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Case Report

Sinonasal Glomangiopericytoma: A Case Report

Raji T Naidu[1], Susan Cherian[2], Vaishnavi Kumba[3], Uma Chaturvedi[4]*

*1*Consultant, Pathology Dept., BARC Hospital, Mumbai.*

2 Consultant & Head, Pathology Dept., BARC Hospital, Mumbai.

3 Post graduate DNB student (Pathology), BARC Hospital, Mumbai.

4 Consultant, Pathology Dept., BARC Hospital, Mumbai.

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Abstract

Sinonasal glomangiopericytoma is defined as a low-grade malignant tumor demonstrating a perivascular myoid phenotype. We report a case of sinonasal glomangiopericytoma having a diffuse architecture with bland spindle cells arranged in short fascicles, storiform, whorled, reticular pattern, separated by a vascular plexus ranging from capillaries to large patulous spaces. Immunohistochemistry is required to differentiate it from other perivascular tumours and solitary fibrous tumour. However, differentiating glomangiopericytomas and solitary fibrous tumours based on immunohistochemistry is a challenge. The tumour expressed vimentin, focal smooth muscle actin and CD34, diffuse, strong Bcl2 and Beta catenin and negative STAT6. Immunohistochemical expression of STAT6 is useful as a negative marker in the diagnosis and separation of glomangiopericytoma from solitary fibrous tumour.

Key Messages: Sinonasal glomangiopericytoma (SNGP), a rare tumour with low malignant potential as per WHO classification is frequently confused with sinonasal solitary fibrous tumour (SFT). This article emphasizes the importance of using a broader immunohistochemical panel of positive and negative markers for differentiating SNGP from SFT. The most useful among them being CD34, SMA, BCL2, Beta Catenin and STAT6.

Keywords: sinonasal glomangiopericytoma, immunohistochemistry, solitary fibrous tumour

Address for Correspondence:
Dr Raji T Naidu,
Consultant, Pathology Dept.,
BARC Hospital, Anushakti Nagar, Mumbai- 400094
Email-Id: drrajipillai@gmail.com
Tel: 022-25598351, Mob: 9619920707.

INTRODUCTION

Sinonasal glomangiopericytoma (SGNP) or sinonasal hemangiopericytoma- like tumour is defined as a low grade malignant/ borderline sinonasal tumour demonstrating a perivascular myoid phenotype.[1] It has a "patternless" diffuse architecture on histology with cells arranged in short fascicles, storiform, whorled, reticular pattern, separated by a vascular plexus similar to soft tissue hemangiopericytoma or solitary fibrous tumour (SFT). Hence differentiating glomangiopericytomas from SFT requires ancillary techniques like immunohistochemistry (IHC) or molecular biology.[1] In the present case report we have used IHC markers to differentiate SNGP from SFT.

CASE REPORT

A 81-year-old male presented with nasal obstruction since a year, left more than right associated with right nostril mucoid discharge. On anterior rhinoscopy, a fleshy reddish mass was observed in the left nostril with mild DNS to right anteriorly. Fiber-optic endoscopy through the left nostril showed mild DNS to right, pale nasal mucosa, a fleshy pinkish red mass. The endoscope could not be passed beyond the mass. Right nostril endoscopy showed no mass in right nostril and nasopharynx. On CT evaluation- unilateral mass pushing septum to opposite side and extending up to posterior ethmoid and sphenoid, not eroding the bones was evident. Endoscopic excision of left nasal mass under general anaesthesia was done. Grossly, we received grossly polypoid, friable and haemorrhagic bits aggregating to (2.7x 2.5x 2.2) cm. On microscopy, an unencapsulated tumour was seen under an intact mucosal epithelium, having a "patternless" diffuse architecture with spindle cells arranged in short fascicles, storiform and whorled pattern separated by vascular plexus ranging from capillaries to large patulous spaces (Figures 1). Peritheliomatous hyalinization was noted. The spindle cells had moderate cytoplasm, indistinct cell borders with minimal atypia (Figure 2). There was no necrosis; mitotic figures were rare. The differential diagnosis included SNGP and soft tissue type hemangiopericytoma or solitary fibrous tumour (SFT). Results of immunohistochemical evaluation are as given in Table 1. The tumor expressed strong and diffuse positivity for Beta catenin (Figure 3), weak and focal positivity for CD34 (Figure 4) and CD99 (Figure 5), strong and diffuse positivity for BCL2 (Figure 6) and was negative for S100, Desmin and STAT6 (Figure 7). A final diagnosis of SNGP was made based on immunohistochemistry.

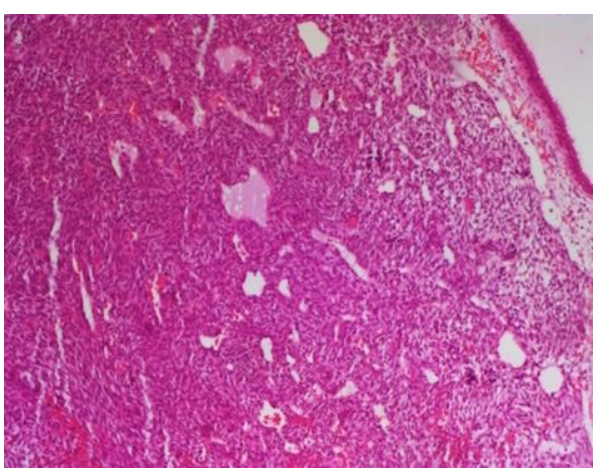

Figure 1: Submucosal tumour having patternless, diffuse architecture and prominent vascular spaces, H & E x50

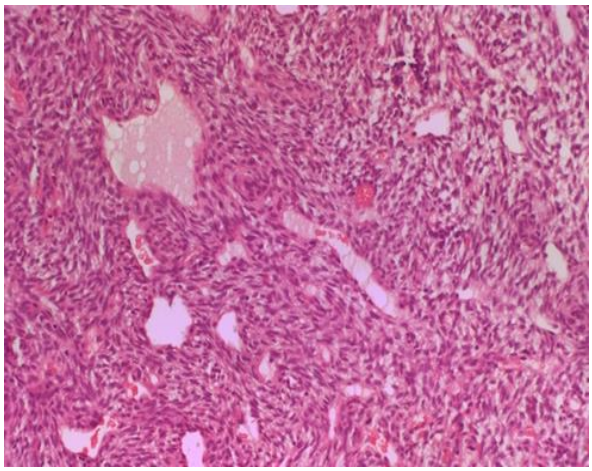

Figure 2: Spindle cells of tumour have moderate cytoplasm, indistinct cell borders with minimal atypia and no necrosis, H & E x200.

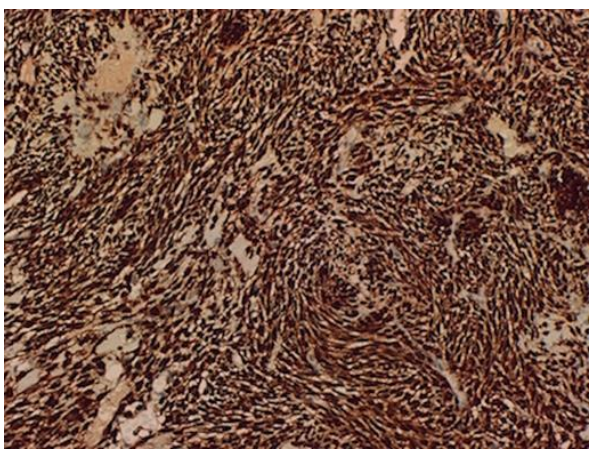

Figure 3: Immunohistochemical staining showing diffuse nuclear positivity for Beta catenin expressed in tumour cells, 200x

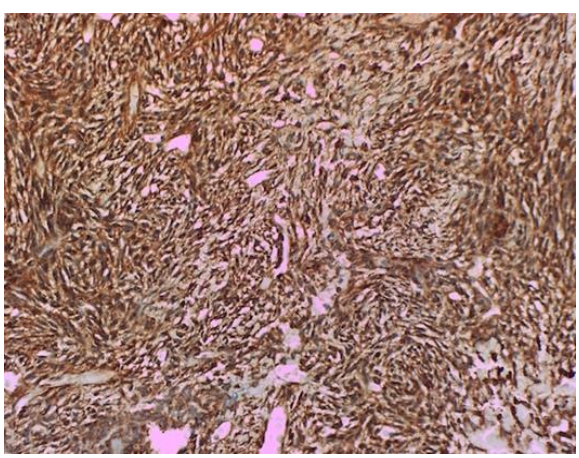

Figure 4: Tumour cells showing focal positivity for CD99, 200x

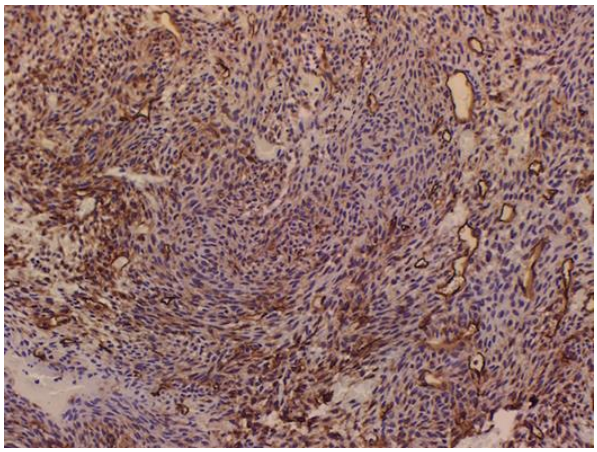

Figure 5: Tumour cells showing Weak focal positivity for CD34, 200x

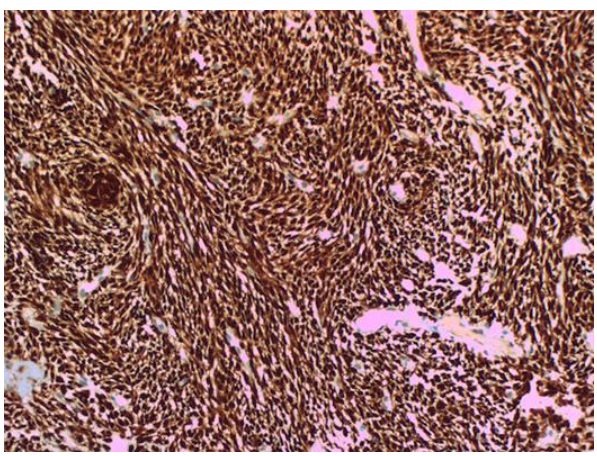

Figure 6: Strong and diffuse positivity is expressed by tumour cells for BCL2, 200x

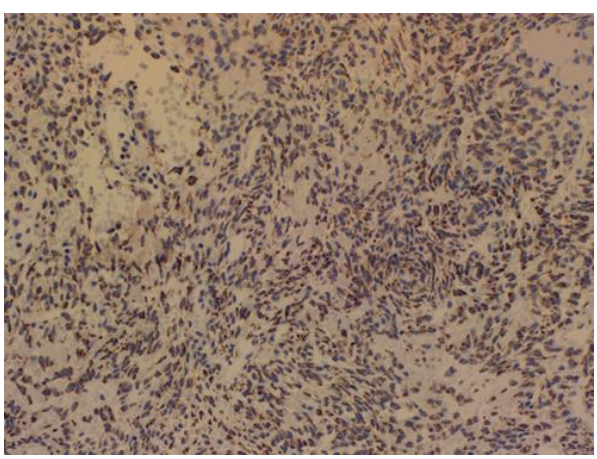

Figure 7: Tumour cells negative for STAT 6 nuclear stain by immunohistochemistry, 200x

Table 1: Results of immunohistochemistry (Original)

Positive markers	Negative markers
Vimentin	S 100
Beta catenin (diffuse nuclear positivity)	Desmin
BCL2 (diffuse and strong)	STAT 6
CD99 (weak, focal)	-
CD34 (weak, focal)	-

DISCUSSION

SNGP accounts for less than 0.5% of all sinonasal tract neoplasms with a slight female predilection and a peak incidence in the seventh decade of life, although individuals of any age may be affected.[1] The tumour is nearly always unilateral (only 5% are bilateral), affecting the nasal cavity alone and frequently extending into paranasal sinuses. Etiology of SNGP remains unknown , however past trauma, hypertension pregnancy, and use of corticosteroids are considered predisposing factors.[2] The most common presentation includes unilateral nasal obstruction, usually over 6 months to one year and epistaxis. Radiological evaluation by CT scan or MRI is needed to know the location and extent of tumour. Histopathologic evaluation is necessary for diagnosis. The tumour is sub-mucosal and consists of closely packed bland spindle cells. The cells are separated by distinctive vascular network comprising of variably sized vascular channels. Peritheliomatous hyalinization is a characteristic feature. Mitoses are less than 3 per 10 high-power fields and nuclear pleomorphism is absent to mild.[1] Mast cells, eosinophils, and extravasated erythrocytes are variably present. Malignant glomangiopericytomas show profound pleomorphism, necrosis, and increased mitoses.[3] Mutation in CTNNB1 gene that encodes Betacatenin protein has been described in SNGP.[4] The tumour needs to be differentiated from other perivascular mesenchymal tumours like glomus tumours and myopericytoma. In general, typical glomus tumour consists of more rounded cells with well-demarcated cell membranes arranged in a perivascular growth pattern. Myopericytoma and myofibroma tend to have larger, less rounded cells. Additionally, the myofibroma typically consists of a biphasic growth of primitive short spindled cells growing around hemangiopericytomatous vascular spaces and fascicles and whorls of plump spindled cells. The myopericytoma is characterized by the presence of plump spindled cells that grow in a concentric fashion around open vessels. Nuclear b-catenin immunoreactivity is not observed in myoperictoma, which may be helpful in its distinction from SNGP.[5]

SNGP is most frequently confused with solitary fibrous tumours.[6] By immunohistochemical analysis, glomangiopericytomas usually show diffuse reactivity with SMA, nuclear betacatenin, cyclin D1, factor Xllla, and vimentin, and lack significant expression of CD34, CD31, CD117, STAT6, BCL2, cytokeratin, EMA,

desmin, or S100 protein.[1] In the present case differentiating the tumour from SFT was a challenge. SFT tumour has variable cellularity with hypo and hypercellular areas and shows presence of similar vascular plexus as SNGP.[7] On immunohistochemical analysis, results obtained were as given in Table 1.

Though the tumour cells expressed diffuse positivity for beta catenin, its expression is not unique to SNGP, its expression has also been seen in SFT. [5,8] CD34 expression was focal. Various reports in literature mentions focal weak CD34 expression to diffuse and strong expression in SNGP.[9] Diffuse BCL2 expression, as seen in soft tissue type hemangiopericytoma was observed. Recently STAT6 has emerged as a more reliable marker for the diagnosis of SFT. Nuclear expression of STAT6 associated with NAB2-STAT6 gene fusion is a specific marker for SFT.[8] Hence additional IHC for STAT6 was carried out, which was negative. A final diagnosis of SNGP was made based on positive and negative markers. The treatment of choice is endoscopic complete resection. Preoperative embolization of afferent vessels can facilitate surgical resection by reducing the blood supply to the tumour.[6] The tumour, classified as tumours with low malignant potential has excellent prognosis, however reported rate of recurrence in literature ranges from 30% to 50%, attributable to incomplete resections. [1,3,4] The present case has been followed up for a year and there is no metastases or recurrence till date.

CONCLUSION

SNGP, a rare tumour with low malignant potential as per WHO classification is frequently confused with sinonasal SFT. Differentiating SNGP from SFT requires a broader immunohistochemical panel of positive and negative markers. The most useful among them being CD34, SMA, BCL2, Beta Catenin and STAT6. It is an indolent tumour with excellent prognosis. Recurrence due to incomplete resection is observed, requiring post-operative follow up.

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Conflicts of interest

There are no conflicts of interest

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