Huge Pleomorphic Adenoma of Sub Mandibular Gland

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ABSTRACT: Pleomorphic adenoma is the most common salivary gland tumor and submandibular pleomorphic adenoma accounts for only 8% of them. The annual incidence is approximately 2-3.5 cases per 100,000 population. Pleomorphic adenoma occurs in individuals of all ages. However, it is most common in the third to sixth decades; the average age at presentation is between 43 and 46 years. Pleomorphic adenoma is seen more often in females than in males (2:1 ratio). Large masses appear heterogeneous on imaging secondary to hemorrhage, necrosis and cystic changes. Lobulated contour of the tumor favors the diagnosis of pleomorphic adenoma and they appear as multiple adjacent masses rather than single on CT scan. Treatment is essentially surgical with post operative radiotherapy is advisable in larger masses to prevent recurrence.

Key Words: Pleomorphic adenoma, huge submandibular gland, mixed salivary tumour.

INTRODUCTION: Pleomorphic adenoma is the most frequent benign tumour of the salivary glands. It is a slow growing tumour and is also called mixed tumour because histologically they are solid, tubuloglandular structure alternating with myxoid and chondroid zones. The name pleomorphic adenoma was suggested by Willis.

CASE HISTORY: A 50 yr old male presented with history of swelling over front of the neck from last 15 yrs gradually increasing to present size (15cm by 10 cm approximately). (Fig.1) On examination the large lobulated swelling was seen over front of neck, predominantly on right side and extending into the right submandibular region up to the angle of the mandible. On palpation it was firm to hard in consistency. The patient was referred to radiology department, CSS Hospital, Meerut for ultrasound examination of neck. On ultrasound examination there was a large heterogeneous predominantly echogenic mass seen overlying right side of neck extending up to angle of mandible. On palpation it was firm to hard in consistency. The patient was referred to radiology department, CSS Hospital, Meerut for ultrasound examination of neck. On ultrasound examination there was a large heterogeneous predominantly echogenic mass seen overlying right side of neck extending up to angle of mandible. Multiple echogenic nodules and anechoic areas were seen within it. Right submandibular gland was not seen separately from the mass. Both parotid glands and left submandibular gland were normal. No lymphadenopathy detected in the neck. Both sided carotid arteries and jugular veins were normal, no evidence of thrombosis seen within them. On ultrasound possibility of a large mass arising from the left submandibular gland was kept and CT scan neck was advised but patient refused citing financial problems. FNAC of the mass suggested the diagnosis of pleomorphic adenoma. The patient was operated at ENT department GMC Srinagar and a large encapsulated mass well separated from the surrounding tissue was removed. (Fig.2) Post operative histopathological examination of specimen confirmed the diagnosis of pleomorphic adenoma arising from the submandibular.

DISCUSSION: Pleomorphic adenoma (benign mixed tumor) is the most common salivary gland tumor and represents 70-80% of all benign tumors of the major salivary glands. Of all the pleomorphic adenomas over 85% occur in the parotid gland, 8% occur in the submandibular gland, and 6.5% occur in the minor salivary glands and 0.5% in the sublingual glands. The submandibular gland is involved in only 5% to 10% of the salivary gland tumors, and
pleomorphic adenoma (PA) is the most common tumor affecting it\textsuperscript{4}. Sarcomatous transformation is seen in only 2-5% of cases and is usually associated with tumors that have been present for 10-15 years\textsuperscript{5}. Tumors arising in the minor salivary gland account for 22% of all salivary gland neoplasms. Majority of them are malignant with only 18% being benign. Of the benign tumors pleomorphic adenoma is the commonest\textsuperscript{5}. The most common site of a pleomorphic adenoma of the minor salivary gland is the palate followed by lip, buccal mucosa, floor of mouth, tongue, tonsil, pharynx, retromolar area and nasal cavity\textsuperscript{5}. Pleomorphic adenomas are usually seen in middle aged women and present as a painless slowly growing mass. The lesions are usually solitary, ovoid, well demarcated masses\textsuperscript{2}. The larger tumors may have pedunculated outgrowths from the main lesion that grossly simulate multiple masses\textsuperscript{1}. On ultrasound they are seen as well defined rounded hypoechoic lesions with lobulated or bosselated contour and may have posterior acoustic enhancement. They may appear heterogeneous secondary to hemorrhage, calcification, and necrosis as seen our case. CT scan is an important diagnostic tool in these tumors because it helps in determining the extent of disease, local spread and also helps to some extent in determining the type of tumor\textsuperscript{1}. Presence of intact fat plane helps in distinguishing benign tumors from malignant\textsuperscript{1}. Most small tumors are smoothly marginated, spherical having higher attenuation value than surrounding normal gland however low attenuation area mimicking cyst may be seen. The larger masses most often have a heterogeneous appearance with sites of low attenuation representing areas of necrosis, old hemorrhage and cystic changes as seen our case\textsuperscript{1}. Localized areas of increased attenuation most often represent sites of recent hemorrhage. Dystrophic calcifications or ossification can occasionally be seen scattered throughout the tumor\textsuperscript{1}. Our case showed a single calcific focus. The larger tumors tend to develop a lobulated contour that, when present, is highly suggestive of the diagnosis as seen in our case\textsuperscript{1}. Such a lobulated mass on CT appears as multiple adjacent masses rather than a solitary lesion\textsuperscript{1}. CT shows well defined enhancing soft tissue mass. Contrast enhanced MR imaging delineates tumor margins better, necrotic and cystic areas can be differentiated from enhancing mass, also skull base invasion and vessel involvement can be better depicted. High grade tumor have intermediate to low signal intensity on all pulse sequences while low grade tumors are dark on T1W and bright on T2W images. The treatment of pleomorphic adenoma is essentially surgical. Though these benign tumors are apparently well encapsulated, resection of the tumor with an adequate margin of grossly normal surrounding tissue is necessary to prevent local recurrence as these tumors are known to have microscopic pseudopod like extension into the surrounding tissue due to “dehiscences” in the false capsule. Postoperative radiotherapy to the parapharyngeal space could possibly reduce the recurrence rate in such tumors\textsuperscript{6}. The dose of radiotherapy recommended in such cases is 50 Gy in 15-16 divided doses\textsuperscript{6}. It is unusual that despite of such a large mass of long duration which seemed to be originate from left submandibular gland on imaging, turned out to be benign pleomorphic adenoma of submandibular gland. The patient was from a far flung area with no access to medical facilities that is why presented at such an advanced stage.

REFERENCES: