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Universal Neonatal Auditory Screening Program in Pakistan - A need of time

Iqbal Hussain Udaipurwala

Hearing is the most significant and essential among all the five senses gifted by God to human beings. Congenital hearing loss is one of the most frequent congenital defects with a incidence of about 30 per 10,000 childrens¹. Significant hearing loss is occurring in 1 to 2 newborns per 1000 in the general population, and 24% to 46% of newborns admitted to neonatal intensive care unit^{2,3}. Hearing loss or impairment is associated with substantial social and psychological consequences. Failure to detect children with congenital or acquired hearing loss may result in lifelong deficits in speech and language acquisition, poor academic performance and personal, social or behavioral problems^{4,5}. Lack of auditory stimulation leads to retrograde degeneration in the cell body and axons of the auditory pathway⁶. Normal speech and language development depend upon a child's ability to hear spoken language and early infancy is the most appropriate time to acquire the foundation of language and communication. The most imperative period for language and speech development is largely regarded as the first three years of life⁷, so, early detection and early identification of hearing loss is very crucial and fundamental. This should be followed by a timely and effective therapeutic intervention and rehabilitation programs to minimize the negative effects of hearing loss on the development of cognitive, psychosocial and verbal communication skills and social interactions. Early intervention by hearing rehabilitation gives positive results in language development. Children undergoing hearing rehabilitation before 11 months of age have a stronger vocabulary and verbal reasoning skills at 5 years of age than those intervened later⁸. Thus, early identification followed by proper intervention as early as six months of age results in essentially normal language acquisition later and minimizes the negative effects of hearing loss^{9,10}. In contrast, a delay in detection of up to 2 to 4 years may result in delayed language acquisition.

World Health Organization estimates that 278 million people worldwide have disabling hearing impairment and two-thirds of these people live in developing countries of which about 50 percent can be prevented¹¹. Hearing loss in newborns and infants are not readily detectable by routine clinical procedures and behavioral observation, although parents often report the suspicion of hearing loss, inattention, or erratic response to sound before hearing loss is confirmed¹². Regardless of the screening method chosen, hearing screening, though critical, is only the first stage of a comprehensive early intervention plan. Screening alone is useless unless appropriate diagnostic testing services and high-quality amplification and rehabilitation services are in place and are implemented in a timely fashion. Early screening does not substitute for further periodic childhood hearing screening. The universal neonatal hearing screening (UNHS) is aimed to identify babies with moderate to profound hearing impairment at birth¹³. It must be considered as a first step of a program for the identification, diagnosis, treatment and habilitation/rehabilitation of children with hearing impairment. Progressive or late onset hearing loss represents a relatively large proportion of childhood deafness and may be not identified by the screening. To early identify those cases an audiological follow-up program is necessary, as well as an audiological surveillance protocol¹⁴. Universal Neonatal Hearing Screening uses electrophysiologic measurements of evoked otoacoustic emissions (OAE) and/or auditory brainstem responses (ABR) to detect hearing loss in newborns and infants. Vibrations of

which is a widely accepted as gold standard measure of hearing sensitivity in newborns and infants.

Both AABR and AOAЕ are noninvasive, rapid screening tests that are rapid and can be performed at the bedside in inpatient or outpatient settings, in term and preterm newborns. The tests are performed on any newborn who is asleep, or at least at quiet rest, in a moderately quiet test environment. Both provide measurements of physiologic activities that are easily recorded in newborns and that correlate highly with the degree of peripheral hearing sensitivity. They are not mutually exclusive, but complementary to each other. Many countries have implemented UNHS as part of early identification and intervention programs. Prior to the implementation of UNHS, only newborns identified as being at high risk such as those in NICU, were screened. In comparison to the resources required for UNHS, substantially fewer resources are required to screen only the high-risk group. However, a major limitation of selective or targeted screening is that as many as 50% of infants with congenital permanent hearing loss have no known risk factor. The relatively high incidence without known risk factors, and the introduction of new and automated screening technology over the last 20 years, has led prestigious bodies in many countries worldwide to recommend UNHS as an alternative to selective/targeted screening. Since 2007, the development and implementation of sophisticated physiological hearing screening techniques has contributed to the feasibility of UNHS worldwide. An increasing number of countries with different healthcare systems and different economic and social circumstances have implemented or are in the process of implementing UNHS programs. Very few studies are carried out in Pakistan about the auditory screening program in neonates^{15,16}. Congenital hearing loss is typically predominant in low-income population. Annually, about 740,000 children (roughly six per 1,000 live births) are detected to have sensorineural hearing impairment in low and middle-income class countries as compared with 28,000 (around two per 1,000 live births) in high income class countries¹⁷. So, it is the time to start universal neonatal hearing screening program all over the Pakistan. It may be difficult to start this program in every hospital and maternity center, but at least one dedicated center can be developed in each region or area that can perform screening of all newborn babies in that locality.

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Universal Hearing Assessment of Neonates: A pressing need of Time.

Iqbal A. Muhammad Khyani

Hearing plays an important role in mental and cognitive development of children. Hearing loss greatly affects linguistic capabilities and speech comprehension. Congenital hearing loss is one of the most common birth defect affecting 2-4 neonates among 1000 live births¹. It has been shown from studies that detection of hearing loss and correction within age of six months leads to better outcomes of hearing and social integration than cases treated later².

Many risk factors have been identified to cause congenital hearing loss that includes genetic factors, maternal factors and perinatal causes. Genetic type of hearing loss makes up to more than 50% of all congenital sensorineural hearing loss³. Much research is being done to identify genetic makeup causing hearing loss when child is still in uterus. Till now, 125 deafness loci have been reported in the literature⁴. Chromosomal studies in intrauterine life and in immediate post natal period can provide information about hearing loss.

Maternal factors such as TORCHS infection (toxoplasmosis, rubella, cytomegaloviruses, herpes type 1 & 2 and syphilis), use of mutagenic drugs during pregnancy and exposure of mother to radiation puts fetus on risk of congenital hearing loss. A test done on viral DNA from neonatal dried blood spots on Guthrie cards and its amplification by means of a nested PCR (DBS test) provides the best identification for use in screening for Cytomegalovirus infection of the new born babies and subsequent treatment for hearing loss⁵. Perinatal stress on infant such as anoxia during birth, prematurity, low birth weight and kernicterus requiring exchange transfusion also account for small amount of cases of hearing loss.

An important issue pertaining to our region of Southeast Asia is consanguineous marriages. Data has shown that there is 76% increased risk for consanguineous marriage children to develop sensory neural hearing loss when compared to non-consanguineous marriage children⁶. Consanguinity has a multiplicative effect on genetic mutation and this practice has to be discouraged actively. Children with syndromic pattern of hearing loss are identified early because of other associated features of a syndrome. Difficulty arises in identifying children with non-syndromic type of hearing loss and those with mild form of hearing loss. A child that is not hearing correctly will have difficulty in understanding speech and delayed development of speech. Such children if not identified and rehabilitated early can be labelled as retarded.

Failure to develop speech by 1 to 2 years of age warrants investigation. Currently, neonatal hearing assessment is only done in "high risk" newborns. Problem with this targeted screening of high risk newborns is that it can only detect 50% of the cases of congenital hearing loss⁷. There is need for universal screening of neonates for earlier detection of hearing loss and providence of rehabilitation.

Objective tests are needed for universal screening of neonates. Otoacoustic emissions (OAE) provide an assessment of inner ear function. It is being used internationally to assess neonatal hearing. Hearing loss above 30 dB can be detected by OAE. Referral rate of 4% was observed in a study for cases screened by OAE in first 48 hours of life⁸. Referred cases are further evaluated by brainstem evoked response audiometry (BERA).

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Bilobed Flaps; A versatile Reconstructive Technique in Fascial Defects Experienced in 50 Cases.

Habibullah Khajjak, Sher Zaman, Shoaib Ahmed Lahri, Munir Ahmed Baloch

ABSTRACT : SUMMARY: Lesions on the face such as Basal Cell carcinomas, Squamous cell carcinoma, moles, post lachmeniac scars and other acquired defects are numerous with great disfigurement and concern for the patient. They also pose a major problem for the clinician, to address the defect properly so that the reconstructive outcome is acceptable to the patient, functionally and aesthetically. In this regards Bilobed flap technique has been used on a variety of defects on different areas of the Body and their utility, advantages, disadvantages and outcome discussed.

Key Words : Cervical lymphadenitis, Lymphadenopathy, Chronic granulomatous inflammation, Tuberculosis.

INTRODUCTION : Skin flaps have been used, for repair of fascial defects since 6th century BC. But more generalized application has been in use since the beginning of century. The earliest description of Bilobed flap is believed to be of Esser in 1918, as a double transposition flap, in the reconstruction of Nasal tip defects^{1,2}. As a re-constructive surgeon we come across cutaneous lesions on the nose, cheeks, and adjoining areas, which pose a serious disfigurement to the patient. Added surgical excision of the defect, specially over nose and adjoining cheeks creates a defect, posing strong challenges to reconstructive surgeon. Use of bilobed flap in this situation allows transfer of skin and soft tissues in an area where

deficient local skin. This technique has been used commonly in reconstruction following resection of malignant disease and defects created after resection of other benign conditions with aesthetic and cosmetic issues. Having advantage over other surgical options such as Full skin grafts and single lobed flaps which are either lacking the natural bulk or can not stand the tension placed on the flap and in conditions where radiotherapy has to be initiated early. Esser described the flap initially consisting of two tongues of a flap, primary lobe and a secondary flap having a common base rotated one behind the other along the long axis of the defect. He emphasized that the primary and the secondary flap should be of the same

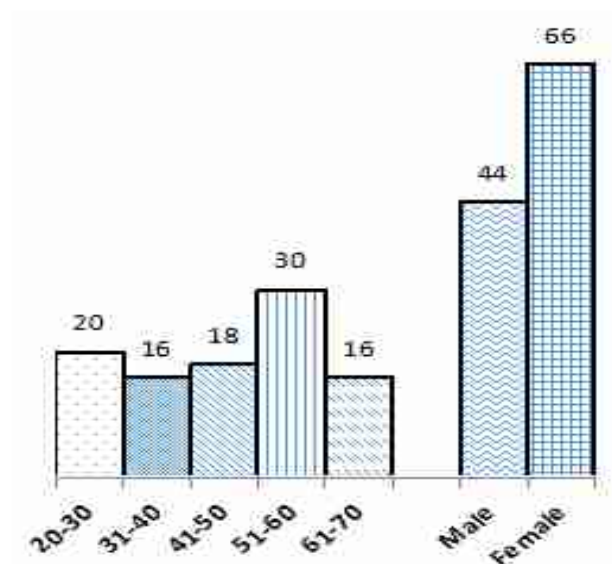


Figure 1 : Age and gender distributions of patients.

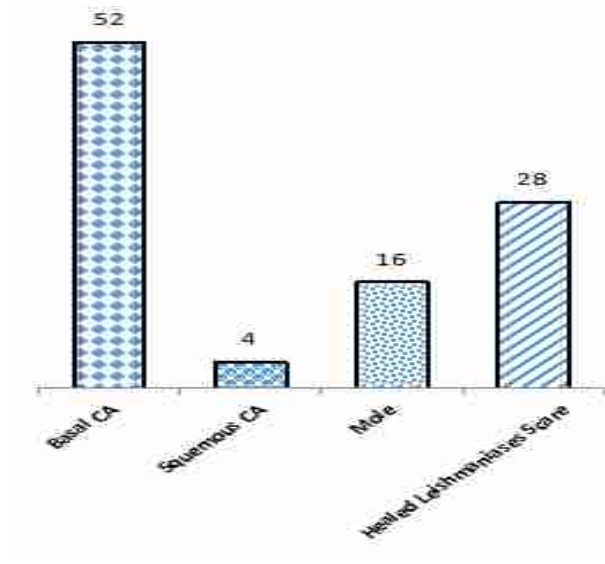


Figure 2 : Sites and cause for bilobed flap reconstructive.

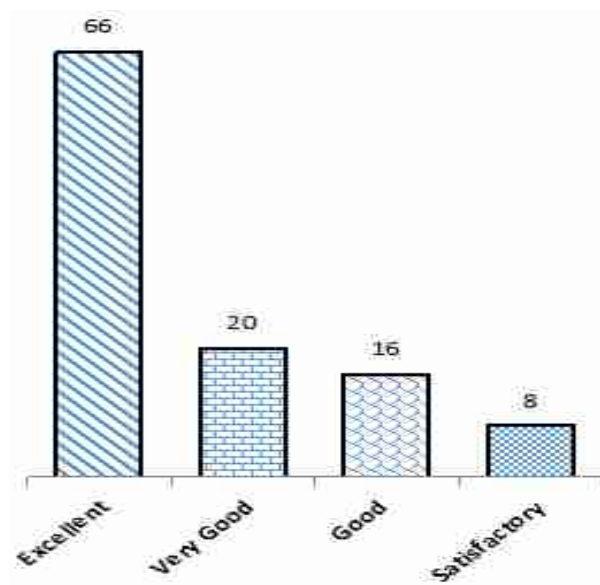


Figure 3 : Patients post operative satisfactory.

flap consisting of two or more flaps or lobes separated around 90 degrees or less based on a common arterial supply and with such dimensions that tension is evenly distributed on all sides. He also added that the angle between the primary and secondary flap may vary between 45- 180 degrees. Gratter the angle between the primary and secondary flap gratter the Dog Ear at the point of rotation. He also felt that the secondary flap should be smaller than the primary flap^{3,6}. McGregor and Soutar also analyzed the design and which resulted in less rotational deformity⁸. The added work by Zetelli suggested an arc of

arc between flaps⁹⁻¹¹. The skin laxity in the area from which flap is to be mobilized can be ascertained by pinching the skin between Thumb and index finger held 1-2 cm apart, this manœuvre will help determine loose and transferable skin^{6,7}.

PATIENTS AND METHODS : This prospective case series was conducted in Bolan Medical Complex Hospital, Quetta. In plastic surgery department and ENT head and neck surgery department during the three years period from January 2013 to December 2015. Patients were enrolled from outpatient department and indoor patients of the respective departments. The sample size was 50 patients (n=50). The sampling technique was convenient sampling. The inclusion criteria was the patient who give the informed consent to be included in this study. Patient who have lesion and needs cosmetic surgery decided by the consultant surgeon. The exclusion criteria was patients who were previously operated with multiple scars make. Those patients who were diagnosed as a case of carcinoma and involvement of the deep muscular or fatty tissues. The selected patients were inquired about other diseases and their complete examination was performed. The selected patients were undergone surgeries and the lesions resected and bilobed flap was performed. The patient was evaluated for the result of the flap. The study variables were the age, gender and primary diagnosis of the disease, site of the lesion, size of the lesion, post-operative complications and results of the flap (excellent, very good, good and satisfactory). The results were analyzed statistically using SPSS version 20 and



A lady with a Basal cell lesion on nose pre, post and follow up photos. Slight post operative dehiscence following infection is noted.
 A Basal cell lesion on the lower Eye lid Excision, and a Bilobed flap reconstruction.
 A Basal cell lesion on the Zygoma Excision bilobed flap reconstruction.

Good: Infection of the flap. Satisfactory: Initial infection and neerosis, bleeding by the secondary infection.

RESULTS: The age-group and the gender distributions are shown in Figure 1. The majority of the cases (30%) were in the age group of 51-60 years. The age group of 31-40 showed the lowest percentage (16%). About two-third subjects (66%) belonged to female patients. Figure 2 illustrates the sites of the Bilobed flap used. More than half (52%) subjects had have Basal cell carcinoma. However, the squamous cell carcinoma showed the lowest prevalence (4%) among the collected samples. Furthermore, healed Leishmaniasis scars belonged to more than one-fourth (28%) of the patients. Patients post-operational responses are shown in Figure 3. About two-third patients were extremely happy with the operation and indicated as 'Excellent' results. Only 8% patients indicated as 'Satisfactory'.

DISCUSSION : The study comprises of application of technique in management of defects, majority of them being Malignant ulcers on Nose, lower Eye lids, cheeks and lips. Size of the defect varied from 1.5 cm to large surface area defects on the Nose, cheek and lips. This flap is mostly used when a defect cannot be closed primarily or adjacent tissues cannot be mobilized or when mobilization of tissues results in gross distortion of regional anatomy. Introduction in majority of the cases, the use of this flap is over the distal nose, here the skin is firmly adherent to the underlying nasal framework, whereas the skin in the middle third and globellar region is comparatively loose allowing mobilization distally in the form of sliding flaps

outcome. Moderate to large sized defects are also reconstructed using Bilobed flap technique with good functional and cosmetic outcome¹². We have presented a broad application of this technique and its utility. Other than these areas this procedure is applicable on other parts of body, with general guide lines.

CONCLUSION : Bilobed flaps are versatile and robust flap. In experience hand, this flap technique has excellent results in reconstruction of fascial defects.

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Comparison of Results of Brain Evoked Response Audiometry with Natural Sleep and after Sedation

Dileep Khatri, M. Iqbal Rao, *Irfan Ahmed Shaikh, *Atif Hafeez Siddiqui

ABSTRACT : *OBJECTIVE:* To compare the results of brain stem evoked response audiometry in patients during natural sleep and after midazolam induced sedation. *SUBJECTS AND METHODS:* The study was conducted at Department of ENT, Peoples University of medical and health sciences, Nawabshah. Total 50 patients were selected below the age of 4 years. The BERA test was performed on these patients two times. Once with natural sleep and later with midazolam induced sedation. The sedation with midazolam was induced in fully equipped operation theatre in presence of qualified anesthetist. *RESULTS:* Out of 50 patients enrolled mean age of patients was 2.48 ± 1.78 yrs. Most of the patients belong to <3yrs of age contributing to 78% cases. There were 28 (56%) males and 22 (44%) females. The results of both methods were almost same in all patients. Although 3 patients required oxygenation during the sedation with midazolam, rest of the patients performed uneventful. *CONCLUSION:* It is concluded that there is no difference between the test results of these two methods. Test with sedation is time saving although there is additional risk of over sedation with midazolam which can easily managed by anesthetist available at that time in operation theatre.

Key Words : Brainstem Evoked Response Audiometry (BERA), Natural sleep, Midazolam Sedation.

INTRODUCTION : Worldwide, Auditory brainstem response audiometry (ABR) is done over the years to analyze functional response to auditory stimulus¹⁻³. It was first discovered in 1971 by Jewet and Williston. It is used to examine integrity of vestibulocochlear nerve, brainstem auditory connections and higher centres to test the sensitivity of hearing^{2,3}. It is usually used when other methods like conditioned play audiometry, hand raising techniques and visual reinforcement audiometry don't reveal conclusive results or these cannot be performed due to developmental status or child's age^{3,4}. ABR process involves transmission from an acoustic transducer i.e; head phone or ear phone of an evoked potential produced by a click which is then measured by electrodes present on the scalp, vertex and ear lobes⁴⁻⁷. The micro voltages are then charted with time in milliseconds. Normally wave I to IV are found in normal hearing individuals. While type V which is strong wave of ABR related to behavioral thresholds. It is hypothesized that longer duration of developmental plasticity in lateral lemniscus and increased myelination with development is associated with speedy connections⁵⁻⁸. Different anesthetic agents and changes in temperature are also associated with increased nerve conduction time⁴⁻⁸. Sedation after midazolam suppresses slow waves and REM sleep in intensive care setting however an oral midazolam decreases stages 1 and 3 and increases stage 2. Increased latencies have been seen in patients with anesthetic use compared

it, and local data statistics are required to bring awareness so that patients can utilize its availability.

SUBJECTS AND METHODS : The study is a comparative study conducted at Department of ENT Peoples University of Medical and Health Sciences, Nawabshah from January 2016 to May 2016. Approval from institutional Ethical committee was taken. Informed consent was taken from patient or from next to kin. Total 50 patients were selected below the age of 4 years of either gender. The BERA test was performed on these patients two times. Once with natural sleep and later with midazolam induced sedation. The sedation with midazolam was induced in fully equipped operation theatre in presence of qualified anesthetist. Patients with mental retardation were excluded.

Patients were sited in a quiet room and were allowed to recline in a comfortable chair and were asked to close eyes in order to relax. For midazolam induced ABR patients were placed in pre-operative area in recovery room after anesthesia review by anesthetist. The lottery method used to divide individuals with n=25 with sedation induced by natural sleep while other n=25 by midazolam. Crash cart was placed together with anesthetist for midazolam induced sleep.

For all patients single recording with electrode placed on forehead, with stimulus given to same side of mastoid or ear lobe is used as inverting electrode while opposite side earlobe is used as ground electrode. The resistance or impedances

10-20milli volts were used.

RESULTS : Out of 50 patients enrolled mean age of patients was 2.48 ± 1.78 yrs. Most of the patients belonged to <3yrs of age contributing to 78% cases (fig.1). There were n=28 (56%) males and 22(44%) females.

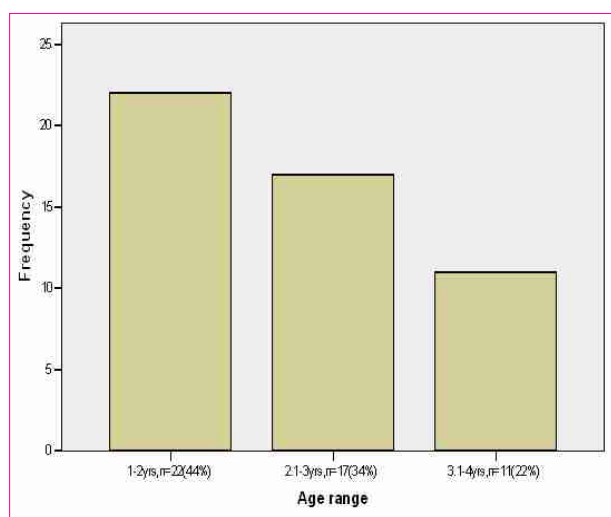


Figure-1 : Age range.

The latencies were classified into I, II, III, III-IV, III-V, I-V. The frequency of wave latencies was 8,14,6,4,14 and 4 respectively. The mean and standard deviation was I, 1.46 (.032), II 3.01(.237), III, 2.21(.980), III-IV 2.41(.109), III-V 2.17(.672) and I-V 4.47(.017) respectively (Table-1)

Variables	I	II	III	III-IV	III-V	I-V
	n=8	n=14	n=6	n=4	n=14	n=4
Mean	1.46	3.01	2.21	2.41	2.17	4.47
Standard deviation	.032	0.237	.980	.109	.672	.017

Table-1 : Mean wave latencies.

Comparison of natural sleep compared to sleep through sedation showed statistically non-significant results with no marked difference in latencies was found. However the latencies of patients undergoing sedation with midazolam appeared to have increased latency periods. The results of both methods were almost same in all patients. Although 3 patients required oxygenation during the sedation with midazolam, rest of the patients performed uneventful.

DISCUSSION: Auditory brainstem response audiometry is a potent non-invasive, reliable clinical assessment technique for hearing impairment mainly under 3 years of age^{1,2,10-14}. Good relaxation and therefore of highest quality recording were obtained with sedation induced by midazolam otherwise inaccuracy and threshold overestimation will occur due to

between sedation through natural sleep and by midazolam. Different studies have shown brain evoked potentials to be very useful parameters for assessment of hearing impairments in addition to audiometry¹¹⁻¹³. It is useful for assessment of deafness due to, peripheral and central nervous system, and in intensive care units. In our study different wave latencies were compared between two groups. There were increased frequency of wave II and III-V seen most commonly in study population. In our study there is slight predominance of males compared to females with male to female ratio of 1.3:1. The age most commonly being tested was <3yrs. reported study has found females to have shorter A wave latencies compared to males¹⁵. Another study has found increased incidence among males compared to females with ratio of 2:1 and above 5 years in 82.65%⁷. Hearing loss was most commonly profound 29.79% compared to milder and moderate cases (4.57%, 7.52%) respectively. In the above study it was found 25.40% mixed cases of conductive and sensorineural hearing loss compared to cases alone. In our study mostly enrolled patient have milder to moderate hearing impairment, In our study the sleep latencies of different wave forms were compared and did not show any statistical significance compared to each other. The sleep latencies were slightly prolonged in patients with sedation under midazolam however normal natural sleep latencies were found in rest of patients. A reported study has also not found any statistically significant result and concluded different other factors responsible for prolonged sleep latencies¹. The results of both methods were almost same in all patients. Although 3 patients required oxygenation during the sedation with midazolam, rest of the patients performed uneventful.

CONCLUSION: It is concluded that there is no difference between the test results of these two methods. Test with sedation is time saving although there is additional risk of over sedation with midazolam which can easily managed by anesthetist available at that time in operation theatre.

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High Frequency of Oral Sub Mucous Fibrosis in Gutka Eating Subjects of Mirpur Khas, Sindh

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ABSTRACT : BACKGROUND: Oral Submucous Fibrosis (OSMF) is a pre-cancerous condition of the oral cavity. Chewing of gutka is associated with OSMF. In Pakistan majority of adolescents are indulging in gutka chewing habits which is regarded as potent cause of OSMF. **SUBJECTS AND METHODS:** A cross sectional study was conducted in department of ENT at a tertiary care hospital of Mirpurkhas Sindh from January 2010 to December 2013, using convenience sampling technique. Gender consuming areca nut, gutka, mainpuri, or paan chewers for atleast one year on a daily basis were included in the study. Patients suffering from oral cancers, smokers and naswar users were excluded. Diagnosis of OSF was based on clinical examination. Data was collected using purpose made questionnaires. Approval was taken from institutional ethical review board. **RESULTS:** A total of 106 patients participated in this survey. Most of the patients belong to age group 31 to 40 years (40%), with marked male preponderance (81%). Out of 106 patients, 51(48.11%) participants were consuming gutka, 34(32.08%) were using areca nuts, 13(12.26%) were paan chewers, 8(7.5%) were mainpuri chewers. Total of 21(14%) patients were diagnosed with OSMF. Statistically significant difference was found in Hemoglobin levels and duration of oral problem amongst patients with and without fibrosis. Strong association was found between intake of gutka and OSMF. **CONCLUSION:** The results of this study suggest that the frequency of OSMF in Mirpurkhas was significantly high as compared to other available reports. Habitual chewing of the commercially available gutka (mixture of areca nut, tobacco and lime) was the most common factor associated with the development of OSMF.

Key Words : Oral submucosal fibrosis, Areca nut, Smokeless tobacco, Betel quid.

INTRODUCTION : Worldwide it has been observed that 58% cancers of head and neck occurs in South Asia and Southeast Asia¹. Throughout the world, greater than 500,000 incident cases and 320,000 fatalities occur on a yearly basis from oral cancer². According to studies in Pakistan Oral cancer ranks second most common cancer in both genders³. Oral submucous fibrosis (OSMF) is a condition that is chronic, progressive and possesses pre-cancerous potential for the oral cavity⁴. OSMF has been well recognized for its malignant tendency⁵. OSMF is insidious, non-reversible, crippling disorder of the oral cavity characterized by fibrotic change with severe burning sensation and limited mouth opening⁶. In Pakistan, studies have shown that 40% of adolescents and adult population residing in Karachi were consuming minimum one product of chewable tobacco on a routine basis⁷. In Pakistan and South Asian regions, the most common and frequently used chewing products are tobacco, betel quid, chalia(betel nut), gutka and niswar⁸. Eating betel quid, gutka, niswar and tobacco is acceptable in Pakistan as it is taken a usual cultural practice⁹. Chewing of these substances has been well established with

rural population¹¹.

Significant difference has been observed in the prevalence of OSMF among different communities, regions, and countries which are related directly to the utilization of areca nut, age of first encounter and differences in availability of product¹². Keeping the above points in view the present study was carried out with the aim to determine the frequency of OSMF and its associated risk factors in patients coming to the tertiary care government hospital of Mirpurkhas Sindh.

SUBJECTS AND METHODS : This cross sectional survey was conducted on total of 106 patients visiting outpatient clinics of ENT at tertiary care hospital of Mirpurkhas, Sindh. The study period was from January 2010 to December 2013. Sample was inducted in the study using convenience sampling technique. Patients of both genders consuming Areca nut, gutka, mainpuri or pan for at least one year on a daily basis were included in the study. Patients who already had oral cancer, naswar users and smokers were excluded from the study. Data analysis was conducted on SPSS version²⁰. For numerical variables descriptive analysis was carried out and then independent t test was used

categorical variables was taken out through application of Chi square test. P value less than 0.05 was taken as significant. Permission was taken from the institutional ethical review board. **RESULTS :** A total of 106 patients were inducted in this study. Out of total, 15(14.2%) participants were less than 20 years, 42(39.6%) patients were 21-30 years of age, 35(33%) participants were 31 to 40 years of age and 14(13.2%) patients were greater than 40 years of age (Table-1). There were 86(81.1%) males and 20(18.9%) females (Fig-1). Out of 106 patients 51(48.11%) patients were chewing gutka, 34(32.08%) were in the habit of consuming areca nuts, 13(12.26%) were pan chewers, 8(7.5%) were chewing mainpuri. Table-2 demonstrates the association between different chewing habits and fibrosis status of participants. However, a positive association was only found with gutka while no association was observed for areca nuts, mainpuri and paan chewers. Patient characteristics presenting with and without fibrosis were compared using Independent t test. Out of 21 patients with OSMF, 11(52.3%) had involvement of buccal mucosa, 9

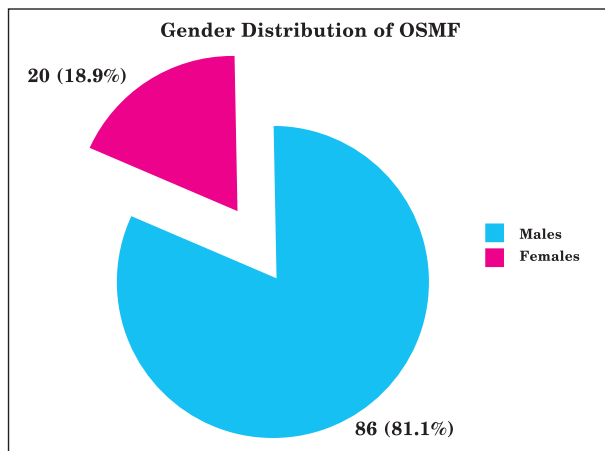


Figure-1 : Gender distribution of oral submucous fibrosis.

Age Range	< 20 Years	21- 30 Years	31- 40 Years	> 40 Years
No of Cases	15(14.2%)	42(39.6%)	35(33%)	14(13.2%)

Table-1 : Age range.

Variables	With Fibrosis		Without Fibrosis		P-Value
	n	%	n	%	
Areca Nuts	6	28.6	28	32.9	0.57
Gutka	11	52.4	42	49.5	0.05
Manpuri	2	9.5	4	4.7	0.19
Pan	2	9.5	11	12.9	0.88

Table-2 : Habits of participants with and without fibrosis.

Site of OSMF	No. of Patients (Percentage)
Buccal Mucosa	11(52.3%)
Soft Palate	9 (42.9%)
Tongue	1 (4.76 %)
Total	21 (100 %)

Table-3 : Site involvement of OSMF.

DISCUSSION : Oral submucous fibrosis is a disabling condition which is common in our subcontinent with an incidence of 0.2-1.2 %¹³ Studies have elucidated that this is not a new disease for this part of the world¹⁴. In our study frequency was high as 19.9% of consumers were seen to have developed fibrosis. In this study majority of the participants were 20 to 40 years of age group. Higher usage of gutka in young generation has been shown by reported studies¹⁵. A study conducted in India demonstrated mean age of OSMF as 38.8 years with cases ranging from 22-61 years of age¹⁶. Another study in Patna, India showed similar results with age range from 21-40 years¹⁷.

In our study 52.4% participants with fibrosis were consuming gutka whereas 26.4% was utilizing areca nut with 9.5% cases of betel quid (Pan) chewing. Gutka eating habit is part of culture in South Asia with children showing predilection towards this habit¹⁸. Our study supported previous data where 24% of gutka consumers had OSMF. Another study from India showed high relative risk ($p < 0.01$) of OSMF with gutka¹⁹. Betel quid is used by 10-20% of global population and approximately 600 million users consume areca nut²⁰. Areca nut association with OSMF has been well established by various studies^{21,22}.

Other studies have stated that areca nut/betel quid is the major risk factor for OSMF with evidence from epidemiological studies in last 40 years²³. However gutka is related to higher severity and faster occurrence of OSMF. In one study mean duration of OSMF development among gutka chewers was 8.2 years, and much earlier compared to habit of betel quid and areca nut users (21.8 years and 10.9 years respectively¹⁶. In our study most OSMF was seen in oral cavity and soft palate. It is considered as pre-malignant and pre-neoplastic condition that may convert to OSCC. Studies in Karachi have shown cancer development mostly in buccal mucosa while in western countries primary areas of involvement are tongue and floor of the mouth^{24,25}.

Our study had few limitations. First the sample size was limited. We utilized a convenience sampling technique with a cross sectional design which makes it prone to bias. Interaction among different substances was not seen. However the strengths cannot be ignored. Proper oral examination of participants was performed by consultants with confirmation based on lab results. This is one of the first studies on OSMF in this area as few studies have been conducted in this district of Sindh.

Early diagnosis and treatment is the only cost effective method for improving survival rate and life quality of patients suffering from OSMF

increasing tax, halt advertisements and control smuggling through law enforcement. There is urgent need for primary prevention among high risk people and different communities²⁹.

CONCLUSION: The results of this study suggest that the frequency of OSMF in Mirpurkhas was significantly high as compared to other available reports. Habitual chewing of the commercially available gutka was the most strongly associated factor with the development of OSMF.

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Increasing Incidence of Dealing Paraphenylene Diamine (Kala Pathar) Poisoning at Tertiary Care Hospital of Nawabshah

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**Iqbal A. Muhammad Khyani, **Nargis Izhar, **Sharjeel Anis

ABSTRACT : *OBJECTIVE:* The aim of our study is to see the incidence and outcome of Paraphenylene Diamine (PPD) Kala Pathar poisoning in our Hospital. *STUDY DESIGN:* Prospective observational. *SUBJECTS AND METHODS :* Subjects with PPD poisoning admitted in Surgical ICU of Peoples University of Medical Health Sciences (PUMHS) Hospital Nawabshah, between 2014 to 2015 were included in our study. The presentation and outcomes of the patients, after management of PPD poisoning was recorded on approved performa. *RESULTS:* 50 cases of PPD poisoning were encountered during the period of one year from 2014 to 2015 at PUMHS Hospital Nawabshah. Out of 50 subjects included 45 (90%) were females and 5 (10%) were males. After examination and complete management of patients, it was found that out of 50 patients 25 (50%) patients cured and 25 (50%) patients died during the management. Tracheostomy was done in 30 patients to relieve the respiratory obstruction. *CONCLUSION :* Poisoning due to PPD was associated with high mortality. Therefore, early management is lifesaving. There is no specific antidote for PPD. Asphyxia is the major early challenge, which require tracheostomy and ventilatory support.

Key Words : Paraphenylene diamine (PPD) poisoning, Kala pathar (black stone), Tracheostomy, Suicide.

INTRODUCTION : Chemically, Kala Pathar is a derivative of Paraphenylene Diamine. It is a brown or black colored solid substance, easily soluble in hydrogen peroxide and not in water^{1,2}. PPD poisoning is amongst one of the emerging causes of poisoning in Asian countries^{2,3}. It is easily available in market at a low cost. Kala Pather (PPD) is crushed and mixed with henna to increase the intensity and fastness of color of henna^{3,4}. It is also used in hair dye for the same purpose since more than a century^{5,6}. PPD compound is highly toxic when taken orally. Death occurs within 24 hours of ingestion due to multiple organ dysfunction. It causes allergy and renal tubular toxicity leading to angioneurotic edema and rhabdomyolysis⁷. First PPD poisoning was reported in 1924 in a hair salon owner^{5,6}. Common presenting features are edema of face, neck, tongue, larynx, sinus bradycardia, sinus tachycardia and acute renal failure⁸.

SUBJECTS AND METHODS: The study was conducted at department of ENT, Head and Neck Surgery of PUMHS Hospital, Nawabshah, Sindh, Pakistan between 2014-2015. Consecutive cases of PPD (kala pathar) poisoning encountered in emergency of PUMHS hospital and admitted in SICU for management were included after taking informed consent from the attendants of patients. Cases where consent could not be taken and SICU admissions other than PPD poisoning were

made IRB approved performa. The patients diagnosed as PPD poisoning and were admitted in SICU were attended by senior consultant on call for the assessment of airway and for making the decision of intubation/tracheostomy. The data was entered and analyzed using SPSS program version 16 (SPSS Inc. Chicago, IL, USA). P-value of 0.05 was taken as a standard reference. **RESULTS:** Total of 50 cases of Paraphenylene Diamine (PPD) poisoning were included during the period of one year from 2014 to 2015. Out of 50 patients, 45 were females and only 5 patients were males (Fig-1)

Age range was from 15 to 50 years. Majority of the patients presented with cervico-facial edema, dysphagia, sore mouth and throat. Dark colored urine, rhabdomyolysis, acute renal failure and hemodynamic shock were also presented as depicted in Table-1

Tracheostomy was performed in 30 (60%) patients by senior consultants to secure the airway as a part of management. Out of 30 tracheostomized patients, 25 (83.33%) patients survived and 5 (17.7%) died (Fig-2)

DISCUSSION: Paraphenylene diamine (Kala Pather) is one of the common constituent of hair dye formulations. It is a coal-tar derivative. Kala Pathar is emerging as a new trend in suicidal poisoning due to easy availability, low cost and salty taste rather than bitter^{9,10}. The symptoms are dose related and patients with ingestion of

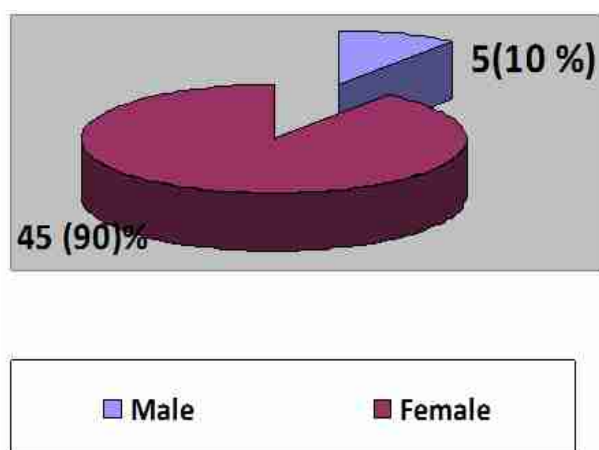


Figure 1 : Gender distribution.

S. No.	Clinical Feature	Patients
1.	Cervico facial Edema	50 (100%)
2.	Dysphagia	50 (100%)
3.	Difficulty in opening of mouth	50 (100%)
4.	Dark Urine	25 (80%)
5.	Rhabdomyolysis	15 (30%)
6.	Acute Renal failure	09 (18%)
7.	Hemodynamic Shock	09 (10%)

Table 1 : Clinical features of Kala Pathar Poisoning.

Poisoning can lead to many symptoms, like angioneurotic edema, sore mouth and throat, acute respiratory failure, rhabdomyolysis, myocarditis, arrhythmias, renal tubular, necrosis, intravascular hemolysis and hypotensive shock¹². Cervico-facial edema was the first symptom to develop. Respiratory failure is the main threat to life. Endo-tracheal intubation, tracheostomy and assisted ventilation are crucial and life saving measures¹³. There is no specific antidote available for this poisoning. PPD is frequently used in Henna and hair dye. Contact allergy to this compound is well recognized problem¹⁴. A study from India showed that increase need of ventilatory support, duration of hospital stay and mortality was dependent on the dose taken by the patients¹⁵. In this study, we could not measure and record the actual amount consumed by the patients due to lack of resources and illiteracy amongst the patient's families presented with the suicidal attempts. It is assumed that patient who could not survive had taken large doses of the poison¹⁶. In a study from morocco during 1996 to 2007 found that, 14.7% patients died due to PPD poisoning out of 102 cases¹⁷. In a seven years retrospective study it was found that the mortality rate was 16% in total¹⁸. Early diagnosis of PPD poisoning and prampt decision for endotracheal intubation or tracheostomy to main join and support airway is essential in terms of

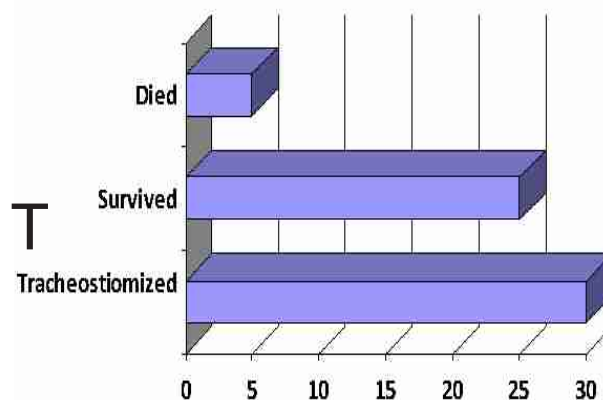


Figure 2 : Survivals In Tracheostomized Cases.

increasingly utilized for suicidal attempts due to its unrestricted free availability, low cost and salty taste rather than bitter. Human consumption is highly toxic and there is no specific antidote available. Therefore early management is life saving. Asphyxia is the major early presentation. Airway should be closely monitored and maintain with timely endotracheal intubation or tracheostomy¹⁹. Most of the reports about PPD poisoning have been reported in Africa and Asia, and its importance need to be highlighted further in view of use of Henna in our society²⁰.

CONCLUSION: There is alarmingly increased cases of Kala pathar poisoning. It is increasingly utilized for suicidal attempts as it is freely available, having low cost and by virtue of its salty taste rather than bitter. Human consumption is highly toxic with high mortality rates. Therefore, early management is lifesaving. There is no specific antidote. Asphyxia is the major early challenge, which required tracheostomy and ventilatory support.

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Experience of Managing Branchial Cleft Anomalies at Civil Hospital Karachi

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ABSTRACT : INTRODUCTION : The branchial apparatus appears between the fourth and fifth week of fetal development. It consists of six paired branchial arches separated by brachial clefts externally and branchial pouches internally. In human branchial cleft contribution to the formation of variety of head and neck structures including cervical skin, jaw, middle and external ear. **STUDY DESIGN:** Retrospective analytical. **SETTINGS:** Department of ENT, Head & Neck surgery, Dow Medical college Dow University of Health Sciences Karachi. **DURATION:** January 2004 to January 2013. **SUBJECTS & METHODS:** A total of 22 cases of branchial cleft anomalies were identified who visited outpatient department. They were diagnosed on the basis of history, clinical examination including location of the lesion and radiological investigation including sino-fistulogram and / or CT scan for cystic lesion. **RESULTS:** Out of total 22 subjects, 14(63.6%) were males and 8 (36.4%) were females, with male to female ratio of 1.75: 1. On presentation majority of the cases (n-17) were having cervical sinus ; 4 cases presented with cyst out of which 03 cases were infected with history of incision and drainage. Only one case of clear cut fistula was identified. The age range was between 15- 35 years with a mean of 19.4 years. Majority were second arch defect with cervical sinus or cyst with only one case having third arch defect with fistulous communication between pyriform fossa and cervical skin. First arch defect was not identified in present series. All the cases were managed surgically with standerd stepladder technique. Recurrence was recorded in 02 (9%) cases on long followup. **CONCLUSION:** Branchial cleft anomalies are rare and not confined to pediatric population. Second brachial cleft anomalies are the commonest of the true Branchial cleft abnormalities and the diagnoses is usually straight forward. Sinuses are more frequent than cyst and fistulae are extermely rare. The management of this rare lesion is challenging and requires knowlegge of the embryological anatomy of branchial apparatus. With the classical approach complete excision must be ensured.

KEYWORDS: Branchial cyst, Congenital anomalies of neck, Benign neck swellings.

INTRODUCTION : The term branchial cyst was first used by Ascheron in 1832. He suggested that these cysts were results of impaired obliteration of branchial clefts. Incomplete, failed or persistent embryonic development of these arches results in several anomalies or defects in the neck^{1,2}. The most widely held belief is that incomplete obliteration of the cervical sinus plays an important role in this process³. Theories of origin of branchial cysts includes. Branchial apparatus theory suggests that branchial cysts represent the remains of pharyngeal pouches or bronchial clefts or a fusion of these two elements. The cervical sinus theory suggests that branchial cysts represent the remains of the cervical sinus of His. The thymopharyngeal duct theory suggests the cysts are remnants of the original connection between the thymus and third branchial pouch. The inclusion theory theory suggests that branchial cysts are epithelial inclusions within a lymph node^{4,5}. Branchial cleft anomalies are the second most common head and neck congenital lesions seen in children⁶.

without any side predilection, right or left in which they develop⁸. They are located in the cervical region, parotid and mediastinum. Many developmental anomalies of the branchial apparatus have been identified: cysts, fistulas, sinuses, ectopic glands, and malformations of head and neck structures. Branchial cleft cysts are congenital lesions often presenting as lateral neck masses.

The branchial cysts were classified into four types. First branchial cleft cysts are divided into type I and type II. Type I cysts are located near external auditory canal. Most commonly, they are inferior and posterior to the tragus. Type II cysts are associated with the submandibular gland or found in the anterior triangle of the neck. The second branchial cleft accounts for 95 percent of branchial anomalies. Most frequently, these cysts identified along the anterior border of the upper third of sternocleidomastoid muscle, adjacent to the muscle. However, these cysts may presents anywhere along the course of the second

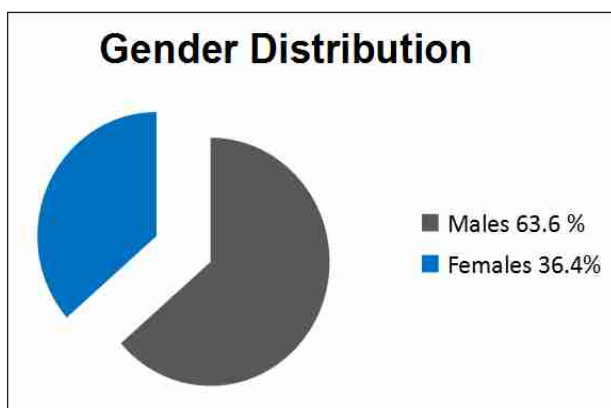


Figure 1 : Gender Distribution.

cleft anomalies are the commonest of the true branchial cleft abnormalities, and the diagnosis is usually straightforward¹⁰. Good knowledge of anatomy and embryology are necessary for proper treatment¹¹. Of the second branchial cleft anomalies, sinuses are more frequent than cysts, and fistulae are extremely rare¹². Bailey classified second BCCs into four types. The Bailey type I cyst is the most superficial and lies along the anterior surface of the sternomastoid muscle, just deep to the platysma muscle. The type II cyst is the most common and is found in the classic location for these cysts, which is along the anterior surface of the sternomastoid muscle, posterior to the submandibular gland. A type III cyst extends medially between the bifurcation of the internal and external carotid arteries to the lateral pharyngeal wall. The type IV cyst lies in the pharyngeal mucosal space and is lined by columnar epithelium¹³. The third and fourth branchial cleft cyst are rare. Cysts that originate from the second or third cleft are in anatomic intimacy with vital neck structures such as the carotid artery, jugular vein and hypoglossal nerve¹⁴. Branchial cleft cysts are benign; however, superinfection, mass effect, and surgical complications account for morbidity. Surgical excision is the definitive treatment for branchial anomalies and is often performed without complication^{15,16}. The rationale of this retrospective review is that, all the admitted cases of branchial cleft anomalies in our setup should be analyzed for presentation and surgical treatment of these patients in our setup.

SUBJECTS AND METHOD: Study was conducted at department of ENT, Head & Neck Surgery, Dow University of Health Sciences & Civil Hospital Karachi. Twenty two patients of suspicious branchial cleft anomalies of any age and sex who were admitted in ENT ward of Civil Hospital Karachi were included in the study from January 2004 to January 2013. All who were treated elsewhere and having no documentary

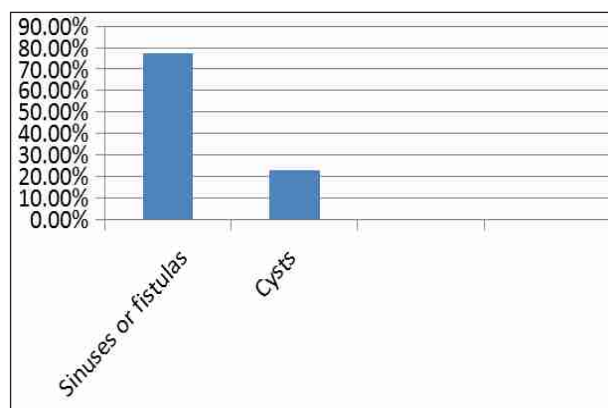


Figure 2: Comparison of branchial cyst and sinuses or fistulae.

proof, recurrent or residual diseases after surgery were excluded from the study. Demographic data, clinical findings of branchial cleft anomalies (site whether right or left side of neck, location on sternocleidomastoid muscle, site of internal and external opening of fistulous tract) & sinogram findings if needed were recorded. All cysts were excised externally through neck while sinuses and fistulae were excised with standard steppled incision. The extent of neck incision for branchial cyst depends upon its size while number of incisions for branchial sinus and fistulae depends upon their vertical length.

RESULTS: A total of 22 cases, out of which 14 (63.6%) were male and 8 (36.4%) were females with male to female ratio of 1.7: 1(Fig 1). The age range was between 15- 35 years with a mean of 19.4 years(77.27%). On presentation majority of the cases (n-17) were having cervical sinus; 4



Figure 3 : Branchial sinus (a) with sinogram (b) on right side of neck

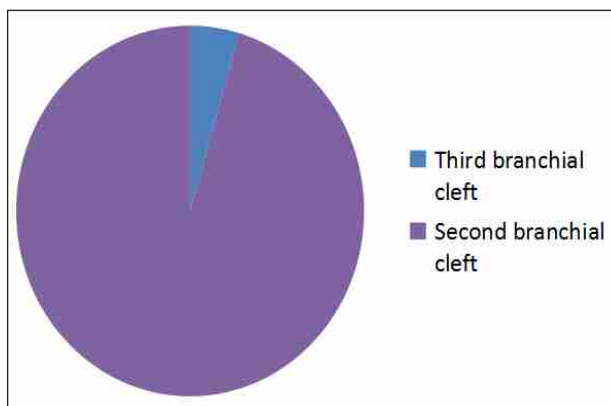


Figure 4 a: Frequency of different branchial clefts.



Figure 4 b :



Figure 5: Peroperative photograph of branchial cyst and (b) fistula with tract.

cases presented with cyst out of which 03 cases were infected with history of incision and drainage (Fig 2). Only one case of clear cut fistula was identified. Majority of the cases n=21 (95.45%) were second arch defect with cervical sinus or cyst. 4 (18.18%) cases presented with branchial cyst located at upper 1/3rd of anterior border of SCM (Fig 3). 17 cases (77.27%) presented with off and on discharge from sinus located at junction of upper 2/3rd & lower 1/3rd of anterior border of SCM. Only one (4.54%) case have third arch defect with fistulous communication between pyriform fossa and cervical skin (Fig 4). First arch one defect was not identified in present series. All the cases were managed surgically. Cysts with its track excised completely through neck. Sinuses & fistulas were excised with standard stepladder surgical technique (Fig 5). Recurrence was recorded in 02 (9%) cases on long followup.

DISCUSSION: Ninety-five percent of abnormalities of the branchial cleft apparatus arise from the second cleft. At least 75% of all second branchial cleft abnormalities are cysts¹⁷, which typically present when an individual is between 10 and 40-years-old. Second branchial cleft fistulas and sinuses are less common and usually present during the first decade of life¹⁸. No gender prediction has been reported¹⁹. In our study approximately 81.81% of the patients had branchial sinuses (there was one case of branchial fistulae) and the rest of 18.19% had branchial cyst, contradicting thus the speciality literature data that show a greater incidence of branchial cysts than branchial sinuses and fistulae. Besides, most of the patients belonged to the first and second decades-70%-(specialty literature data show a higher frequency of branchial cysts at adults-decade two and three). Our data showed more male predominance (male female ratio 1.7: 1). Branchial cleft abnormalities are usually small, but can enlarge enough to cause difficulty swallowing and breathing. Sixty percent of branchial cysts are located in the upper third of neck, at the anterior margin of sternocleidomastoid muscle²⁰. Small lump or fluctuant masses in the lateral portion of the neck adjacent to the anteromedial border of the sternocleidomastoid muscle at the mandibular angle²¹⁻² (usually only one side of the neck, rarely on the both sides; usually painless, unless infected). There is another symptoms: hoarseness (attributed to palsies of cranial nerves IX, X and XII), a bulge in overlying pharynx²³⁻⁴. Palsy of cranial nerves IX, X, XI or XII is extremely rare in benign tumors or cysts and usually suggests direct infiltration of local malignancies or metastatic lymph nodes²⁵⁻⁷. Small opening in the skin that drains mucus or fluid near the front edge of the sternocleidomastoid

muscle. The ostium is usually noted at birth just above the clavicle in the anterior neck²⁷. Third and fourth branchial sinus and fistulae are more rare, opening low in the neck and pyriform fossa²⁸⁻⁹. Besides complete clinical examination, ultrasound neck and fistulography were needed to obtain an accurate positive diagnosis. At ultrasonography, a second branchial cleft cyst is seen as a sharply marginated, round to ovoid, centrally anechoic mass with a thin peripheral wall displaces the surrounding soft tissues. The mass is compressible and shows distinct acoustic enhancement. Occasionally, fine, indistinct internal echoes, representing debris, may be seen.

At CT, these cysts are typically well circumscribed, homogeneously hypoattenuated masses surrounded by a uniformly thin wall²². The mural thickness may increase after infection. The cyst typically displaces the sternocleidomastoid muscle posteriorly or posterolaterally, pushes the vessels of the carotid space medially or posteromedially, and displaces the submandibular gland anteriorly²⁷. It may also be seen more medially within the parapharyngeal space after extending through the stylomandibular tunnel and middle constrictor muscle³⁰. MR imaging better depicts the deep tissue extent of a second branchial cleft cyst, which allows accurate preoperative planning. The cyst fluid varies from hypointense to slightly hyperintense relative to muscle on T1-weighted images and is usually hyperintense on T2-weighted images²⁰.

All patients of suspected branchial cyst should be investigated with at least an fistulogram and ultrasound guided FNAC³¹. For the studied group, as paraclinical exploration we did sinogram-18 cases (the patients diagnosed with branchial sinuses) and ultrasonography-4 cases (the patients with branchial cysts). The ultrasonographic aspect of the cervical tumor, that of cystic formation, facilitated the diagnosis very much. Some authors recommend radiology studies (CT, MRI)³²⁻⁴. At the studied group, we did not make such advance imaging studies as the clinical data; ultrasonography and sinogram were considered sufficient in diagnosing the patients accurately. Pathologically characteristic branchial cysts are usually filled with a turbid, yellowish fluid that may contain cholesterol crystals. Their walls are thin and lined with stratified squamous epithelium overlying lymphoid tissue³⁵. Columnar respiratory epithelium is occasionally present. Management of second branchial cleft anomalies is surgical excision³⁶. Surgery should ideally be performed on the uninfected neck. Previous authors have described the oral approach to management of a second branchial fistula.

Stripping of the fistula was first described by Heanley in 1976, but this requires a correct diagnosis and complete fistula with both internal and external openings, an extremely rare clinical occurrence³⁷. Surgical management of second branchial sinus presents challenge, partly because, unlike the complete branchial fistula, there is difficulty in visualizing the cephalad end of the sinus. As such, surgical excision using the classical approach is analogous to a partial amputation, without the benefit of including, within the excised tissue, the blind tonsillar fossa ending. This may explain the high recurrence rates recorded in some series. The combined approach technique overcomes this limitation by ensuring complete excision of the branchial sinus tract. With the classical approach, complete excision must be ensured but this is doubtful in some cases as recurrence rates following complete excision can be as high as 22 percent³⁸. Olusesi also described the combined approach branchial sinusectomy which ensures the complete excision of the entire tract of the branchial sinus³⁹. The studied patients were performed complete excisions of fistulous or sinus tracts (18 cases) and there was no recurrence within 1-5 years of followup.

CONCLUSION: Branchial cleft anomalies are rare and not confined to paediatric population. Second branchial cleft anomalies are the commonest of the true Branchial cleft abnormalities and the diagnosis is usually straight forward. In second branchial cleft anomalies, sinuses are more frequent than cyst and fistulae are rare. The management of this rare lesion is challenging and requires knowledge of embryological anatomy of the branchial apparatus. With the classical approach complete excision must be ensured.

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Clinical Profile of Oral Pre-neoplastic Lesions at Tertiary Care Hospital

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Irfan Ahmed Shaikh, M. Umar Farooq

ABSTRACT : *BACKGROUND* Oral pre-neoplastic lesions are at rise in our population and alarming numbers of these Pre-malignant lesions (PML) are being documented in younger adults in recent years. The major risk factors in our region for these lesions are the increasing use of areca nut (chalia, supari, betel nut), betel quid with or without tobacco and poor nutrition. Wide majority of them may convert to frank squamous cell carcinoma. **OBJECTIVE:** To determine the clinical profile of oral pre-neoplastic lesion in patients presenting at tertiary care hospital. To determine the frequency of different oral pre-neoplastic lesions in our etiological distinct population. **STUDY DESIGN:** Non-interventional observational study. **PLACE OF STUDY:** Department of ear, nose, throat, head and neck surgery, Dow Medical College (DMC), Dow University of Health Sciences (DUHS) and Civil Hospital Karachi – Pakistan. **DURATION:** One year from July 2015 to June 2016. **SUBJECTS & METHODS:** Total of 400 consecutive cases were included in the study, detailed history and clinical examination was performed and findings were recorded on performa. **RESULTS:** Out of 400 cases, 305 were males and 95 were females. Age range was 14-65 years with the mean of 28 years. Oral submucosal fibrosis was observed in majority of the cases (n-232) followed by leukoplakia seen in 108 subjects. Erythroplakia was found in 45 cases and 15 subjects were diagnosed as lichen planus. **CONCLUSION:** It is a common observation that males are more susceptible to oral PML than females. One of the probable reasons of male preponderance is that females are less privileged to access for healthcare and diagnosis due to socio-cultural reasons being a male dominated society. Majority of the oral PMLs are habit related and poor oro-dental hygiene was observed in those cases.

Key Words : Pre-malignant lesions, Leukoplakia, Erythroplakia, Lichen planus, Oral submucous fibrosis.

INTRODUCTION : Pre-malignant lesions also referred to as pre-cancerous or pre-neoplastic lesions arises frequently from oral epithelium and these epithelial lesions precedes and ultimately converts to OSCC in 90% of the cases. Classification of these lesions of oral cavity followed in day to day clinical practice is typically based on clinical appearance to determine its type¹. World Health Organization (WHO) defines leukoplakia as “white patch or plaque that cannot be characterized clinically or pathologically as any other disease”. Precise definition of leukoplakia is under continuous refinement for long in an attempt to distinguish benign from pre-malignant lesions. It has no distinctive histologic features that define it as a unique histopathologic process. Leukoplakia has been classified in clinical subtypes as simplex, verrucous, erosive, granular / nodular and proliferative verrucous leukoplakia (PVL)². Erythroplakia is a clinical term used to describe a velvety red patch that cannot be clinically or pathologically distinguished as any other definable disease. Similar to leukoplakia, the erythroplakic lesion is considered as a diagnosis of exclusion because numerous other disease entities must be excluded before erythroplakia

lateral tongue, retro-molar pad and soft palate^{3,4}. Lichen planus is a muco-cutaneous disorder affecting 0.5 to 1% of population worldwide⁵. These are heterogeneous group of muco-cutaneous lesions varies in spectrum from hyperplasia to dysplasia and hyperkeratosis. Mucosal lichen planus is more common than cutaneous type which may persist for more than 20 years without spontaneous remission. Common mucosal sites of oral cavity involved are buccal mucosa, hard palate, retro-molar trigone, lips. Different forms of lichen planus have been described; strial type, plaque like, reticular type, ulcerative type, erosive type, hypertrophic type, vesicular and bullous types etc⁶. Oral submucous fibrosis (OSF) is a chronic progressive condition found frequently in people of Asian descent. This pre-cancerous lesion is thought to be caused by areca nut, catechu, betel quid along with slake lime used for recreation in these geographic regions. OSF is considered to be due to consumption of areca nut products with resultant disruption of extracellular matrix affecting collagen synthesis. OSF may leads to changes in fibro-elasticity of oral mucosa causing burning sensation, spice intolerance and restricted mouth opening. The condition often

trismus⁷. OSF has been tried to be staged as; stage-1 stage of stomatitis, stage-2 stage of fibrosis and stage-3 sequelae of fibrosis⁸. **SUBJECTS AND METHODS:** This is a non-interventional observation study of 400 cases of pre-malignant lesions. The subjects included were the cases of any age, sex or ethnic group presented in outdoor department of ENT, Head and Neck Surgery, Civil Hospital, Karachi during 2015-2016 with history of exposure to risk factors, with symptoms and strong clinical evidence of pre-neoplastic lesions.

Subjects who did not gave consent to be included in the study, patients with subsequent or concomitant primary malignant lesion in the regions other than oral cavity, patients with prior history of treatment for loco-regional or distant malignancy, patients who have clinically obvious growth of oral cavity and patients with the previous history of immune deficiency or autoimmune disorders were excluded from the study. Detail history and clinical examination was carried out in all the cases. Findings were recorded on IRB approved performa.

RESULTS: The total of 400 cases were included in the study, 305 were males and 95 were females. (Fig-1) The age range was 14 – 65 years with the mean age of 28years and the youngest being 14 years old. OSF was seen in majority of the cases (n=232) followed by leukoplakia seen in 108 cases. Erythroplakia was found in 45 cases and 15 patients had lichen planus (Fig-2). Sites of oral cavity involved were buccal mucosa in 30%, tongue in 50%, floor of mouth in 40%, lower alveolus in 15%, upper alveolus in 5%, hard palate in 3% (Fig-3).

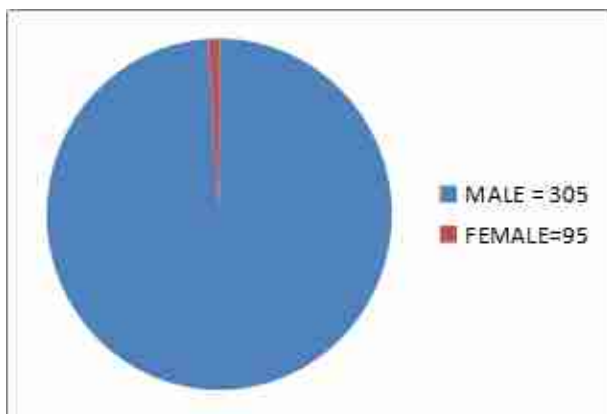


Figure 1 : Gender distribution.

DISCUSSION: It is a common observation that males are more susceptible to oral PML and oral cancer than females^{9,10}. One of the probable reasons of male preponderance is that females are less privileged to access for healthcare and diagnosis due to socio-cultural reasons being a male dominated society. However, the difference in the male to female ratio has become less

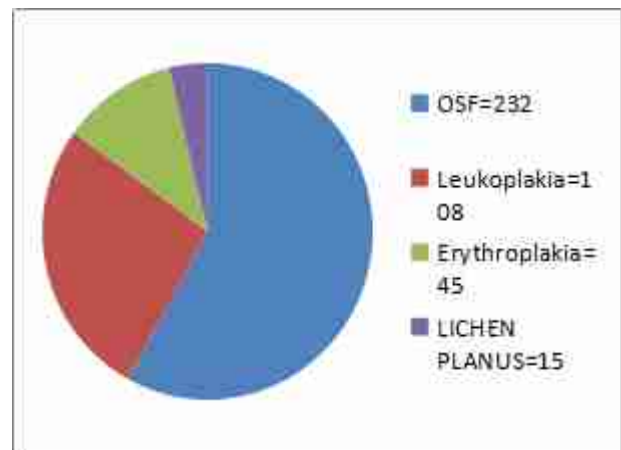


Figure 2 : Types of pre-malignant lesions encountered.

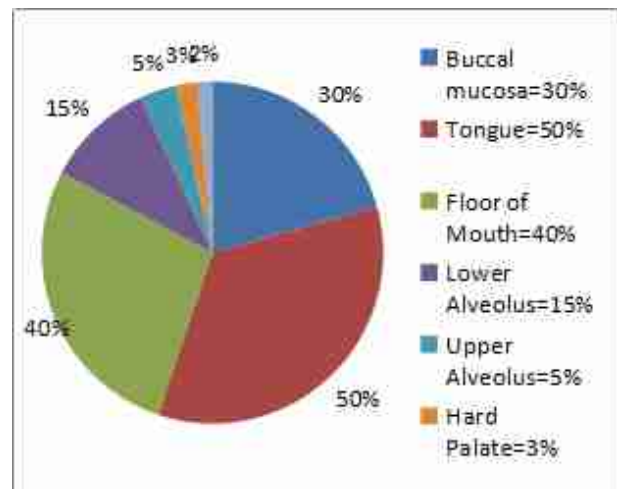


Figure 3 : Subsites of oral cavity involved in oral PML

women have indulged in risk factor habits exposing them to known oral carcinogens¹¹. Leukoplakia is the term best described as white patch or plaque of oral mucosa that cannot be removed by vigorous scraping and cannot be characterized clinically or pathologically as any other specific disease entity. Up till now it remains a clinical diagnosis of exclusion. Leukoplakia should be used only as a clinical term with no specific histopathological appearance and it may never be used as a microscopic diagnosis.

Approximately 80% of leukoplakia are benign with no histological evidence of dysplasia with none or less tendency to be converted in malignancy, while 10-20% of white lesions are either dysplastic or changed to carcinoma in situ or early invasive carcinoma that warrants histological evaluation. Mild, moderate or severe dysplasia rarely occurs and are indicative of carcinoma in situ. Risk factors for malignant

Leukoplakia occurs more commonly in younger and middle-aged men and arises most frequently on buccal, alveolar and lower lip mucosa. However, the lesions arising on mouth, lateral and lower lip are most likely to harbor dysplasia or progress to malignancy. There is no reliable method of predicting malignant transformation in PML. The rate of progression to malignancy has been reported to be between 3.6% and 17.5% over 10 years¹².

Erythroplakia and leukoplakia lesions are sometimes categorized together as speckled leukoplakia or speckled erythro-leukoplakia. It is not possible to separate two entities definitely in many clinical situations. Although less common than leukoplakia; erythroplakia is a worrying clinical condition that often harbors dysplasia. On histological analysis, 51% of erythroplakic lesions have been found to demonstrate invasive squamous cell carcinoma, with 40% demonstrating carcinoma in situ and 9% exhibiting mild to moderate dysplasia. It is considered to be carcinoma in situ and is more likely to transform into invasive OSCC⁴. Oral lichen planus is a relatively common inflammatory disorder of stratified epithelium with a prevalence of 0.5-2.2% in adult population¹³. Definite etiology of lichen planus is still not known. An autoimmune cause triggered by stress, trauma, viral infection or drugs has been proposed^{14,15}. Pre-malignant or malignant potential of lichen planus is in dispute. Some believe that occasional epithelial dysplasia or carcinoma found in patients with this relatively common lesion may be either coincidental or evidence that initial diagnosis of lichen planus was erroneous. It is difficult to differentiate lichen planus from epithelial dysplasia. It was found that 24% of oral lichen planus cases had 5 out of 12 World Health Organization (WHO) diagnostic criteria for epithelial dysplasia and only 6% had no histologic features suggestive of that disorder¹⁶. Oral sub mucous fibrosis (OSF) is a chronic progressive condition found frequently in people of Asian descent. It is predominantly reported from Pakistan, East India, Sri Lanka and South East Asian cultures. This pre-cancerous lesion is thought to be caused by areca nut, catechu, betel quid along with slake lime used for recreation in these geographic regions. OSF is considered to be due to consumption of areca nut products with resultant disruption of extracellular matrix affecting collagen synthesis. OSF may leads to changes in fibro-elasticity of oral mucosa causing burning sensation, spice intolerance and restricted mouth opening. The condition often manifest with diffuse involvement of oral cavity and pharynx that appears clinically as thickened whitish mucosa

muscle atrophy. Chronic inflammatory cells infiltration and epithelial changes ranging from atrophy to hyperkeratosis. Epithelial dysplasia has been found in 7-26% of OSF tissues. Long-term studies suggest a malignant transformation in approximately 7% of these lesions. The concept of a two-step process of malignancy development in the oral mucosa, i.e., the initial presence of a precursor subsequently developing into malignancy, is well-established. Salivary levels of cytokines, interleukin 6 and 8 were found raised in cases of oral PML but to a lesser extent than frank oral cancer¹⁸⁻²². This salivary detection would probably serve as a useful gadget for diagnosis and malignant transformation in future.

CONCLUSION: It is a common observation that males are more susceptible to oral PML than females. One of the probable reasons of male preponderance is that females are less privileged to access for healthcare and diagnosis due to socio-cultural reasons being a male dominated society.

Up to 80% of the cases of oral pre-malignant belong to low socio-economic status where, poor nutrition, compromised oro-dental hygiene and indulgence in the risky habits may contribute to its development. Nutritional deficiency may play a role in the development of oral pre-malignant lesions.

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Ameloblastoma of Ramus of Mandible Amalgamating as Parotid Tumor.

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ABSTRACT : Ameloblastoma is a benign locally invasive epithelial odontogenic tumour comprising 1% of all tumors and cysts arising in the jaws. It is commonly found in the third and fourth decade in the ramus of the mandible. In this report we present a 50 years old female with a large ameloblastoma in the region of right mandibular ramus along with cortical disruption. This lesion was diagnosed with the help of C.T scan and the patient was successfully managed by wide local excision with hemimandibulectomy. Literature is reviewed.

KEYWORDS: Ameloblastoma, Odontogenic tumor, Hemimandibulectomy.

INTRODUCTION : Ameloblastoma is the most common benign odontogenic tumor that constitutes about 1% of all cysts and tumors of the jaws. It is generally a painless, slow growing, locally aggressive tumor causing expansion of the cortical bone, and infiltration of the soft tissues. It has peak incidence in third and fourth decade of life but can be found in any age group with equal gender predilection (1:1). The relative frequency of mandible to maxilla is reported to be varying from 80-20% to 99-1%. In the mandible majority of ameloblastomas are found in the molar ramus region^{1,2}. Conventional radiograph is sufficient for small mandibular lesions but maxillary lesions and extensive lesions require CT and MRI to establish the extent of the lesion. We present a 50 years old female with a large ameloblastoma in the region of right mandibular ramus along with cortical disruption. This lesion was diagnosed with the help of C.T scan and the patient was successfully managed by wide local

presented with right cheek mass of 5 years duration. It was a slow growing painless mass which was not associated with any overlying skin changes. It did not increase in size with mastication and was associated with facial asymmetry. On physical examination it was found to be about 8x6 cm, irregular in shape with well circumscribed margins, firm to hard, non-compressible, non-reducible, non-tender, non-translucent mass over the right parotid area, extending from the zygomatic arch to the lower border of the mandible and extending 3cm lateral to the angle of mouth up to the post auricular area horizontally. The mass was fixed to mandible. Facial nerve function was intact with no facial muscle weakness observed Figure-1. The patient was otherwise healthy and was not taking any medications.

The patient was subjected to radiographic and routine hematological examination. The hematological findings were unremarkable. OPG



Figure 1 : Clinical appearance of the lesion.



Figure 2 : OPG Findings.



Figure 3 : C.T Scan Findings.

coronoid and condyloid process on the right associated with soft tissue swelling on the right side lateral wall of maxillary sinus was also appeared to be eroded and the zygomatic arch was not defined (Fig-2). C.T scan showed a large expansible destructive lesion appreciated in right mandibular ramus with cortical disruption. It was associated with a large soft tissue component involving the right parotid gland and extending into the infratemporal fossa proximally. It was involving the right masseter and right temporalis. The mass was showing multiple areas of necrosis and specks of air communicating with right second molar teeth. This mass was measuring 8.2x6.0x7.3 cm in APxTSxCC dimensions. Imaging appearances are suggestive of malignant neoplastic lesion (Fig-3). The differential diagnosis includes odontogenic keratocyst, odontogenic myxoma, central giant

cell granuloma and Brown's tumor of hyperparathyroidism. FNAC showed neoplastic lesion of low malignancy, probably of salivary gland origin. Differential diagnosis includes; polymorphous low grade adenocarcinoma, papillary adenocarcinoma (low grade), epithelial or myoepithelial carcinoma.

Considering the merit of the situation, modified Blair's incision was given, Branches of facial nerve were exposed due to stretching effect of the growth. Right ramus of mandible with part of the body, condylar and coronoid processes were involved by the tumor. Whole mandible ramus removed together with body and the whole specimen with growth was delivered between the retracted branches of Facial nerve. The specimen was sent for histopathology (Fig-4). Histopathology sections examined revealed a neoplastic lesion comprising of islands of columnar cells showing peripheral palisading



Figure 4 : Specimen showing growth along with hemimandible.

lymphovascular invasion or evidence of polymorphous low grade adenocarcinoma were observed. Final diagnosis of Ameloblastoma, follicular variant was made. Post operatively patient had slight neuropraxia of right marginal mandibular nerve. The patient was discharged on 5th post. operative day (Fig-5).

DISCUSSION: Ameloblastoma is a benign epithelial odontogenic tumour often aggressive and destructive with the capacity to erode bone and invade adjacent structures. Ameloblastoma of the lower jaw can progress to variable sizes (1-16 cm) and cause facial asymmetry, displacement of teeth, malocclusion and pathological fractures¹. In the present case also the patient's clinical examination revealed a large hard swelling in the ascending ramus of



Figure 5 : Post operative appearance of the patient.

development. Ameloblastomas occur with almost equal frequency in both genders slightly higher in women than men. The majority (70%) are located in the molar ramus region and 10-15% are found in association with an unerupted tooth³. In the present case also ameloblastoma was found in the molar ramus region and was not associated with any unerupted tooth. Ameloblastoma are classified as either extraosseous (peripheral) or intraosseous. Peripheral one presents as either sessile or pedunculated, slow growing mass that is confined to the gingiva or alveolar mucosa, without involving the underlying bone. Intraosseous ameloblastomas of the jaws are further classified as unicystic, desmoplastic, mixed cystic and solid types⁴. The mixed cystic and solid forms demonstrate more aggressive behaviour and are known for recurrence⁶. The histopathological variants are the follicular and plexiform types, followed by the acanthomatous and granular cell types. Uncommon variants are desmoplastic, basal cell, clear cell ameloblastoma, keratoameloblastoma and papiliferous ameloblastoma⁷. Among all these variants, plexiform pattern is less aggressive with low recurrence

Ameloblastoma usually presents as a painless, slow growing hard mass⁸. It was painless and hard swelling which took about 5 years to attain the presented size and there was no other clinical feature. In mandibular ameloblastoma half of the patients presented with swelling of the affected region (38.3%), paraesthesia of the innervated region of the mandibular nerve (13.3%) and altered occlusion of teeth in 10% of cases. In a conventional radiograph, ameloblastoma can present as either unilocular or multilocular; the bony septae results in a honey comb or soap bubble appearance, or tennis racket pattern. In some places, cortical plates are spared and expanded where as in other region they are destroyed. Root resorption is a common finding with buccal and lingual cortical plate expansion is common in ameloblastoma than in other tumors.

The challenge in managing ameloblastoma is in achieving complete excision and reconstruction of the defect when the tumor is large¹⁰. Ameloblastoma is treated by enucleation, curettage or surgical excision depending on size and type of the lesion. The rate of recurrence

includes resection with or without continuity defect.

For solid-multicystic ameloblastoma of the mandible, a resection of the jaw should be approximately 1.5-2¹¹ cm beyond the radiological limit, in order to ensure that all the microcysts and daughter cysts are removed. In our case also marginal clearance of 2cm was achieved and histopathology of specimen revealed it to be follicular variety.

There are different methods of mandibular reconstruction of large defect with microvascular surgery using donor site from fibula, iliac crest, scapula and radial forearm. In our case although tumor was treated with hemimandibulectomy as it was quite an extensive lesion involving body as well as the ramus of the mandible but the reconstruction of the mandible was not attempted as yet due to patient disagreement to undergo second stage surgery.

The chance of recurrence seems to be more dependent on the method of surgical treatment. In general, annual follow-up for at least 10 years is recommended. Few authors have recommended annual follow-up for 5 years and thereafter once in every 2 years till 25 years. Our patient is also under follow-up with no evidence of recurrence.

CONCLUSION: Ameloblastoma is the most commonly occurring odontogenic tumor in the mandibular body and ramus region in the middle age group, yet other differential diagnosis such as odontogenic keratocyst, odontogenic myxoma, central giant cell granuloma has to be ruled out with the help of advanced radiodiagnostic tools. The best treatment of Ameloblastoma is aggressive en bloc resection with simultaneous reconstruction.

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An Unusual Case of Intra-oral Epidermoid Cyst Mimicking Plunging Ranula

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ABSTRACT : Epidermoid cyst in the oral cavity is uncommon. Among the cysts in the floor of the mouth, they represent only 0.01% cases. We report the case of a 30 years old male presenting with 5x6cm swelling in the floor of the mouth extending into the submental and submandibular region. The midline swelling was painless, soft and dome-shaped. CT scan contrast revealed the site and extent of swelling. Complete surgical excision of the lesion was performed transcervically. Histopathology revealed cystic fibrocollagenous tissue covered by squamous epithelium containing some keratin flakes .

Key Words : Epidermoid cyst, Plunging ranula, Floor of mouth.

INTRODUCTION : Epidermoid cysts are benign lesions comprising solely of ectodermal lining¹. They may develop anywhere in the body, most commonly involving the ovaries, testis and sacrum. However, they may occur in the head and neck region in 7% of cases, with very few occurring intraorally (1.6%)². Only 0.01% of all cases are found in the floor of the mouth³. Though usually congenital, they often came into notice in the 2nd or 3rd decades of life². Epidermoid, Dermoid and teratoid are the three types of these cysts according to Meyers classification, with Dermoid cyst often used as a broader term encompassing all of these. Etiologically, they may develop due to trauma, dysodontogenesis and from thyroglossal cyst³. In the floor of the mouth, they may occur above or below the mylohyoid muscle presenting as an asymptomatic painless midline swelling with slow but progressive growth⁴. When the cysts enlarge in the oral cavity, they may push the tongue posteriorly and interfere with

CASE SUMMARY : A 30 years old male presented to the outpatient Department of Otorhinolaryngology at Civil Hospital Karachi with complain of painless swelling in the floor of the mouth extending into the neck in the submental and submandibular region. The swelling started 18 years back and was initially small in size. However, it gradually increases in size and over the course of last 3 years it grew very large as such it began to interfere with mastication, swallowing and speech. Fortunately, there was no interference with breathing. The patient told that the swelling burst and discharged spontaneously 7 years back exuding about 0.5 liter of greyish watery fluid containing white granules. Neither there was any significant medical history nor history of any previous trauma. On examination of oral cavity, there was a dome-shaped pinkish swelling observed in the floor of the mouth obliterating the entire gingivolingual sulcus and extending upto the anterior faucial pillars. It was a single 5x6cm



Figure-1 : Large sublingual epidermoid cyst extending into submental and submandibular region.



Figure-2 : CT scan coronal view showing non-enhancing mass in the floor of mouth extending into the neck.



Figure-3: Perioperative picture showing transcervical approach for the removal of the large epidermoid cyst.

was an inverted dome-shaped 13x11cm swelling in the submental and submandibular region extending upto the line below the angle of mandible superiorly and upto the hyoid bone inferiorly. It was soft, fluctuant, non-tender, non-reducible but compressible. Color and temperature was same as the surrounding skin. Transillumination was absent (Fig-1). CT scan of head and neck with contrast was performed. It showed a large mass 10x7x5cm containing fat and fluid with a fat fluid level. Extend from soft palate to floor of the mouth and submental region. Abutting the maxilla and mandible anteriorly while pushing the tongue posteriorly (Fig-2). All the laboratory reports were unremarkable Surgical removal of the lesion along with the sublingual gland was planned. General anesthesia was given with



Figure-4 : H&E stained section shows a cyst lined by stratified squamous epithelium without any associated skin adnexal structures with underlying fibromuscular tissue showing inflammatory cells. The luminal side of cyst shows keratin flakes.



Figure-5: Post-operative picture of the patient showing complete removal of the epidermoid cyst.

incision and was exposed by subplatysmal dissection (Fig-3). By careful dissection, the thin wall of the cyst was separated from surrounding tissues and was excised in-toto. Sublingual gland was also removed transcervically. The excised specimen was sent for histopathological evaluation which revealed cystic fibrocollagenous tissue focally covered by squamous epithelium. The cyst showed denuded lining and some keratin flakes. The tissue showed fibrosis, congested vessels and mild inflammation. Focal salivary gland parenchyma was also seen. The finding were consistent with Epidermoid cyst (Fig-4). Post-operative recovery was uneventful with remission of misery of the patient to his utmost satisfaction (Fig-5).

DISCUSSION : Epidermoid cysts are non-malignant inclusion cysts lined by ectoderm alone⁵. Etiologically, they may be dysodontogenetic originating from the entrapment of ectoderm during fusion of the first and second branchial arches in the 3rd and 4th weeks of embryonic life. They may also develop later in life, as an acquired lesion, secondary to some trauma resulting in embedment of epithelial cells into the deeper tissues. Another theory suggests thyroglossal anomaly as a cause^{2,6}. Epidermoid cyst usually presents in the 2nd and 3rd decades. It is rare in childhood. It has no sex predilection affecting both the genders equally^{6,7}. Epidermoid and dermoid cysts occurring in the head and neck constitute only about 7% of these cysts. They are uncommon intraorally (1.6%) with only 0.01% cases reported in the floor of the mouth⁸. Anatomically, cysts in the floor of the mouth can be categorized into three types; sublingual, submental and submandibular cyst; depending upon their relationship with the mylohyoid, geniohyoid and genioglossus muscles^{2,6}. Histologically, Meyer

have their lining derived from all the three germinal layers⁹. Clinically, epidermoid cysts are usually painless and asymptomatic. However, large ones may push the tongue backwards interfering with mastication, articulation, swallowing and sometimes, respiration. Moreover, they are well-encapsulated, slow growing cysts, soft to doughy in consistency, and usually present without lymph node involvement⁵⁻⁷.

The differential diagnoses of epidermoid cyst can broadly be classified as infective (cellulitis of the floor of the mouth, necrotizing lymphadenitis, and abscess formation-Ludwig's angina), neoplastic (salivary gland tumors), developmental (branchial cleft cyst, cystic hygroma, thyroglossal cyst), anatomic abnormalities (lymphangioma, mucocele, haemangioma, lipoma, neurofibroma) and mucous extravasation (ranula). The absence of inflammatory signs helped us excluding infectious lesions. While the benign appearance and lack of lymph nodes involvement reduced the likelihood of malignancy in such cases^{3-5,7-11}. Since, the lesion in our case had a prominent oral component along with submental and submandibular swelling, we ruled out developmental and anatomic abnormalities of neck. Hence, we were left with diagnostic possibility of epidermoid cyst and ranula. Plunging ranula topped our differential list because of the clinical appearance of the lesion and also because its more common. In few cases of sublingual swellings where it is difficult to establish diagnosis clinically, the imaging techniques are employed to reach conclusion. Though, US, CT and MRI, all are helpful in identifying the lesion and establishing its size, site and extent but none of these are conclusive. FNAC also has a low sensitivity and specificity for diagnosis of these types of lesions. Hence, histopathology is the only effective method in accurately diagnosing the disease⁴. Complete surgical excision of the cyst is the recommended treatment in these cases to validate the diagnosis

those below the mylohyoid are best approached extraorally via submental or submandibular incision^{4,9}. Malignant transformation of the lesion is rare. After adequate removal of the lesion, prognosis is good with very low incidence of relapses^{1,9}.

CONCLUSION: Although, intraoral cystic lesion in the floor of the mouth are rare, epidermoid inclusion cyst should be considered in differential diagnosis. Moreover, the treatment of epidermoid cysts of the floor of the mouth is surgical and can be intraoral or extraoral according to the localization and the size of the lesion.

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