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Case Report

Solitary fibrous tumor of nasal cavity: A case report

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ABSTRACT

Solitary fibrous tumor is fibroblastic tumor, located mostly in deep soft tissue as well as extrapulmonary sites. In the present case, a 27-year female presented with complain of bilateral nasal blockage (right >left) with history of nasal bleed. The CT scan report revealed p/o hypervascular nasal polyp and sinusitis. Patient underwent excision of granulomatous mass arising from lateral nasal wall in right nasal cavity. The pathological report stated nasal tumor with hemangiopericytoma like pattern with differential diagnosis of 1) Solitary fibrous tumor – right nasal cavity 2) Glomangiopericytoma; further Immunohistochemistry was positive for CD34 (membranous) and STAT6 (nuclear) which leads to final diagnosis of Solitary fibrous tumor – right nasal cavity. Histopathological characteristics serve as the primary criteria for differentiating this condition from other entities.

1. Introduction

Solitary fibrous tumor (SFT) is primarily a tumor of adult life, affecting both the sexes equally. It is predominantly situated in deep soft tissue with particularly predilection for thigh, pelvic fossa, abdominal cavity, retroperitoneum and serosal surfaces.[1] Tumors in these locations often chiefly show cellular features which are now considered SFT (previously labelled as 'hemangiopericytoma'). Although previously thought to be confined to the pleura, tumors exhibiting characteristics of classic solitary fibrous tumors (SFT) are now also increasingly identified in extrapulmonary locations like head and neck, trunk.[2–5]

2. Case Report

A 27-year old female came to Otorhinolaryngology (ENT) OPD at tertiary care hospital, Ahmedabad with chief complain of B/L nasal blockage (right>left). Patient had a history of nasal bleed.

Computed tomography of para nasal sinus (CT PNS) findings showed homogeneous enhancing polypoidal soft tissue lesion in right anterior nasal sinus, suggest possibility of hypervascular nasal polyp. Also, there is presence of Right maxillary sinus polyp. Deviated nasal septum towards left side with bony septal spur impinging inferior turbinate. Mucosal thickening of right ethmoid sinus, suggest possibility of sinusitis.

Patient underwent excision of granulomatous mass arising from lateral nasal wall in right nasal cavity.

3. Result

3.1. Gross examination

Specimen consist of multiple whitish soft tissue portions measuring 2.0 X 1.5cm in aggregate.

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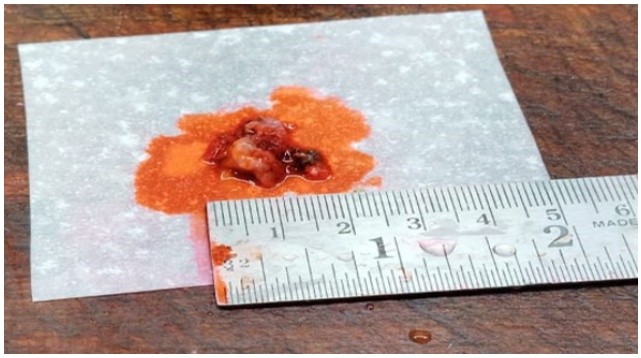

Figure 1: Gross image of nasal biopsy specimen

3.2. *Microscopic examination*

Section reveal tumor to be composed of cells that are ovoid to spindle-shaped with indistinct borders, arranged in a haphazard manner and forming ill-defined fascicles. It shows presence of dilated, branching, hyalinized staghorn like (Hemangiopericytoma like) vasculature.

Findings are suggestive of nasal tumor with Hemangiopericytoma like pattern with following differential diagnosis:

1. Solitary fibrous tumor- right nasal cavity
2. Glomangiopericytoma

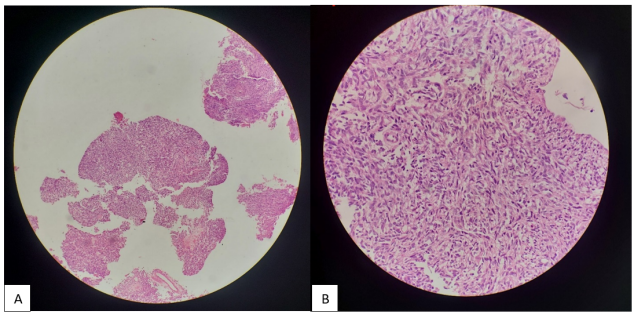

Figure 2: A,B): H&E stain showing ovoid to spindle cells arranged haphazardly and as ill-defined fascicles (10X, Left) and (40X, Right) respectively

3.3. *Immunohistochemistry*

1. Light microscopy:
2. CD34: Positive (membranous)
3. STAT 6: Diffusely positive (nuclear)

3.4. *Diagnosis*

Immunohistochemistry findings are suggestive of Solitary fibrous tumor of nasal cavity. No e/o significant mitosis or necrosis is seen.

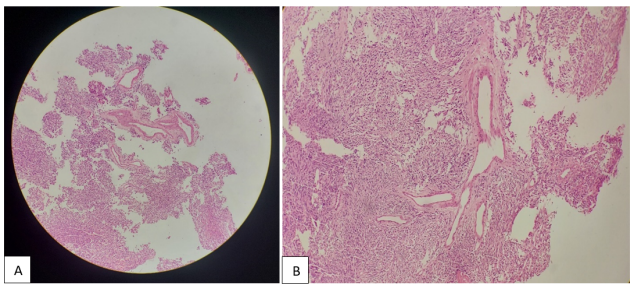

Figure 3: A, B): H&E stain showing staghorn like blood vessels (10X, Left) and (40X, Right) respectively

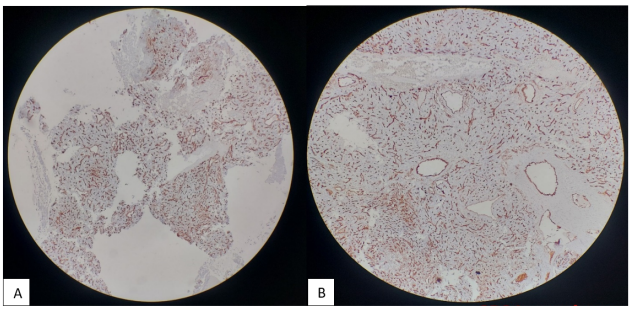

Figure 4: CD34 positive (10X, Left); CD34 positive (40X, Right)

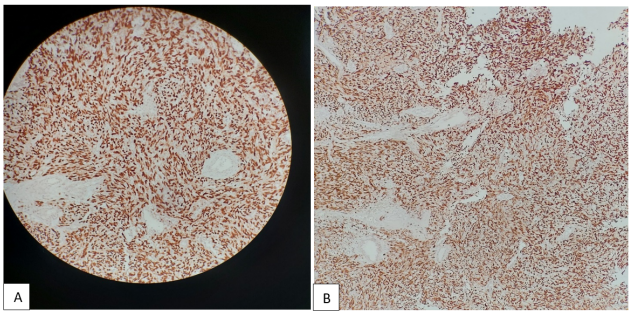

Figure 5: STAT6 positive (40X, Left); STAT6 positive (10X, Right)

4. Discussion

The tumor may occur in any anatomic site, with approximately 5–27% of SFTs arising in the head and neck region.[6–8] Within this anatomic region, preferred sites of involvement include the oral cavity and orbit.[7–10] In contrast, SFT infrequently affects the sinonasal tract (SNT). Due to its relative rarity and variable morphologic appearance, sinonasal tract SFT may be difficult to distinguish from other mesenchymal lesions that are more commonly recognized at this site.

In solitary fibrous tumors of the nasal cavity, the vessels form a continuous, ramifying vascular network that exhibits marked variation in size and shape i.e caliber. Typically the dilated, branching vessels divide and communicate

with smaller vessels, accompanied by surrounding cellular proliferation which seems to obscure and partially compress it. The dividing sinusoidal vessels often exhibit a "staghorn" or "antler-like" configuration.

The cellular phase of SFT consists of tightly packed, round to fusiform cells with indistinct cytoplasmic borders which surrounds an elaborate vasculature. The cells, ranging from spindled to oval, are arranged in short fascicular, storiform, whorled or mixed patterns. The thin-walled vessels are occasionally staghorn shaped and hyalinized. Atypia is generally absent and mitotic activity low (1/10hpf).

Immunohistochemistry was positive for CD34 (membranous) and STAT6 (nuclear) which leads to final diagnosis of Solitary fibrous tumor – right nasal cavity.

Molecular Genetics shows NAB2-STAT6 fusions with breakpoint between NAB2 exon 4 and STAT6 exon 2 which is characteristic of Solitary fibrous tumor.

Differential Diagnosis:

1. Glomangiopericytoma: Minority of cases express CD34, the classic marker of the solitary fibrous tumor ("hemangiopericytoma") family of lesions and STAT6 expression is absent. [11,12]
2. Deep benign fibrous histiocytoma: Usually show more prominent, uniform spindle cell pattern than SFT. Tumour cells form well developed storiform growth pattern.
   (a) They show variable expression of CD3 and STAT6 negative. STAT6 may be helpful in difficult cases.
3. Synovial sarcoma (monophasic): Hypercellular spindle cell neoplasm with variable fascicular growth; prominent staghorn vasculature is uncommon but present focally. They are associated with distinct spindle cells and hyalinised calcified areas.
   (a) Focal cytokeratin (+) and/or EMA (+) expression, diffuse TLE1 (+) is seen. CD34 expression is not seen.
   (b) Molecular genetics studies show t(X; 18) with SS18-SSX1/2 fusions.
4. Mesenchymal chondrosarcoma: It shows Hemangiopericytoma - like vascular pattern in closely packed, small cell areas. It also shows island of well differentiated cartilage or much less frequent bone.

5. Conclusion

Solitary fibrous tumor is fibroblastic tumor characterized by prominent, branching, staghorn vasculature. It was originally reported as localized fibrous mesothelioma. Nuclear immunoreactivity for STAT6 protein, represents NAB2-STAT6 fusions that is characteristic of SFT, has been reported in almost 100% of SFTs in several large recent studies. [13,14] Cellular solitary fibrous tumors (SFTs) express CD34, though typically in a smaller percentage of cases and to a lesser extent compared to more classic, hyalinized tumors.

6. Source of Funding

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7. Conflict of Interest

No competing interest.

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