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Sinonasal - Type Hemangiopericytoma of Nasal Cavity: A Rare Neoplasm- Case Report with a Brief Review of Literature.

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Case Report

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ABSTRACT

Sinonasal type hemangiopericytoma, now called as glomangiopericytoma is a rare neoplasm arising from the pericytes surrounding the capillaries. It accounts for less than 0.5% of all sinonasal tumors. This tumor is different from conventional soft tissue hemangiopericytoma and hence considered as a distinct entity. It behaves in a benign fashion. Local excision with regular post-operative follow-up is recommended. We present a case of this rare tumor in a 55 yr male with its clinicopathologic and immunohistochemical features.

INTRODUCTION

Sinonasal type hemangiopericytoma was first reported and described as hemangiopericytoma in 1942 by Stout and Murray [1]. It is a rare neoplasm accounting for less than 0.5% of all sinonasal tumors. It arises from the pericytes surrounding capillaries and exhibits perivascular myoid phenotype [2]. The World Health Organization(WHO) classified this tumor as glomangiopericytoma in 2005. Sinonasal hemangiopericytoma is typically characterized by a benign course, low tendency of metastasis and a recurrence rate of approximately 25% [3,4]. Less than 250 cases have been reported and published in literature in past two decades making it a rare tumor entity [5].

We report this case for its rarity and describe the clinicopathologic and immunohistochemical features along with a brief review of literature.

Case Report

A 55 yr male, presented to the OPD with complaint of rt. sided nasal block since 10 months, insidious in onset, gradually progressive and aggravated on exposure to dust. This was associated with recurrent upper respiratory tract infections and watery nasal discharge. There is no significant past and personal history. Anterior rhinoscopy showed a polyp obstructing entire right sided nasal cavity. Hematological, microbiological and biochemical parameters were within normal limits. On radiology, CT scan showed polypoidal mucosal thickening in the right maxillary sinus ostium obstructing the right nasal cavity and causing deviation of the nasal septum to the left (Figure1, 2). Features were suggestive of Antrochoanal polyp.

Functional endoscopic sinus surgery was performed with excision of right nasal mass, right partial middle turbinectomy, right maxillary sinusotomy, balloon assisted sphenoidotomy and right frontal recess

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clearance. On histopathology multiple irregular grey white tissue bits together weighing 16 grams were received. Microscopy revealed a circumscribed unencapsulated cellular tumor composed of diffusely arranged spindle cells with uniform round nuclei, vesicular chromatin and indistinct nucleoli arranged in storiform pattern and short fascicles with interspersed blood vessels, areas of hyalinization and perivascular hyalinization (Figure 3, 4, 5, 6, 7).

Immunohistochemistry showed diffuse positivity for vimentin and smooth muscle actin in the tumor cells (Figure 8, 9). Based on histology and immunohistochemistry a diagnosis of sinonasal type hemangiopericytoma was given. Patient is on regular follow up, no evidence of recurrence is reported.

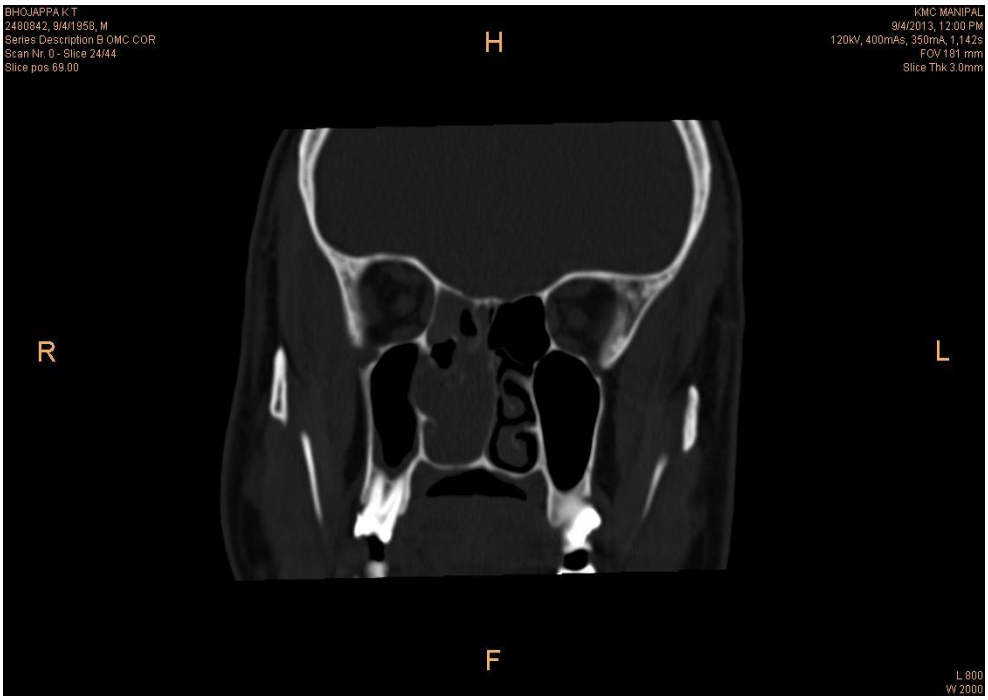

Figure 1: CT scan, Coronal section- Soft tissue density mass filling the right nasal cavity partly extending into the right maxillary sinus through inclined maxillary ostium and infundibulum with deviation of the nasal septum to the left

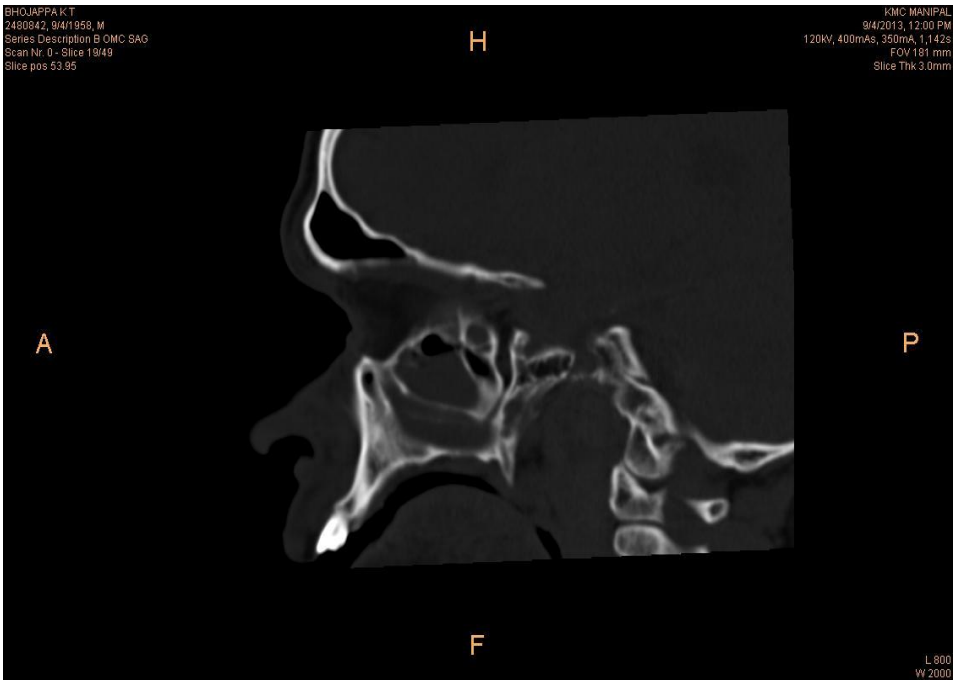

Figure 2: CT scan, Sagittal section- Intranasal extension of the lesion extending posteriorly through choana into the nasopharynx

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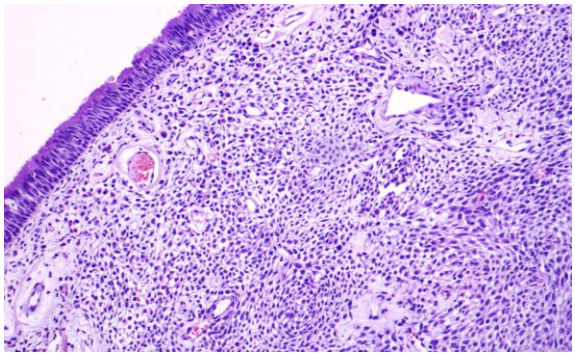

Figure 3: Unencapsulated cellular tumor beneath the respiratory epithelium H&E x100

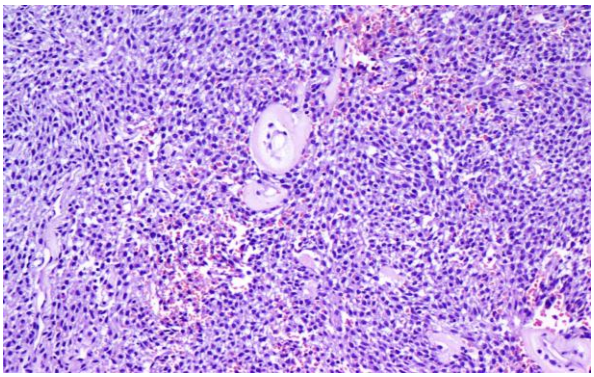

Figure 4: Cellular tumor composed of diffusely arranged spindle shaped cells H&Ex100

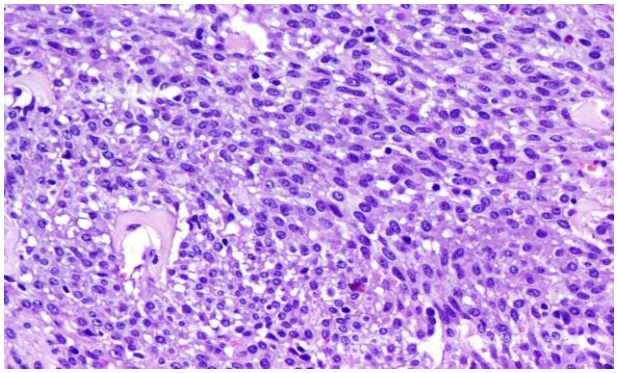

Figure 5: Blood vessels with perivascular hyalinization H&E x 200

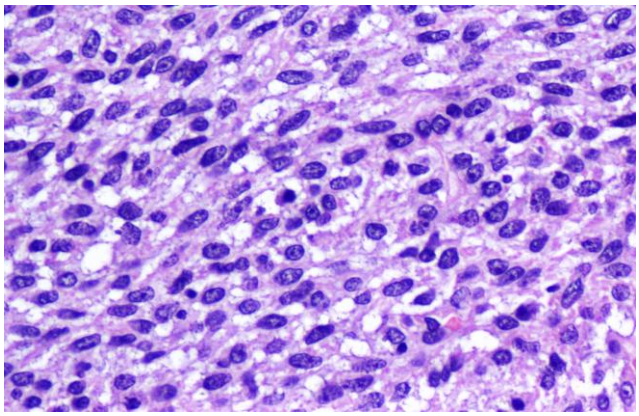

Figure 6: Spindle shaped cells with uniform nuclei , indistinct nucleoli H&E x400

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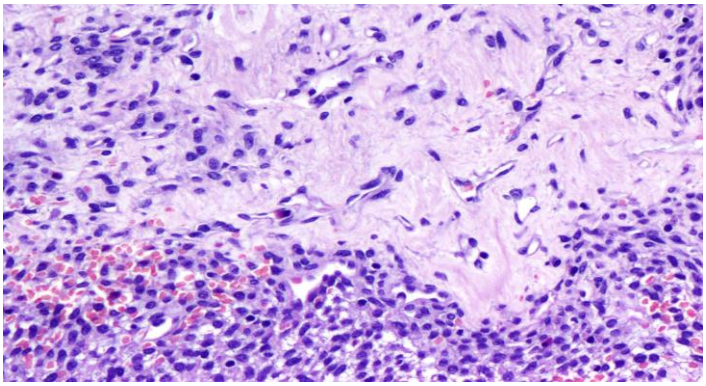

Figure 7: Areas of hyalinization H&E x200

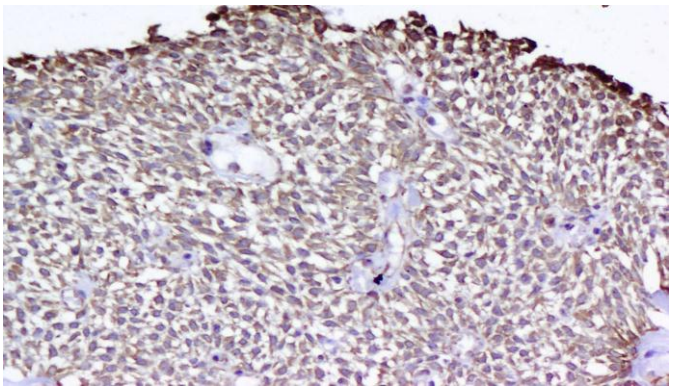

Figure 8: Diffuse positivity for vimentin x200

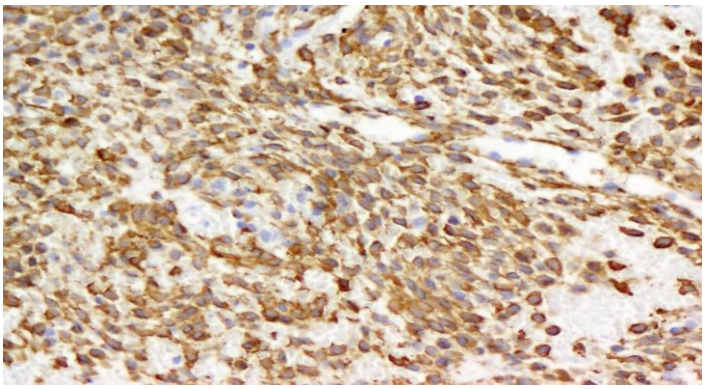

Figure 9: Tumor cells diffusely positive for SMA x200

DISCUSSION

The term hemangiopericytoma was first described in 1942 by Stout and Murray [1] as a soft tissue tumor with characteristic vascular proliferation including branching vessels and small vessel perivascular hyalinization. Around 5% of hemangiopericytomas are reported to occur in the nasal cavity and paranasal sinuses[5]. In 1976, Compagno and Hyams [3,4] first described the histological findings of sinonasal hemangiopericytoma and due to its typical morphologic features, term "hemangiopericytoma-like" was introduced. Sinonasal hemangiopericytoma is different from conventional soft tissue hemangiopericytoma in its location, biologic behavior and histologic features [2,6]. The World Health Organization in 2005 proposed that sinonasal hemangiopericytoma should be named glomangiopericytoma as the studies revealed their similarity and close relationship with glomus tumors [7,8]. This tumor is categorized as borderline and low malignant-potential soft tissue tumors of the nose and paranasal sinuses with a perivascular myoid phenotype. The proposed cell of origin is a modified perivascular glomus like myoid cell[9]. Sinonasal hemangiopericytomas accounts for less than 0.5% of all sinonasal neoplasms [2].

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This tumor affects any age, with a reported peak incidence in the seventh decade of life. In literature it has been reported as early in 18 years of age. There is a slight female predominance. However, this gender predilection is not well accepted. The etiology is not yet established completely, previously proposed predisposing risk factors like trauma, long-term steroid use, arterial hypertension, and hormone imbalance are now not accepted [2,5,9]. However, our patient did not have significant past history.

Mostly this tumor is located in the nasal cavity itself, although a few cases have been reported to occur in the paranasal sinuses [10]. This tumor usually presents with unilateral nasal obstruction, recurrent epistaxis and less frequently with breathing difficulty, visual disturbance and headache. Pain should be regarded as sign of local infiltration [2,5].

For suspected sinonasal tumours investigations include endoscopic evaluation and radio-imaging. The characteristic finding on CT imaging include a soft tissue mass with enhancement following the administration of intravenous contrast, and gadolinium contrast in the case of MRI scanning. MRI is preferred for a clear differentiation between tumor mass and inflammatory fluid in obstructed paranasal sinuses. Few authors also recommend angiography in larger tumours to facilitate pre-operative planning and to enable embolisation, therefore reducing the overall risk of intraoperative haemorrhage [5,11]. On endoscopic examination, sinonasal hemangiopericytomas are polypoid, beefy red, soft, fleshy to friable, edematous to hemorrhagic in appearance and bleeds on touch. They range in size from 1 to 8 cm, with a mean of 3.1 cm. Regional lymph node involvement is rare [2,9]. Clinically they may be mistaken for an inflammatory polyp [10].

On histology, these tumors are usually submucosal with an overlying intact respiratory epithelium. Tumor cells form a closely packed syncytium of uniform, monotonous, spindle-shaped cells with indistinct cell borders, vesicular to hyperchromatic, round to oval spindle-shaped nuclei arranged in short interlacing fascicles, storiform, whorled, and palisaded pattern. Tumor shows variable sized vascular channels exhibiting characterstic “staghorn” or “antler-like” configuration and prominent peritheliomatous hyalinization [9].

Aggressive clinical course and poor prognosis can be expected in tumors with size of >6.5 cm, histological findings of necrosis, nuclear atypia, a high number of mitosis and Ki- index of >10% [9,12]. Metastasis is reported to be approximately 5% which is very less compared to the conventional hemangiopericytomas. Lymphogenous and hematogenous metastases and involvement of lungs, liver and bone are been described in the literature. Less than 5% deaths due to sinonasal hemangiopericytoma are been reported [5,13].

Immunohistochemistry helps to differentiate sinonasal hemangiopericytoma from its histological mimickers like glomus tumors, angiofibromas, solitary fibrous tumors, lobular capillary hemangiomas, hemangioendotheliomas, myoepitheliomas, leiomyomas, olfactory neuroblastomas and rarely leiomyosarcomas and fibrosarcomas. Sinonasal hemangiopericytomas are true pericytic tumors exhibiting a myoid phenotype. On immunohistochemistry, they express vimentin, smooth muscle actin, muscle-specific actin, and factor XIIIa. Staining for desmin, S100 protein, and CD34 is variable. Additional negative stains include Bcl-2, CD99, CD117, and cytokeratins [9,10,14]. Vessels of the tumor exhibits partial positive staining for D2-40, podoplanin antibody and differentiates it from other solitary fibrous tumors [5]. Our case showed diffuse positivity for vimentin and smooth muscle actin in the tumor cells.

Success of the treatment depends on complete tumor resection, traditionally done via an open approach and presently endoscopic surgical techniques are preferred which minimizes the morbidity and facilitates subsequent surveillance of the operative site [11] for evidence of recurrence. 5-year survival is reported to be greater than 90% after complete surgical resection [9]. Chemotherapy and radiotherapy should be combined for preventing recurrences and are indicated as palliative treatment especially for inoperable tumors and cases of metastases. However, recurrences may occur even after a prolonged disease-free interval and has been reported 26 years after the tumor resection. Overall incidence of recurrence varies from 9.5% to 50%, depending on the length of the follow-up. Re-resection is usually performed after recurrence. Since the prognosis is not predictable clinically or by the histological findings, thus a regular, life-long follow up of such patients is suggested [5]. The treatment and follow up for our patient was uneventful.

CONCLUSION

Sinonasal hemangiopericytoma, now called as glomangiopericytoma is a rare, indolent, unique sinonasal tumor of elderly with a characterstic perivascular myoid phenotype. It is different from

conventional soft tissue hemangiopericytoma in its location, biologic behavior and morphologic characterstics. Histology and immunohistochemistry helps in establishing the diagnosis and differentiating it from its histological mimickers. These tumors usually have a benign course and have very less chances of metastasis. Complete local excision and regular, lifelong follow up is suggested.

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