CASE REPORT

Sinonasal Synovial Sarcoma Masquerading as A Common Soft Tissue Tumor: A Diagnostic Challenge and Literature Review

Pranshu Saklani[1], Shruthi N Shetageri[2], S R Raja Parthiban[3]

ABSTRACT

Synovial sarcoma is a malignant soft tissue neoplasm which amounts to 7-10% of all sarcomas. Clinicopathological heterogeneity within this tumour creates a diagnostic challenge in sorting it out from its differential diagnoses. Here we report a 42-year-old female patient presenting with a mass in the right nasal cavity for two months. With an imaging impression of a right ethmoidal polyp with a remote possibility of malignant etiology, a final diagnosis of sinonasal synovial sarcoma was made with the help of a panel of IHC antibodies. A high degree of suspicion along with a detailed work up is required to accurately diagnose synovial sarcoma in unlikely locations such as nasal cavity.

KEY WORDS: Synovial sarcoma, Nasal cavity, TLE1, BCL2, SOX 10.

Background

Synovial sarcoma is an aggressive high-grade sarcoma which amounts to 7-10% of all sarcomas. With a male predilection, tumor is more often seen in the age group of 15 to 40 years. In most series, extremities (85-95%) have been the commonest site followed by head and neck region (5-10%).[1] The hypopharynx is the most frequently involved site in the head and neck followed by masticator space, parapharyngeal space and sinonasal region.[2]Clinicopathological heterogeneity within this tumour poses a considerable diagnostic challenge in sorting it out from its differential diagnoses. In comparison to other adult soft tissue sarcomas, it is more chemosensitive and hence its definitive diagnosis is critical for the patient management.[3]This case report deals with the possible differentials that come into picture while diagnosing synovial sarcoma on a small biopsy in a rare site like nasal cavity and the selection of right set of IHC in reaching the correct diagnosis.

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Case report

We received a nasal biopsy specimen of a forty-two-year-old female who presented with complaints of headache, nasal obstruction and mass in the right nasal cavity for two months. On examination, a mass occupying the right nasal cavity measuring 2x2cm was noted. Patient was advised CT scan of the nasal cavity and paranasal sinuses. On CT scan, a heterogeneously enhancing partially calcified lesion m/s 3.6x 2.4x 2.0 cm epicentered in the posterior right ethmoid blocking the right choana and posteriorly extending to sphenoethmoidal recess and laterally blocking the right OMU (Ostiomeatal unit) was seen. There was mucosal thickening in the right frontal, maxillary and sphenoid sinuses. A possibility of a right ethmoidal polyp with a remote possibility of malignant etiology and to correlate with histopathology was suggested on CT scan. In view of this, the patient was further advised for a Contrast-enhanced MRI of the Brain which showed that there was no intraorbital or intracranial extension with a mass lesion measuring 4.2x 4.0 x 1.9 cm in the right

[1]PG, Department of Pathology, MVJ Medical College & Research Hospital, Hoskote, Karnataka, India, [2]Associate Professor, Department of Pathology, MVJ Medical College & Research Hospital, Hoskote, Karnataka, India, [3]Professor & HOD, Department of Pathology, MVJ Medical College & Research Hospital, Hoskote, Karnataka, India

Address for correspondence:
Shruthi N Shetageri, Associate Professor, Department of Pathology, MVJ Medical College & Research Hospital, Hoskote, Karnataka, India. E-mail: dr.shruthi.n.s@gmail.com

nasal cavity and right ethmoid sinus.

After routine hematological investigations, the patient underwent an incisional biopsy of the lesion. On gross examination, a grey-white, soft and bony tissue bits measuring 0.4x 0.2x 0.1 cm were received which was subjected to histopathological examination.

Microscopy revealed multiple bits of tumor tissue covered by respiratory epithelium with focal ulceration. Subepithelium showed an underlying cellular tumor composed of spindle-shaped cells arranged in interlacing bundles and fascicles with a rich vascular network and at places exhibiting a staghorn like pattern. Tumor cells were spindle, with scant cytoplasm, elongated hyperchromatic nucleus and inconspicuous nucleoli. Areas of hemorrhage, edema and myxoid changes were noted. Mitotic figures of 0-1/ hpf was seen. No necrosis or calcification was observed. Tumour was seen infiltrating the bony trabeculae. Figures 1 and 2

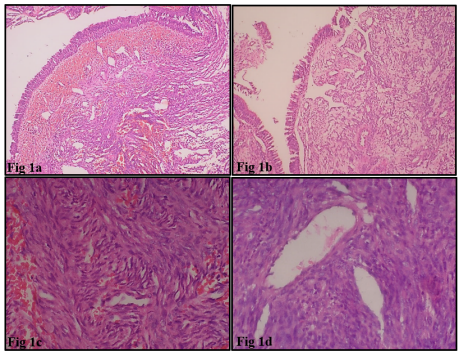

Figure 1: A & B: Bits of tissue covered by respiratory mucosa with an underlying cellular soft tissue tumor. Subepithelium shows hemorrhage and edema with chronic inflammatory cell infiltrate. (H&E, 4x). C & D: Tumor cells which are spindle in shape with hyperchromatic nucleus and moderate eosinophilic cytoplasm and inconspicuous nucleoli. Also seen are slit-like blood vessels. (H&E, 40x)

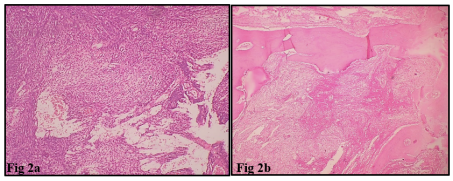

Figure 2: A: Hypocellular areas with myxoid change.(H&E, 10x). B: Tumor cells invading the adjacent bony tissue. (H& E, 10x)

With these histopathological features, a diagnosis of cellular spindle cell neoplasm with the following differential diagnosis was considered.

1. Cellular schwannoma
2. Synovial Sarcoma
3. Solitary fibrous tumor (SFT)
4. Biphenotypic Sinonasal sarcoma (BSNS)

Immunohistochemistry was performed to confirm the diagnosis and rule out the differentials. The primary panel included EMA (epithelial membrane antigen), Desmin, SMA (Smooth muscle antigen), CD 34, S100 and Ki 67.

S 100 was diffusely positive (Figure 3a) while EMA, Desmin, SMA and CD 34 were negative. Tumour showed a low proliferation index of 3.5%. Cellular Schwannoma was considered as the first possibility due to the diffuse positivity of S100. Due to the infiltrative nature and ill circumscription of the tumor mass, we further performed IHC for TLE 1 and BCL 2 to rule out synovial sarcoma. TLE 1and BCL 2 were diffusely and strongly positive suggesting the possibility of synovial sarcoma. (Figure 3b, c)

The diffuse positivity of S100 in synovial sarcoma is a rare entity. Contrary to this, cellular schwannoma can also show TLE 1 positivity. Hence, we performed IHC for SOX 10 which is highly specific and sensitive for Schwannoma. Tumor cells were negative for SOX 10 **(**Figure 3d) which helped us in ruling out the possibility of Schwannoma.

Keeping in consideration of the histopathological features and IHC findings (S 100, TLE 1 and BCL 2 positive and negative for EMA, Desmin, SMA, SOX 10), a final diagnosis of Synovial Sarcoma of nasal cavity involving the paranasal sinus with bone invasion was made.

Discussion

Synovial sarcoma is a misnomer as the tumor does not arise from synovial tissue but instead has its origin from the pluripotent mesenchymal cells which variably can display epithelial differentiation. [2] The first synovial sarcoma was described by Jernstrom in 1954. [4]

According to a review done by Saito et al in 2018, sinonasal synovial sarcoma is an extremely

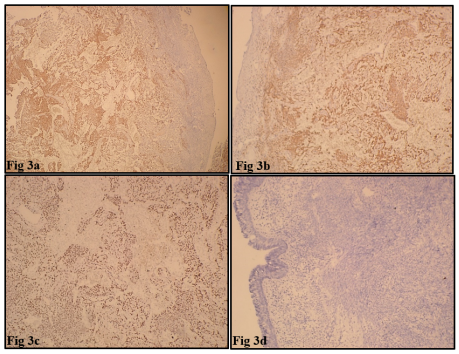

Figure 3: A-D: Tumour cells show strong positivity to S100, BCL2 and TLE 1 and negative to SOX 10 (IHC; 4x)

rare tumor, with only11cases being reported till date. The conventional treatment for any sarcoma is complete resection with leaving a safe margin, which becomes difficult in head and neck cases because of the complex anatomy. Management of head and neck sarcomas becomes quite challenging with a requirement for a multimodal approach in many cases. [5]

Synovial sarcoma may reveal a SFT type morphology with hemangiopericytomatous vessels and share positive immunostaining for EMA, CD99, and Bcl-2. However, the negative immunoreactivity to CD34, aids in differentiating it from SFT. [6] The recent novel marker identified for synovial sarcoma is Transducin-like enhancer of split 1 (TLE1) which is a repressor of transcription and is important for hemopoiesis, neuronal and terminal epithelial differentiation. [7] Literature indicates 85 to 97% of synovial sarcomas to beTLE1 positive; however, the positivity is not limited to only synovial sarcoma. TLE-1 can be variably positive in other tumors like, Schwannoma, Malignant peripheral nerve sheath tumor, Endometrial stromal sarcoma, Ewing sarcoma, Epithelioid sarcoma, Solitary fibrous tumor. [6]

An important differential diagnosis in our case was cellular schwannoma which usually poses a diagnostic difficulty due to its overlapping histological and immunohistochemical features with synovial sarcoma. This tumor was first described by Woodruff et al in 1981. [8] Though, head and neck region is a common site (25-50%), for the occurrence of schwannomas, only 4% are known to have origin from the paranasal sinuses or nasal cavity. [9] Histologically has hypercellular Antoni A areas with hyperchromatic cells showing minimal pleomorphism and mitotic activity admixed with thick-walled hyalinized blood vessels. Thus, alarming features in a case of a cellular schwannoma include increased cellularity, increased mitoses, lack of encapsulation, locally aggressive nature and bone erosion. [10] Based on these aggressive features, approximately one-third of cellular schwannomas were misdiagnosed as malignant in the past. Immunohistochemistry usually reveals a strong and diffuse expression of S100 and SOX 10 in these cases. In all suspected cases of cellular schwannoma, the absence of SOX10 immunoreactivity helps in differentiating tumors with monophasic histology like synovial sarcoma, fibrosarcoma and leiomyosarcoma. [11]

S100 expression in synovial sarcoma is seen in upto 30% of cases. [1] Distinguishing S100 positive cellular spindle cell neoplasms on small biopsy specimens is quite challenging. Hence, whenever we encounter these lesions, t (X;18) demonstrated by fluorescence in situ hybridization (FISH)or immunohistochemical staining with TLE1 is important to exclude synovial sarcoma from the differentials. [12] Karamchandani et al in 2012 reported 15% of the total 75 cases of synovial sarcoma to be S 100 positive with no evidence of SOX 10expression. A combination of negative staining to SOX10 and positivity to TLE1 helps in providing a strong support for the diagnosis of synovial sarcoma and thereby ruling out the possibility of a cellular schwannoma. [13]

Depending on the site and morphology of the tumor, BSNS was one of the differentials which needed to be excluded in the study. BSNS is a rare low-grade sinonasal sarcoma first described by Lewis et al in 2012 which has both neural and myogenic differentiation. [14] The tumor is infiltrative and has hypercellular fascicles of bland spindle cells with consistent immunoreactivity to S100 and smooth muscle actin (SMA) thus, describing the term "Biphenotypic". [15] It can be locally aggressive and microscopic bone invasion has been documented in almost 20% of cases. Existing data reveals that most BSNSs can be diagnosed by positive expression of S100 with either actin or calponin and absence of cytokeratin and SOX-10. [16] In our case, EMA, SMA and SOX 10 were negative, thus helping us in ruling out this entity.

Conclusion

Most of the sinonasal tract tumors are epithelial in origin and soft tissue tumors when present belong largely to vascular or fibrohistiocytic category.

As there are numerous differentials for spindle cell lesions that simulate synovial sarcoma, a thorough histopathological examination is of prime significance. While evaluating spindle cell tumors in an odd location like nasal cavity, an adequate number of IHC markers are essential in arriving at a conclusive diagnosis as the initial IHC panel can be misleading. Thus, this case report highlights not only the importance of considering synovial sarcoma in the differential diagnosis of a spindle cell tumor in sinonasal space but also the value of a complete IHC workup in all these challenging cases.

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