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CASE REPORT

Ethmoidal Sinonasal Angioleiomyoma: A Rare Case With A Diagnostic Challenge

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ABSTRACT

Sinonasal Angioleiomyomas are rare benign mesenchymal tumours consisting of well differentiated smooth muscles proliferation with vascular component that represent less than 1 percent of all the vascular leiomyomas. The most effective way of detecting sinonasal angioleiomyoma preoperatively is by computed tomography however the postoperative histopathological remains the gold standard in diagnosis of vascular leiomyomas. In this case report we present a case of 45 years old female with positive history of nasal obstruction, epistaxis, hyposmia and facial pain. Physical examination revealed whitish mass in both nasal cavities with decrease nasal patency in both nostrils. After endoscopic resection, the diagnosis of sinonasal angioleiomyoma was established based on histopathology. Since ethmoidal sinuses are relatively uncommon area for the sinonasal angioleiomyomas to occur therefore we suggest that one should keep them in mind when making differential diagnosis of nasal obstruction with epistaxis. It is also worth noting that this is the first reported case of sinonasal angioleiomyoma arising from the ethmoidal sinuses from Pakistan.

Key Words: Nasal angioleiomyoma, paranasal sinuses, smooth muscle tumour, vascular leiomyoma

INTRODUCTION: Angioleiomyoma, also known as vascular leiomyoma, is a rare benign tumour that most commonly develops in the uterus (95%), skin (3%), and digestive system (1.5%). It is caused by smooth muscle cells in the arterial wall or by residual embryonic tissue. Nasal cavities host less than 1% of all vascular leiomyomas[1]. The inferior turbinate's, septum, and vestibule are where Sino nasal angioleiomyoma development is most frequently recorded. Epistaxis and nasal blockage are frequent presenting symptoms, and women are effected more frequently than males[2].

Due to the lack of smooth muscle in the nasal cavity, angioleiomyoma of the nasal cavity has an unknown etiology. Smooth muscle tumours in the nasal cavity have been theorised to originate from abnormal undifferentiated mesenchymal cells, smooth muscle components found in the walls of blood vessels and piloerector muscles, or both of the aforementioned theories concurrently[1]. According to certain studies, nose angioleiomyoma can be caused by Epstein-Barr virus infection and sexual hormones[3].

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The most effective way to understand the extent of the tumour's growth is computed tomography (CT), but it can be sometimes misleading causing misinterpretation of sinonasal angioleiomyoma as more of typical nasal lesions such polyps, fibroma, papilloma, hemangiomas, and angiofibroma because of their vague appearance and imaging findings. A conclusive diagnosis is provided by histopathological analysis[2].

In this paper we reported a case of nasal mass which was later found out to be Sino nasal angioleiomyoma arising from the ethmoid sinus and based on our literature search this is the first of ethmoidal Sino nasal angioleiomyoma to be reported from Pakistan.

CASE PRESENTATION: A 45 years old female known case of hypertension presented in our Otorhinolaryngology department with complaint of bilateral nasal obstruction for 1 year and recurrent nose bleeding for 6 months. The patient reported that the nasal obstruction was gradual in onset, slowly progressive and had worsened over time with no significant aggravating or relieving factor but on and off associated with mouth breathing and snoring. Our patient also revealed the history of epistaxis for 6 months on and off at least 2 to 3 time a week. The epistaxis happened on its own, but it could also be brought on by trauma to or manipulation of the nasal canal, such as when she blew her nose or sneezes hard.

The patient also complaint right sided facial pain, hyposmia and on and off frontal headache. However, no history of nasal itching, sneezing, post nasal dripping was present. On physical examination there was whitish mass visible in both nasal cavities which was sessile, firm in consistency, insensitive but bleed to touch with a slight left sided nasal deviation and decrease nasal patency in both nostrils. Colour vision, eye movement and corneal reflex all were intact. As a part of investigative proceedings CT Scan Nose and Paranasal Sinuses with contrast was done which showed heterogenous soft tissue density mass in bilateral ethmoids extending into bilateral sphenoid, laterally abutting the medial wall of both sided maxillary sinus and erosion of posterior nasal septum as shown in Figure 1.

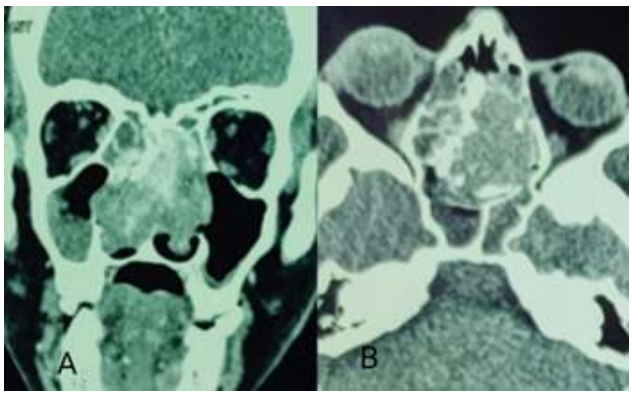

Figure 1: Computed Tomography of the Nose and Paranasal Sinuses in Axial (A) and Coronal (B) Sections, Showing Mixed Density Mass Arising From the Ethmoid Sinuses, Involving the Both Nasal Cavities.

Surgical resection of the mass was done through endoscopic approach. The resected specimen was composed of white globular mass which was quite shiny in appearance (Figure 2) On Histopathology report tissue specimen was composed of haphazard smooth muscles pattern with cytologic atypia, tumour necrosis and increased mitotic activity largely associated with ectatic blood vessels with narrow lumina. Reactivity pattern of immunohistochemical stains were positive for Anti smooth muscle antibodies and negative for S-100. The patient's post-operative recovery went without issue. The initial six months of follow-up revealed no new problems or recurrence. The nasal obstruction of the patient was significantly improved, and endoscopic examination revealed a clear nasal cavity.

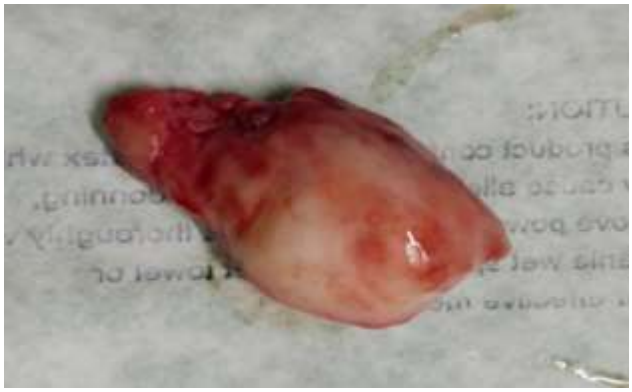

Figure 2: Post Operative Resected Specimen of Sino Nasal Angioleiomyoma

DISCUSSION: Rare benign mesenchymal tumours called angioleiomyomas are made up of proliferating, well-differentiated smooth muscles with a vascular component[4]. Leiomyomas in the nose and paranasal sinuses are relatively uncommon, and their appearance can resemble that of any other benign nasal lesion[5]. The most frequent symptoms in angioleiomyoma patients are pain and temperature sensitivity. When a tumour is exposed to external stimuli or is even slightly touched, pain, which is frequently paroxysmal, might be caused. The histological type of the tumour is correlated with the existence of pain[6].

Only 5 of 54 instances (9%) of sinonasal angioleiomyoma reported pain as a presenting symptom, according to a 2015 analysis of the relevant literature. It is quite uncommon to experience facial pain on its own. We are aware of only one other case where a sinonasal angioleiomyoma only manifested as unilateral facial pain[4].

Vascular leiomyoma is difficult to diagnose clinically. Rarely is it taken into account while making a differential diagnosis. Nasal angiofibroma, hemangiomas, inverted papilloma's, malignant lymphomas, fibromas, leiomyoblastomas, hemangiopericytomas, angiosarcomas, angiomyolipoma, and vascular leiomyosarcomas are some more common cancers of the nasal cavity[1]. Based on the predominant histological pattern, angioleiomyomas can be classified into three subtypes: solid, venous, and cavernous. While the venous and cavernous kinds are marginally more common in males, the most prevalent subtype, Solid, displays a substantial female predominance[6].

Hematoxylin and eosin (H&E) staining can be used to histologically identify the majority of vascular leiomyomas. The smooth muscle bundles that make up a solid or capillary angioleiomyoma are so closely interlocked that they give the vascular channels a slit-like appearance. A venous angioleiomyoma features a thick smooth muscular wall in contrast to a cavernous tumor's thin smooth muscular wall and dilated vascular channel. The smooth muscle cells of angioleiomyomas are mature and well-differentiated, and mitotic signals are typically absent or extremely rare. Rarely, the tumour may show signs of calcification, hyalinization, or myxoid transformation[6].

Vascular leiomyomas are benign tumour lesions that have the capacity to develop into cancer. Although malignant transformation of vascular leiomyoma in the nasal cavity has never been observed, malignant transformation of VL in other places, such as the hands, has only very rarely been described. 31 Angio leiomyosarcoma, the malignant analogue of VL, exists and histopathological has dysplastic characteristics[7]. Angioleiomyoma is best treated with surgical excision. After complete resection, the tumour rarely reoccurs[6].

In one of the studies conducted by Ah-Young Kim *et al* in 2015 on a 70-year-old woman, presented with intermittent spontaneous epistaxis, and left nasal obstruction. A reddish globular mass was discovered near the inferior turbinate after an endoscopic examination. A greatly increased tumour coming from the inferior turbinate and a hemangioma were suspected on the CT image. The histopathology analysis revealed that the mass was a VL, and the patient underwent total endoscopic resection of the tumour. Immunohistochemical labelling revealed that the tumour lacked progesterone and oestrogen receptors. The time following surgery went without incident. Over the course of the 12-month follow-up, there was no local recurrence[7].

In another study conducted by JNS Heyman *et al* in 2020 on a 33-year-old man with complain of right sided facial pain, clinical examination revealed mass arising from medial border of right inferior turbinate. CT scan was done which showed single 13-mm polyp with no radiological signs of aggression growing in the right middle meatus Under general anesthesia, a complete macroscopic excision of the lesion was accomplished using an endoscopic method[4].

In 2015, Lalee Varghese et al. reported a case of a 40-year-old man who complained of recurrent right sided epistaxis. A tumour in the right inferior meatus that was pushing the right inferior turbinate upward was identified during a clinical examination. On a CT scan, the right inferior turbinate was enlarged, the left maxillary sinus was opaque, and the infundibulum was occluded. The right maxillary sinus has a polypoidal mucosal thickness. The right nasal tumour was completely removed, whereas the left's endoscopic sinus surgery was limited. The entire mass was excised after a large arterial bleeder close to the tumor's base was cauterised. Even after a 6-month follow-up with no recurrence, he continued to be symptom-free[6].

One such case was reported by Y W Lau *et al*, a 43-year-old lady presented with the complain of recurrent epistaxis and right nasal obstruction for two months. A large tumor in the right nasal cavity was found during a clinical examination. With the use of endoscopic endonasal surgery, the entire tumor was removed. A Sino nasal angioleiomyoma was identified by histopathological analysis of the tumor. She had a successful postoperative recovery and no recurrence after a year of monitoring[8].

A 45-year-old man who arrived with progressive left nasal blockage and recurrent epistaxis for several weeks was the subject of a case study by Chun-Hsein Ho et al. An examination using a Sinoscopy revealed a soft reddish tissue tumour on the left nasal floor. The computed tomography of the sinuses revealed a soft tissue mass in the lower left nasal floor without any evident bone erosion. Endoscopic removal of the cancer, which started from the anterior left nasal floor, was effective. There were no difficulties after the treatment, it was claimed[9].

The preferred course of treatment for this benign tumor is surgery. The surgical strategy is determined by the tumor's size, location, extent, and the surgeon's level of experience. The surgery of the turbinate has given rise to a number of well-known surgical methods (e.g., resectioning, laser surgical procedures, and coagulation procedures)[10].

CONCLUSION: The extremely rare benign tumour known as a nasal angioleiomyoma develops from smooth muscle cells. Its clinical appearance resembles that of other nasal tumours, making preoperative diagnosis difficult. The location and size of the tumour can be determined with the help of imaging techniques, but a histological analysis is still required for a conclusive diagnosis. Accurate diagnosis and effective care depend on pathologists and clinicians having a

greater understanding of nasal angioleiomyoma. Such occurrences should be reported and documented in order to add to the expanding body of knowledge about this unusual nasal tumour, which will help with comprehension and patient outcomes.

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