SINONASAL MYOPERICYTOMA: A CASE REPORT

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ABSTRACT

AIM: Myopericytoma is a rare vascular neoplasm. It is extremely rare in sinonasal region. We report such a case which was treated by medial maxillectomy via lateral rhinotomy approach.

KEYWORDS
Sinonasal, Myopericytoma.

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INTRODUCTION Myopericytoma constitutes 1% of vascular tumours, 25% of which occurs in head & neck region. These tumours are rare in nose & paranasal sinuses. Histopathologically they overlap the features of myofibroma, angioleiomyoma & hemangiopericytoma. They show a radial and perivascular arrangement of ovoid neoplastic cells called myopericytes (Myocytes).

CASE REPORT: A 35-year-old lady presented with recurrent episodes of bleeding from left nasal cavity of 2 months duration. She also had progressive left-sided nasal obstruction. Anterior rhinoscopic examination revealed a reddish smooth surfaced mass in the left middle meatal area (Figure 1). Anterior end of the mass was about 2 cm behind the anterior nares. Probe could be passed all around the mass except the lateral part. On posterior rhinoscopic examination, both choanae and nasopharynx were free of lesion. Right nasal cavity and the nasal septum were normal. A contrast enhanced CT scan of nose and paranasal sinus was done, and it showed a heterogeneously enhancing, 4 x 2.8 x 3.7 cm sized lesion in the anterior part of left nasal cavity, arising from the left middle meatus destroying the middle turbinate. There was erosion of medial wall of maxillary sinus & widening of the ostium and extension of lesion to the maxillary sinus (Figure 2). Under local anaesthesia, a nasal endoscopic evaluation & biopsy was done. Histopathology report came as Myopericytoma. We proceeded with medial maxillectomy by lateral rhinotomy approach and the mass was completely excised. Final histopathology report confirmed the diagnosis of Myopericytoma. On immunohistochemistry, the tumour cells showed SMA positivity & CD34 negativity (Figure 3). She is now on followup.

Figures & Legends:

Fig. 1: Anterior Rhinoscopy Showing the Reddish Smooth Surfaced Mass in the Left Nasal Cavity

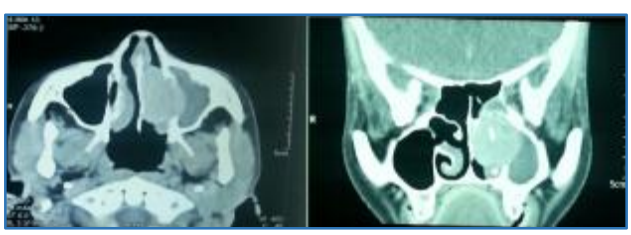

Fig. 2: Axial & Coronal CT Image Showing Heterogeneously Lesion Arising from the Left Middle Meatus Destroying the Middle Turbinate & Medial Wall of Maxillary Sinus with Extension to the Maxillary Sinus

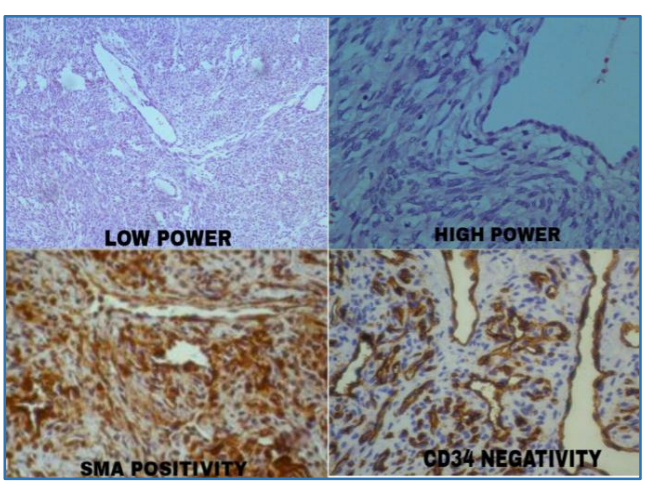

Fig. 3: Histopathology Showing Low Power, High Power and Immunohistochemistry Details

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DISCUSSION: In 1942, Stout[1] described a tumour that he termed haemangiopericytoma (HPC), which characteristically occurred on the extremities, and consisted of plump spindle shaped cells arranged around prominent, thin-walled, branched, blood vessels. Stout believed that HPC was a morphologically heterogeneous group of tumours, and recognised both myoid differentiation and an overlap with glomus tumours. Subsequently, Enzinger expanded the term HPC to describe any tumour characterised by the presence of a prominent, branching, thin-walled vascular pattern aligning it with vascular tumours.[2] More recently, it has been recognised that this vascular pattern is nonspecific and may be seen in a variety of other benign and malignant tumours, including benign fibrous histiocytoma, synovial sarcoma, mesenchymal chondrosarcoma, solitary fibrous tumour, leiomyosarcoma, endometrial stromal sarcoma, and infantile fibrosarcoma.[3] Thus, it is clearly inappropriate to classify tumours solely on the basis of their vascular pattern. The use of immunohistochemistry has assisted the appropriate classification of many tumours displaying this vascular pattern. However, a small subset remains, probably representing examples of myopericytoma (MPC).

Because the term "Haemangiopericytoma" has become nonspecific, there has been a general push to make Myopericytoma as a new term.[4] and this has now been endorsed by the World Health Organisation.[5] In the recent WHO classification of soft tissue tumours, myopericytoma and myofibroma (MF) are listed as separate entities and glomangiopericytoma appears as a subtype of MPC.[5] MPC is recognised on the basis of both immunohistochemical and ultrastructural studies, as a tumour derived from the perivascular myoid cell. Candidates for the progenitor cell of origin for the myopericyte include the myofibroblast or the pericyte. The term "Myofibroblast" is applied to a spindle shaped cell with elongated nucleus and pale eosinophilic cytoplasm that usually shows a desmin negative, actin positive immunohistochemical phenotype. The pericyte is viewed as a pluripotential resting stem cell, capable of differentiating along smooth muscle, pericyte, glomus cell, osseous, fibroblast, and adipocyte cell lines.[4,6,7] Indeed, differentiation of pericytes into myofibroblasts and smooth muscle cells has been documented[7]. This concept accounts for the spectrum of tumours and may explain the distinctive features of each variety.[4,5,6,7,8] Myopericytoma frequently affects soft tissues of extremities, only 25% occurs in head & neck region, rare in nose and paranasal sinuses. Sinonasal myopericytoma usually presents as epistaxis or nasal obstruction.

These are usually benign, and malignant varieties are very rare. Angiographically, these are well-vascularised lesions. Surgical excision is the treatment. Selective tumour embolisation will reduce per operative bleeding.

CONCLUSION: The term myopericytoma is used to describe a spectrum of tumours typified by a haemangiopericytoma like vascular architectural pattern with features of perivascular myoid (Myopericytic) differentiation. Myopericytoma typically affects soft tissues of extremities, rarely located in nose and paranasal sinuses. It should be included in the differential diagnosis of a slow growing bleeding nasal mass.

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