

Resection of Sinonasal Hemangiopericytoma through Transnasal Endoscopic Method

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Abstract:-

➤ **Background:**

Hemangiopericytoma is a soft tissue tumor which is derived from mesenchymal cells with differentiation of pericytes. It represents a limited proportion of tumors in head and neck and most commonly occurs in sinonasal cavities.

➤ **Case Report:**

A case of sinonasal hemangiopericytoma of 75years male patient admitted. He presented with symptoms of frequent episodes of epistaxis, foul smelling nasal discharge. A purple vascular lobulated mass filled in both nasal cavities. The tumor removed en-bloc by endoscopic operation.

➤ **Results:**

Post-operative histopathological and immunohistochemistry examinations confirmed sinonasal hemangiopericytoma. No recurrences seen both endoscopically and radiologically in 3year post-operative follow-up.

➤ **Conclusion:**

Hemangiopericytoma is vascular tumor of uncommon variation, its biological behavior prediction is difficult based on histopathological examination alone. Immunohistochemistry is helpful to diagnose the tumor.

Keywords:- Hemangiopericytoma, Epistaxis, Endoscopy, Paranasal Sinuses, Histopathology.

I. INTRODUCTION

Hemangiopericytomas are unusual vacular tumor which is derived from the mesenchymal cells with pericytes of zimmermann. These were first described by *Stout and Murray* in 1942⁽¹⁾. The contractile smooth muscle cells which are found on the outer surface of capillaries and venules are represented as pericytes.

Hemangiopericytoma can be either benign or malignant. These are 2 types: Infantile hemangiopericytoma and adult hemangiopericytoma. There is 15-30% of hemangiopericytoma occurs in H&N region approximately, most commonly found in parapharyngeal space. 5% of HPC of head and neck region arise from nasal cavity. It occurs in 5th-6th decades of life, there is no sexual predominance. Hemangiopericytomas are resistant to radiotherapy, they are friable and profusely bleed. Prior ligation of vascular bundle that nourishes the tumor followed by wide surgical resection is the treatment of choice. Endoscopic resection of sinonasal hemangiopericytoma is the best method and has many advantages.

II. CASE DESCRIPTION

An irregular tumor in bilateral nasal cavity was found in 75years old man in diagnostic nasal endoscopy (DNE). Nasal endoscopy showed a purple lobulated tumor with purulent discharge in bilateral nasal cavity, the tumor attachment present at lateral wall of nose (Figure1). There is proptosis of left orbit with diplopia.

Imaging modality of CT and MRI shows a soft tissue tumor involving both nasal cavities, all paranasal sinuses eroding nasal septum, lateral wall, lamina papyracea and compressing left medial rectus muscle (Figure 2). Due to risk of epistaxis preoperative biopsy was not performed. As the base of tumor was limited preoperative vascular embolization was not performed. Histopathological examination diagnosed as sinonasal hemangiopericytoma. It showed *Staghorn* pattern and blood vessels has a lining of flat endothelial cells. Tumor cells present as pale cytoplasm with round nuclei (Figure 4).

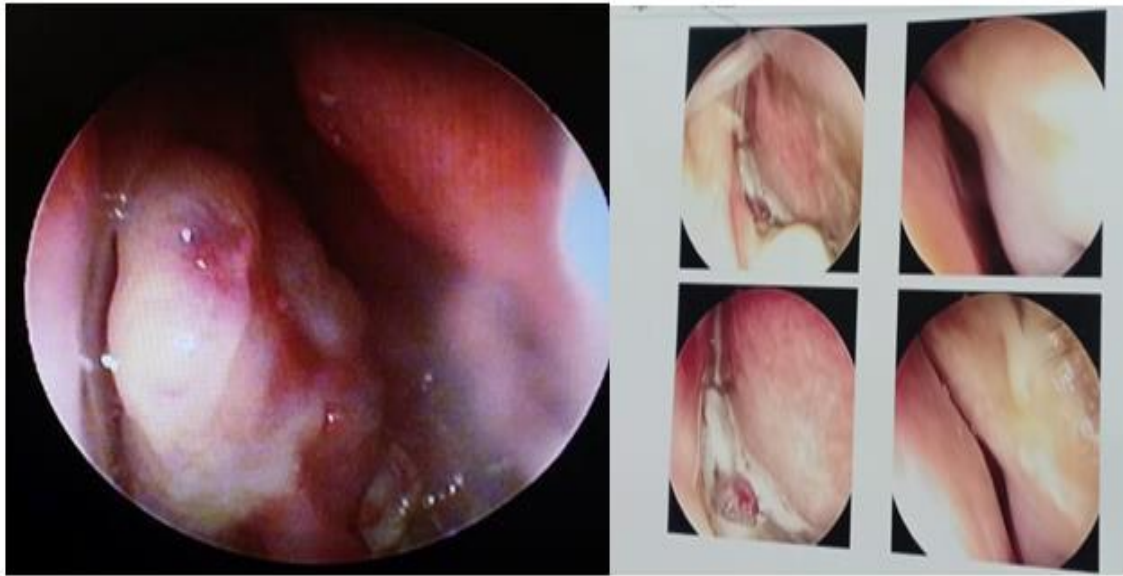
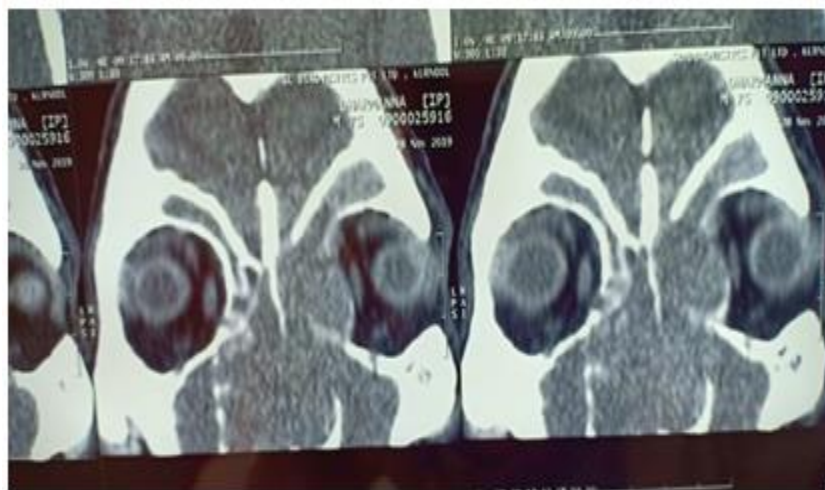
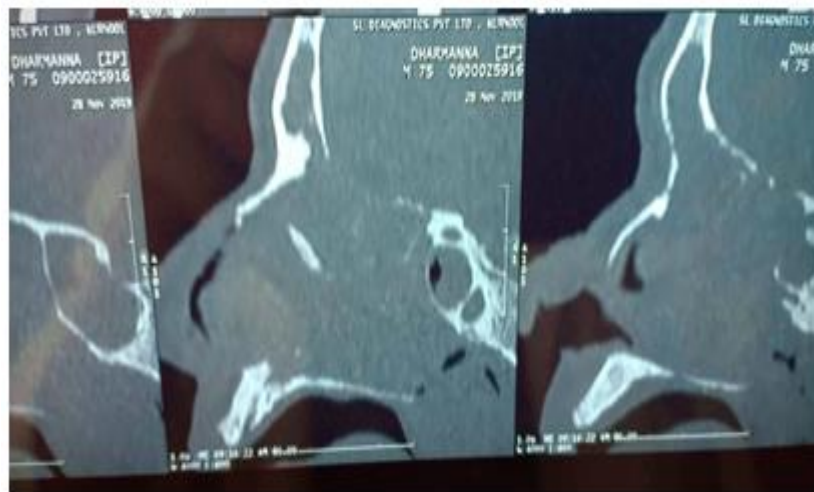


Fig 1 Preoperative Diagnostic Nasal Endoscopy

Diagnostic nasal endoscopy showing purple vascular tumor involving right nasal cavity mostly. There is highly vascular tumor surface. It is admixed with purulent discharge.



A.



B.

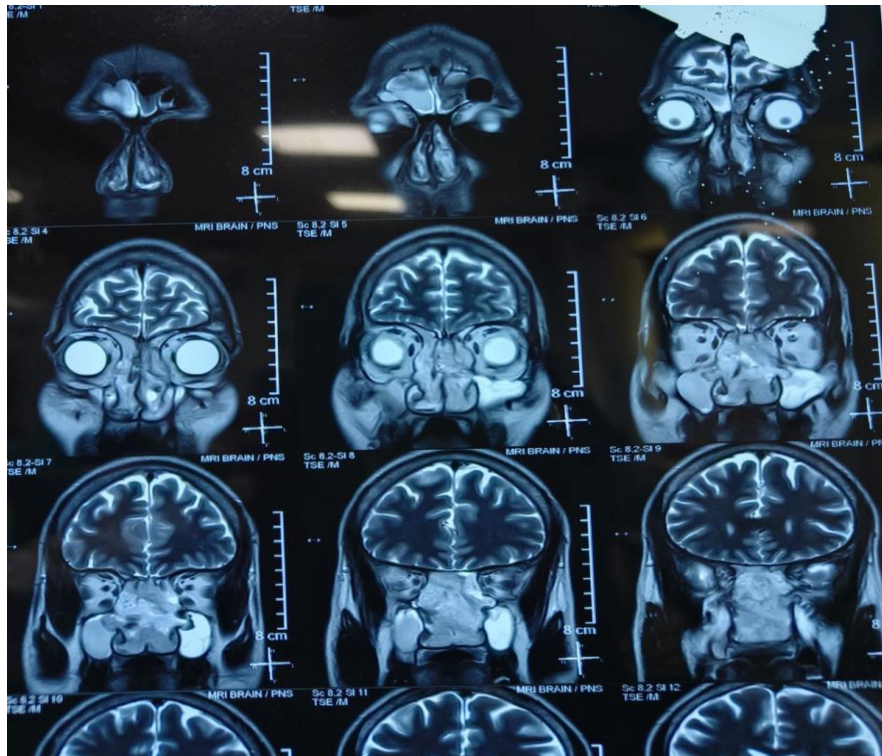


Fig 2 a) Computed tomography (CT) plain of the paranasal sinuses shows a mass involving both nasal cavities. Bony destruction of nasal septum and compression of left lamina payracea is evident. b) Saggital section of CT PNS showing involvement of Frontal, Sphenoidal and Ethmoidal sinuses with intact skull base. c) MRI Brain and PNS showing involvement of B/L nasal cavities, PNS with hyperintensity in left maxillary sinus and right frontal sinus.



A.

B.

Fig 3 Intraoperative Findings

A. Endoscopic Clearance of Tumor with Debrider ; B. Excised Tumor Specimen.

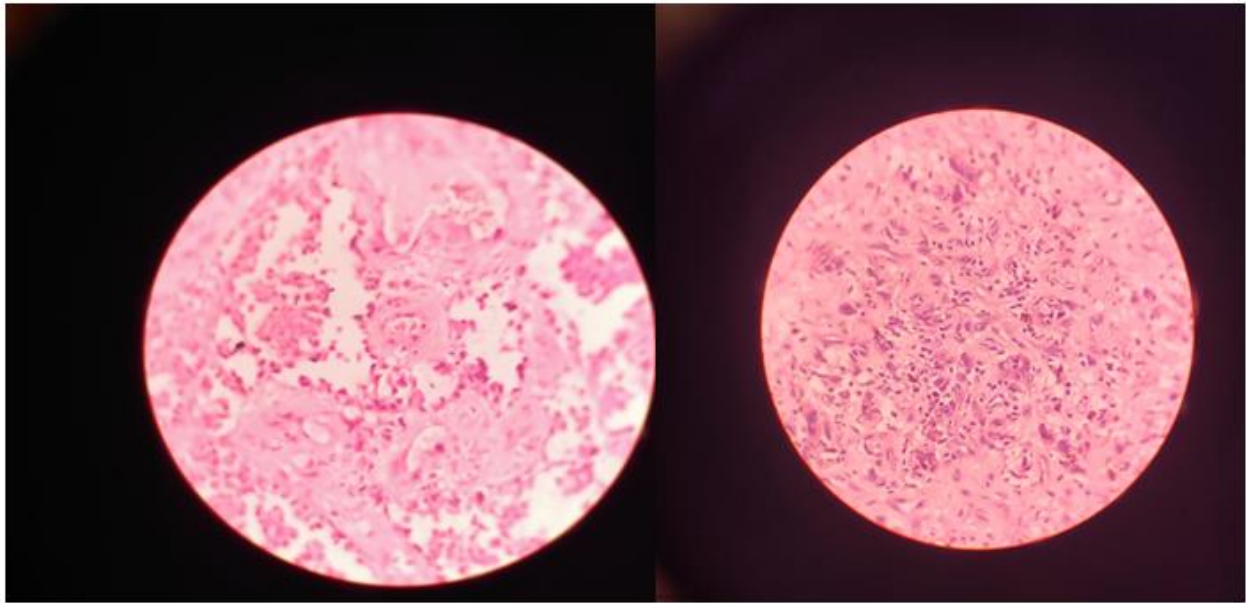


Fig 4 Histopathological Examination

Staghorn pattern and blood vessels are lined by flat endothelial cells. Tumor cells present a pale cytoplasm with round or ovoid nuclei.

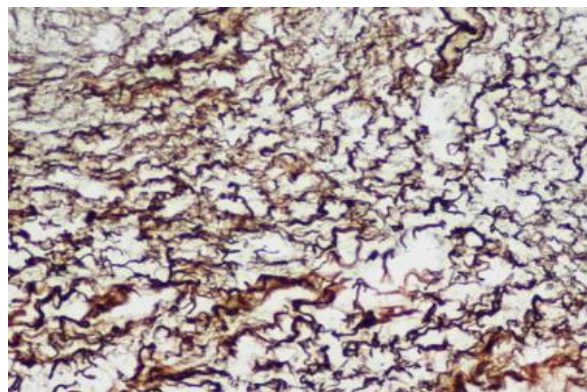


Fig 5 Immunohistochemistry Staining CD 99+Ve.

III. DISCUSSION

Hemangiopericytomas (HPC) are vascular soft tissue tumors of low grade which can arise from anywhere in the body. It can be both benign and malignant. It has a potential to turn into malignancy with late recurrence rates which are reported within the range of 8-53%⁽²⁻⁶⁾ and metastasis ranging from 12-60%^(5,6) of cases. It is very important to remove tumor completely because local recurrences tend to show metastasis.

Hemangiopericytomas are two types- Infantile HPC and Adult HPC. Various etiological factors are included like Hypertension, hormonal or metabolic imbalances.

The other more common pathologies in the sinonasal region (nasal polyposis, solitary fibrous tumor, fungal ball, inverted papilloma, malignancy) as a differential diagnosis should be undertaken preoperatively. Diagnosis is based on clinical and radiological examination. Based on diagnostic nasal endoscopy there is a suspect of vascular tumor. CECT and MRI of PNS revealed vascular tumors as strongly

contrast enhancement. Plain CT PNS showed lesion involving both nasal cavities and all paranasal sinuses, eroding nasal septum, lateral wall of nose, lamina papyracea, comprising left medial rectus muscle. On T2w MRI, tumor mass is iso to hypo intense whereas inflammatory fluids appear hyperintense⁽⁷⁾. The advantage of each sequence and a systematic approach to the evaluation of the images can be very helpful in understanding the characteristics of the type of the tumor. The use of gadolinium chelate contrast agents with fat saturation increases the sensitivity of contrast enhancement, thereby improving the detection of local disease extent and the presence of diseases beyond the paranasal sinuses.

Vimentin and smooth muscle actin are generally positive for the tumour cells in hemangiopericytomas (98% of specimens). A cytokeratin staining that is positive rules out hemangiopericytoma as a diagnosis.

Hemangiopericytoma and single fibrous tumour have been shown to be intimately related. It's crucial to distinguish between these tumours since benign solitary

fibrous tumours of the soft tissues represent the majority of them. The marker CD34 is incredibly helpful for this.

Gross picture of hemangiopericytoma is similar to many other soft tissue tumors. Differential diagnosis can be Synovial sarcoma, Solitary fibrous, Fibrous histiocytoma, Mesenchymal chondrosarcoma.

Poor visualization due to excessive bleeding has an increase risk of intraoperative complications. Strategies to improve surgical field include proper patient positioning, regulation of hemodynamic parameters, adequate topicalisation and the use of electrocautery. 15°-20° of reverse trendelenberg can increase venous drainage without compromising cerebral perfusion. A wide local excision of tumor is the preferred method for Sinonasal Hemangiopericytoma. If cribriform plate or skull base is breached, craniofacial approach is necessary. A tumor localized to nasal cavity and maxillary and ethmoidal sinuses can be removed with endoscopic approach. Complication like cerebrospinal fluid (CSF) leak is not seen in this case. Life long follow-up is necessary due to recurrences which can occur as a result of inadequate surgical excision⁽²⁻⁴⁾.

IV. CONCLUSION

We had one case of sino-nasal Hemangiopericytoma which is removed enbloc by endoscopic method. Complete excision of tumor is necessary to avoid latent malignant behavior of these tumors. External approach is gold standard method, it can be combined with endonasal endoscopic method. In 3 years follow-up no recurrences observed but follow-up for lifelong is necessary.

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