Case Report

Sinonasal Glomangiopericytoma - A Rare Sinonasal Neoplasm

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Glomangiopericytoma of the sinonasal type is a rare sinonasal soft tissue tumor of low malignant potential. Its nosologic nature was controversial after it was first reported by Stout and Murray in 1942, who termed it as sinonasal haemangiopericytoma. It behaves very indolently as compared to its soft tissue counterparts, and now its cell of origin is known to be a perivascular modified glomus-like cell with myoid features. Clinically, it has many mimics ranging from inflammatory nasal polyps to epithelial neoplasms and vascular tumors. Here, we report a case in a 52-year-old male patient who presented with epistaxis and sinonasal polypoid mass. CT/CECT showed a polypoid lesion involving the right nasal cavity between the right middle turbinate and bony nasal septum with the possibility of a small mucosal nasal polyp. Total resection was performed by endoscopic nasal route. On histopathology and immunohistochemical examination, it was diagnosed as sinonasal glomangiopericytoma (GPC). We discuss the histology features and differentials to help in the diagnosis of this rare nasal neoplasm that presents with common symptoms of epistaxis diagnosis.

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Introduction

Sinonasal type glomangiopericytoma is rare, <0.5% of primary sinonasal neoplasms, seen over a wide age range but is more common in the sixth to seventh decade and is slightly more often seen in females (female to male ratio of 1.2: 1).[1] It has unique histology and behavior separating it from its soft tissue counterparts and is biologically close to the glomus tumor.[2] Clinically, it presents as a sinonasal polypoid mass and has an indolent behavior. Cell of origin is a modified perivascular glomus-like cell with myogenic differentiation, and thus, the preferred term is glomangiopericytoma.[1, 3] Patient presents with nasal obstruction, epistaxis, sinusitis symptoms, or even headache.[3] Clinical and radiology imaging (CT and MRI of paranasal sinuses) show polypoid mass, which on histopathology shows a submucosal soft tissue neoplasm with perivascular myoid cells that are positive for β catenin (nuclear staining) and negative for vascular markers. Though it has indolent behavior, but can present with a recurrence rates ranging from 7-40%.[1, 4, 5] The mainstay of treatment is complete resection with free margins.[6] To avoid bleeding during polypectomy surgery (as it is a tumor rich in vessels), endoscopic surgical resection, post embolization, or ligation of feeding arteries is generally done.[3,7]

This case report is in a 52-year-old male patient with a history of hypertension and complaints of right-side epistaxis, who underwent endoscopic nasal excision of the polypoid mass in the right nasal cavity. Its histology showed an unencapsulated submucosal spindle cell tumour with ectatic vessels, and immunohistochemistry was suggested for biphenotypic sinonasal sarcoma and sinonasal glomangiopericytoma. On immunohistochemical evaluation, the tumor cells were positive for β catenin (nuclear staining) and SMA and it correlated with the diagnosis of glomangiopericytoma (GPC), sinonasal type.

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Case report

A 52-year-old male with a history of hypertension presented to the ENT specialist with complaints of right-side epistaxis. His CT scan of paranasal sinuses showed evidence of well-defined polypoid lesion involving the right nasal cavity between the right middle turbinate and bony nasal septum with the possibility of a small mucosal nasal polyp. Deviation of the bony nasal septum was seen towards the right side with a septal spur causing compression of the right inferior turbinate. Deviation of cartilaginous portion of nasal septum was seen towards left side. Minimum mucosal thickening was noted in bilateral maxillary sinuses (more on right side), suggestive of changes of sinusitis. His CECT of paranasal sinuses was also reported as polypoidal, mildly enhancing soft tissue in the superior aspect of the right nasal cavity, placed between the nasal septum and the right middle nasal turbinate. It measured 8x18x19mm (transverse x anteroposterior x craniocaudal dimension). Histopathological correlation was suggested.

The patient underwent complete endoscopic nasal resection of the mass. In histopathology department of our tertiary care institute, we received specimen labelled as excised right nasal bleeding mass -haemangiomatous polyp. On gross examination, there were multiple greyish white soft to firm tissue pieces, aggregate measuring 1.5x1.0x0.3cm. Entire tissue was processed.

Light microscopy examination showed polypoid tissue lined by respiratory mucosal epithelium. Underlying submucosal soft tissue showed proliferation of bland spindled to ovoid stromal cells in fascicles and sheets with focal nuclear overlapping. Mitosis was very scant and occasional, and there was no necrosis. Numerous hemangiopericytomatous blood vessels were seen with sprinkling of plasma cells (figure 1 a and b)

The case was reported as a right nasal polypoid mass with spindle cell neoplasm, and differential diagnosis offered were biphenotypic sinonasal sarcoma and sinonasal GPC. Immunohistochemical markers such as SMA, β catenin, CD34, CD99, TLE 1, Cyclin D1, CK, desmin, SOX 10, HMB45 etc were suggested for categorisation and confirmation.

Immunohistochemical analysis done outside showed that tumor cells were positive for β catenin (nuclear staining), TLE1, CD99, SMA and negative for desmin, CD31, ERG, STAT 6, MyoD1, NKX2.2, S100, HMB45, CD34, and pan keratin. H3K27Me3 was retained. Ki67 proliferation index was 8%. The histologic and immuno profile now supported the diagnosis of sinonasal GPC.

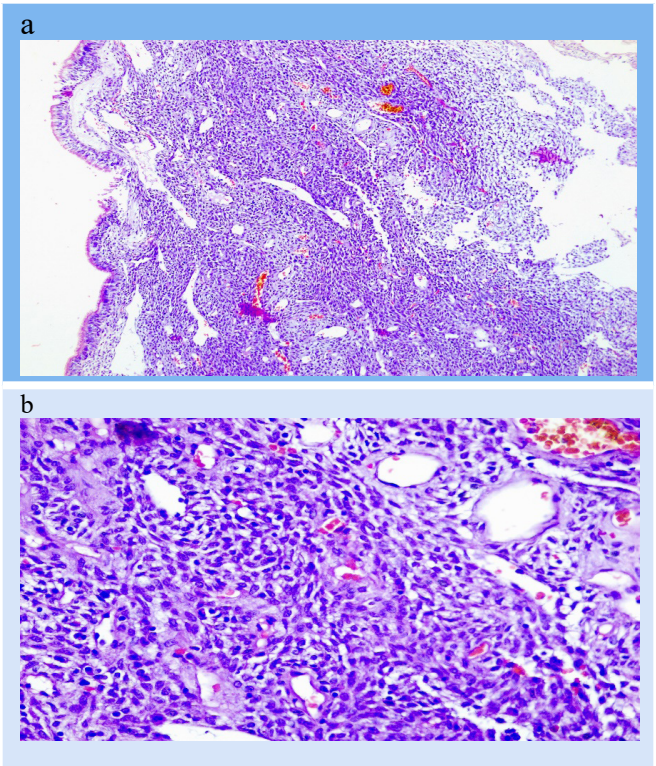

Figure 1 (a): Photomicrograph showing subepithelial unencapsulated tumour with overlying respiratory epithelium (H&E,100X), (b): Photomicrograph showing bland spindle and oval tumor cells arranged around ectatic blood vessels (H&E, 400X)

On follow-up, the patient did not present with recurrence or any symptoms.

Discussion

Sinonasal GPC was earlier also termed sinonasal hemangiopericytoma, glomus tumor, and intranasal myopericytoma, but due to its unique histology and behavior separating it from its soft tissue counterparts, the 4th edition of WHO classification of head and neck tumors (tumors of nasal cavity) has classified this tumor as a distinct entity.[8] It has been assigned ICD coding of ICD-10: D38.5- neoplasm of uncertain behavior of other respiratory organs.

The tumor occurs in a wide age range but is more commonly seen in older age (6th to 7th decade) and slightly more in females than males. Though clinical history is generally of epistaxis with nasal obstruction, these clinical clues are not of much help in distinguishing it from other more common causes of polypoid sinonasal mass. Glomangiopericytoma was first reported in 1942 as sinonasal hemangiopericytoma by Stout and Murray because of its staghorn vasculature.[9] They demonstrated by tissue culture method that the tumor cells originated

from Zimmermann's pericytes. In 1976, it was termed a hemangiopericytoma-like- intranasal tumour by Compagno and Hyams because of its low malignant potential.[10] Its cell of origin shows hybrid differentiation between glomus (myoid) and hemangiopericytoma (pericytic), and hence WHO termed this tumor as glomangiopericytoma.[3, 7, 11, 12]

The underlying cause is not clear, though it has been seen following trauma, in patients with hypertension those on corticosteroid therapy, or during pregnancy. Literature review shows cases associated with other conditions like hypoglycemia, serum hypophosphatemia, osteomalacia, and elevated alkaline phosphatase.[13, 14, 15]

Bilaterality is rare, and the tumor is usually seen in the nasal cavity and rarely in the maxillary, ethmoid, or sphenoid sinuses.

It can be equally challenging to diagnose this tumour on imaging. It is generally localized to the nasal cavity with rare extensión to paranasal sinuses or to the skull base, and if it were not for its submucosal location, it would be difficult to differentiate it from common causes of sinonasal polyps ranging from inflammatory polyps to inverted papilloma and squamous cell carcinoma.[16] Imaging shows a well-defined, round/polypoid or lobulated soft tissue mass with uniform enhancement on contrast administration. MRI provides superior soft tissue contrast and helps to define the tumor's relationship to adjacent structures. T1 images show it to be isointense to muscle, and on T2, it shows heterogenous high signal intensity.[17] In fact, the aggressive nature of lesions can be preoperatively assessed with the help of CT and MRI in order to see the presence and extent of invasion, especially bone erosion.[18]

Its underlying molecular etiology has been attributed to the mutational activation of β catenin with *CTNNB1* mutations and associated over-expression of cyclin D1.[19, 20]

Diagnosis of GPC, sinonasal type, requires histopathology and immunohistochemical workup for confirmation. Hematoxylin and eosin-stained sections on light microscopy show an unencapsulated bland spindle cell lesion in the subepithelial zone, separated from the overlying respiratory epithelium by the Grenz zone. The tumor has characteristic ectatic staghorn-type vasculature with perivascular hyalinization and spindle cells arranged in a fascicular or solid or focally whorled pattern. These lesional cells are bland with oval to spindled nuclei and eosinophilic cytoplasm. Mitosis is rare. Inflammatory cells, including mast cells and eosinophils, can be seen. Malignant GPC shows features of bone invasion with mitotic activity, necrosis, and pleomorphism.[1, 3, 7, 18, 21, 22, 23, 24]

Its differential diagnosis includes vascular and spindle cell tumors like lobular capillary haemangioma, angiofibroma, solitary fibrous tumors, leiomyoma, spindle cell sarcomas (biphenotypic sinonasal sarcoma, MPNST, synovial sarcoma) and melanoma. Immunohistochemistry helps in confirming as these tumor cells show characteristic nuclear positivity for β catenin, and cells are also positive for other markers like smooth muscle actin (SMA), CD99, and cyclin D1. They are negative for endothelial cells or vascular markers like CD34, CD31, ERG, and factor VIII. Lobular capillary haemangioma has a lobulated granulation tissue-like appearance and is positive for CD31 and CD34 and negative for nuclear β catenin staining. Angiofbromas have stellate cells with stromal collagen and are androgen receptor (AR) positive. Another locally aggressive, malignant, low-grade sinonasal sarcoma with focal nuclear β catenin positivity is biphenotypic sinonasal sarcoma. This is a cellular, infiltrative spindled cell proliferation with areas of herringbone pattern and is so termed as it has both neural (S 100 positive) and myogenic (SMA/MSA positive) differentiation with PAX3 positive staining on immunohistochemistry due to recurrent *PAX3* translocations, most commonly partnered with *MAML3.* In contrast, sinonasal GPC does not show a herringbone pattern, and immunohistochemistry shows diffuse SMA and nuclear β catenin-positive staining with the absence of S 100. Ectatic staghorn vasculature is a common feature in solitary fibrous tumors, but this tumor is STAT6 and CD34 positive, whereas these markers are negative in sinonasal glomangiopericytoma. Uncommon tumors for this site, like synovial sarcoma with spindle cell component, are also CD99 positive and show β catenin nuclear staining, but it is usually biphasic and CK positive with characteristic chromosomal translocation t(X;18) (p11; q11). Other pericytic tumors, like glomus tumors, are also SMA positive on IHC but can be distinguished morphologically by the presence of compact epithelioid cells in addition to negative nuclear β catenin staining. Ewing sarcoma is positive for CD99 and several other markers along with EWSR1 rearrangement but does not show nuclear β catenin positivity.

Treatment is mainly surgical as these tumors are radioresistant and warrant complete surgical excision with negative margins. Perioperative bleeding and operation time can be reduced by digital subtraction

angiography (DSA), preoperative embolization, and intraoperative use of the coblation technique. Long-term follow-up is recommended to manage recurrence. [25]

Our case showed bland spindle cell morphology with characteristic sub-epithelial grenz zone and staghorn vasculature. IHC showed β catenin (nuclear staining) with TLE1, CD99, and SMA positivity. Other markers like pankeratin, S100, STAT6, CD31, CD34, ERG, HMB45, NKX2.2, SOX10 were negative and H3K27Me3 was retained. These markers helped to rule out morphologically or immunohistochemically overlapping tumors discussed above that are either specific or rare in the sinonasal regions like biphenotypic sinonasal sarcoma, melanoma, solitary fibrous tumor, synovial sarcoma, Ewing sarcoma, and peripheral nerve sheath tumors.

This case has been discussed here due to its presentation as a nasal mass with epistaxis and nasal obstruction, which in a rural-dominant practice points to more common diagnosis like nasal and sinonasal inflammatory lesions or epithelial neoplasm. Knowledge of this rare sinonasal tumor will help the pathologist diagnose it more accurately and aid in its management and follow-up.

Conclusion

To conclude, sinonasal masses can have a varied differential diagnosis, ranging from inflammatory lesions to neoplastic lesions. Its management depends upon preoperative evaluation with the help of endoscopy, CT, MRI, and angiography, followed by histopathological and immunohistochemical confirmation of diagnosis. Complete surgical resection with regular long-term follow-up is required to diagnose possible recurrence.

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