INTRODUCTION

A variety of pathologies present as swelling in the maxillofacial region. These range from various slow-growing benign tumors and low-grade malignancies to fast-growing high-grade malignancies. Angiofibromas are one such group of tumors arising in this region, which present as swellings. Of these, nasal angiofibromas (NAs) are the most common and NAs occurring in the region other than the nose are called as extranasopharyngeal angiofibromas (ENA).

One such case presented to us as a swelling in the parotid region. Differential diagnosis and a brief review of literature are presented in this case report.

CASE REPORT

A 54-year-old male reported to our department with history of swelling over the right side of face since 5 years. The patient reported insidious onset of swelling with a gradual progress to the present size. He also reported mild, intermittent, and dull pain in the region of swelling.

Clinical examination confirmed a solitary, oval, well-defined swelling measuring approximately 7 × 5 cm in the preauricular region [Figure 1]. The surface over the swelling was smooth without any secondary changes. It extended supero-inferiorly from the level of outer canthus of the eye to inferior border of mandible and anteroposteriorly extended from 2 cm in front of anterior border of ramus to the mastoid notch [Figure 1]. On palpation, the swelling was firm, non-tender, free from skin, and mobile over the underlying structures. Mild pulsations could be appreciated from the swelling. There was no weakness of facial muscles or enlargement of swelling during mastication.

Ultrasonography revealed a large lobulated solid mass of 6 × 4 cm in the right parotid region with multiple vessels with low velocity flow and without any areas of calcification or cystic changes. Findings were suggestive of a low-grade vascular tumor. Computed tomography scan with contrast medium revealed a large well-circumscribed, solitary isodense lesion measuring 6.36 × 4.39 cm [Figure 2]. It was observed to be extending anteriorly over the superficial lobe of the parotid gland, with anterior displacement of parotid gland. A diagnosis of mixed parotid tumor was made.

FNAC revealed numerous vascular channels with occasional singly scattered and single cluster of plump spindle cells. Based on these findings, a lesion of vascular origin was suggested.

Classical superficial parotidectomy was performed under general anesthesia using a modified Appiani incision. Tumor mass was dissected anteriorly and excised along with the superficial lobe of parotid gland [Figure 3]. Facial nerve was located using the anterograde method and all the
branches of facial nerve were preserved. Post-operative recovery was uneventful. Upon six months of follow up, facial nerve function was intact and no sign of reoccurrence was observed.

Histopathological sections revealed connective tissue with numerous vascular spaces of variable sizes ranging from small capillary-like vessels to partly lined vessels. Around the endothelial cells, few vascular spaces showed a rim of circumscribed smooth muscle cells. The supporting connective tissue stroma appeared immature and fibromyxoid with sparse cellularity. Cells were plump or stellate-shaped with mild inflammatory cell response and composed predominantly of plasma cells and a few lymphoid follicles [Figure 4]. Immunohistochemistry revealed strong uptake of CD34 in the endothelial cells [Figure 5]. Findings were suggestive of a vascular lesion compatible with diagnosis of ENA.

**DISCUSSION**

Various soft tissue tumors, both benign and malignant, present as swelling in the parotid region. Clinical examination confirmed a smooth-surfaced, solitary, oval, firm, well-defined swelling measuring approximately 7 × 5 cm in the preauricular region. Based on the clinical findings of location, consistency and borders a differential diagnosis of pleomorphic adenoma, solitary fibrous tumor (SFT), low-grade fibrosarcoma, and lipoma was made.

As the most common tumors found in the region are parotid tumors, a provisional diagnosis of mixed parotid tumor was made. However, FNAC and Contrast-enhanced CT scan were in favor of a tumor with vascular component, shifting the differential diagnosis towards tumors with a vascular component presenting as slow-growing, circumscribed firm swellings.

Histopathological examination of the excised lesion revealed features suggestive of angiofibroma and had to be differentiated from lesions with similar features. Immunohistochemistry with CD34 was also done to confirm the presence of fibrovascular element. The differential diagnosis included low-grade vascular lesions, spindle cell neoplasms, comprising of a large range of benign and low-grade malignant soft tissue lesions, including cellular angiofibroma, SFT, Low-grade fibromyxoid sarcoma (LGFMS), low-grade myxofibrosarcoma, myxoid liposarcoma, giant cell angiofibroma (GCA), angiomyolipoma, and angiomyofibroblastoma. 

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Figure 1: Preoperative

Figure 2: CT-Scan transverse section

Figure 3: Excised tumor mass

Figure 4: Photomicrograph shows numerous endothelial lined blood capillaries with collagen fibres interspersed with fibroblasts and fibrocytes (H&E stain, ×40). (b) Photomicrograph shows endothelial lined blood vessels with connective tissue wall in the background of collagen fibres interspersed with fibroblasts and fibrocytes (H&E stain, ×400)

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Cellular angiofibroma, usually occurs in the pelviperineal region, which would be an unusual location for the tumors reported herein; however, cases have been reported to have occurred in the buccal mucosa,[14,15] raising a diagnostic possibility. Cellular angiofibroma shows more rounded, non-branching vessels, often of medium size with thicker walls. The tumors are generally more uniformly cellular, composed of lesional cells with short, stubby nuclei resembling those of spindle cell lipoma.[10,11,13-15] SFTs occur in a variety of locations in the head and neck region; myxoid examples may be particularly difficult to identify and should be considered in the differential diagnosis. Microscopically, besides the so-called “patternless” architecture, there is pronounced regional variation in cellularity, prominent thick collagen bundles and characteristic branching ectatic staghorn vessels, which are not accompanied by the abundant smaller-sized vessels present in angiofibroma of soft tissue. In addition, the tumor cells in SFT express strong and diffuse positivity for CD34 (IHC stain, ×100) and factor VIII.

LGFMS is a distinctive fibroblastic malignant neoplasm characterized by a peculiar tendency to give rise to very late metastases. Histologically, it shows alternating collagenous and myxoid areas with a usually swirling or whorled growth pattern, a frequently lobular appearance, and deceptively bland spindle cell morphology. The lesions tend to be more hypocellular, with a fibrous component that has uniform collagen deposition rather than the fibrillary or coarsely banded collagen fibers in angiofibroma of soft tissue. Although LGFMS may contain arcades of thin-walled vessels, vascularity is usually not prominent.[11,12] Myxofibrosarcoma is one of the most frequent sarcomas of elderly patients. Usually occurring in subcutaneous tissues of the extremities, it shows distinctive histologic features including a lobulated growth pattern with infiltrative margins and fusiform, round, or stellate tumor cells with frequently slightly eosinophilic cytoplasm and hyperchromatic atypical or pleomorphic nuclei. Obvious features of malignancy are usually present, characteristic elongated curvilinear vessels with perivascular hypercellularity, these features bear little or no resemblance to the rich vascular network of angiofibroma.[10-12]

Myxoid liposarcoma contains a prominent plexiform network of thin-walled capillaries (which has been referred to as a “chicken wire” or “crow’s feet” vascular pattern); hence, the possible diagnostic confusion. However, it also shows scattered univacuolar and multivacuolar lipoblasts throughout, as well as stromal mucin pools not seen in angiofibroma. GCA has been reported as a benign mesenchymal tumor with 2 cases originating in the buccal mucosa. Although benign, the lesion has potential for local reoccurrence, especially after incomplete resection. Histopathologically, it has similar presentation to that of SFT, with presence of multinucleated giant cells not seen in angiofibroma. Angiomyolipoma differs from angiofibromas because of the presence of prominent muscular arteries[9,13] while angiomyofibroblastoma differs because of the presence of fibrovascular component with a loose myxoid fibroelement.[9]

ENAs have interstitial stromal tissue predominance with less vascular elements such as that of long-standing NA.[2-7] Other distinctive histologic features are angiofibroma of soft tissue, consisting of a vaguely lobular, variably cellular proliferation of bland, uniform spindle cells set in an abundant, variably myxoid or collagenous extracellular matrix with numerous small, and thin-walled, branching blood vessels. The complexity of the vascular pattern, often also including larger vessels of varying size and shape, is the most noticeable feature. Immunohistochemical analyses may show that stromal cells have strong cytoplasmic reactivity for vimentin and are generally immunonegative for smooth muscle actin and desmin. Vascular component shows positivity for CD34 and factor VIII.[1] Histopathology coupled with immunohistochemistry usually confirms the diagnosis in favor of Angiofibroma. In our case, the tissue specimen showed features consistent with the diagnosis of ENA.

In 1980, De Vincentiis and Pinelli reviewed a series of 704 cases of angiofibroma and found that 13 cases manifested outside the nasopharynx, thus suggesting that extra nasopharyngeal localization of this tumor is a possible although rare occurrence.[10]

A review article by Windfuhr and Remmert[41] in 2004 summarized 65 patients with ENAs. The most common site reported for ENAs is the maxillary sinus followed by the ethmoid, nasal cavity, and nasal septum. ENAs are also reported to originate from the ethmoid sinus, nasal cavity, nasal septum, larynx, sphenoid sinus, cheek, conjunctiva, oropharynx, retromolar area, middle turbinate, inferior turbinate, and tonsil.[1,2,8] Other rare sites for ENAs occurrence have also been described-external nose, hard palate, external...
ear, lacrimal sac, carotid bifurcation, oesophagus, trachea, facial nerve, middle cranial fossa, and infratemporal fossa.\textsuperscript{[3,4]} The most common age of incidence was the second decade (46\%) in comparison to 80\% of patients with NAs. ENAs have a higher mean age, i.e., 22 years compared to NAs, which have a mean age of 18 years.\textsuperscript{[4,6]} The male to female ratio was 2.75:1.\textsuperscript{[3,4,7,9]} There is a higher incidence of ENAs in females. This is much higher than the rare incidence of NAs in females. The characteristic age and sex distribution of ENAs is not like NAs.\textsuperscript{[3,4]}

This lesion has been described as different from that of the classical nasopharyngeal variety in being more common in females, older individuals with an early presentation, and being capsulated with poor vascularity and infrequent recurrence.\textsuperscript{[4]} Unlike NAs, ENAs present non-specific and numerous symptoms. Clinical manifestation and the onset of symptoms depend on the tumor site.\textsuperscript{[3,5,7]}

Imaging modalities such as CT scan, MRI, CT angiography, and ultrasonography are required for optimal preoperative evaluation of NAs and ENAs.\textsuperscript{[2-6]} Imaging of NAs with a contrast agent leads to diffuse and usually homogenous involvement in CT and MRI scans with strong contrast enhancement.\textsuperscript{[3,2]} In contrast, ENAs enhances moderate amount of contrast or none due to its weak vascular involvement.\textsuperscript{[3,2]}

Surgical excision is the prime modality of treatment for ENAs; radiotherapy maybe applied for non-resectable lesions.\textsuperscript{[1-6]} No reoccurrence of ENAs have been reported in literature thus far.\textsuperscript{[3,4,7]} Occurrence of ENAs, although rare in the maxillofacial region, should be considered in differential diagnosis of swellings in this region.

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