Case Report

A RARE NASAL TUMOR WITH NEURAL AND MYOGENIC DIFFERENTIATION: BIPHENOTYPIC SINONASAL SARCOMA

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ABSTRACT:
Biphenotypic sinonasal sarcoma is a newly recognized mesenchymal tumor that has been included in the upgraded classification of sinonasal tumors by WHO. This is a low-grade spindle cell malignancy previously categorized as fibrosarcoma or peripheral nerve sheath tumor. We report a case of Biphenotypic sinonasal sarcoma arising in a middle-aged female. The patient came with the chief complaints of left nasal discharge and epistaxis. CT report was suggestive of left-sided sinonasal polyposis. After the surgery, histopathological and immunohistochemistry (IHC) analysis confirmed the diagnosis of Biphenotypic sinonasal sarcoma. This highlights the importance of IHC in this new entity to decrease the morbidity and mortality in such cases.

Key Words: Immunohistochemistry, Sarcoma, Mortality

INTRODUCTION:

Biphenotypic sinonasal sarcoma (BSNS) is a recently documented entity in the World Health Organization classification for head and neck tumors. This lesion is a rare form of malignancy arising primarily in the nasal tract.[1] This lesion, exhibiting characteristics of both neural and myogenic differentiation, carries a high preponderance for middle-aged females.[2] Researchers have found that fusion of paired box gene 3 (PAX3) and mastermind like transcription coactivator 3 (MAML3) genes give rise to this Biphenotypic tumor.[3] It has the predilection to quickly invade surrounding facial structures in an outward fashion typically towards each of the nostrils. The failure in early detection of this low-grade sarcoma and its infiltrative pattern makes it difficult to treat. Furthermore, attempts made at surgical removal of this lesion results in facial disfigurement.[4] The patient was a 50-year-old woman. She presented with a history of left nasal discharge for 01 year while bouts of epistaxis for last 01 month.

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CASE REPORT:

On examination, there was a widening and expansion of the left nasal cavity. Computed Tomography of Nose and Paranasal Sinuses (CT PNS) revealed a soft tissue expansile lesion in left-sided maxillary antrum filling maxillary sinus causing the widening of osteomeatal complex. The mass was noted to be extended up to the ipsilateral nasal cavity, ethmoidal air cell, sphenoidal and frontal sinuses. The left Cribriform plate was found to be thinned and eroded. However, there was no evidence of intracranial involvement. CT findings were suggestive of left-sided sinonasal polyposis. Laboratory investigations showed her lab values were in the normal range.

The patient was prepared for the surgical removal of the tumor. After surgery, the excised mass was sent to the Department of Histopathology at Akhter Saeed Medical and Dental College for confirmatory diagnosis.

The gross examination of the mass showed grey-white tissue fragments measuring approximately 1.2 cm in aggregate. While the histopathological examination of excised tissue section revealed a spindle cell neoplasm composed of fascicles and sheets of pleomorphic round to oval cells with uniform elongated nuclei. These cellular

fascicles were surrounded by delicate strands of collagen fibers. However, there were few areas that showed characteristic epithelial proliferation in small cystic spaces often forming glands. The intervening stroma revealed moderate chronic inflammatory cellular infiltrate and congested blood vessels. Mitotic figures were infrequent.

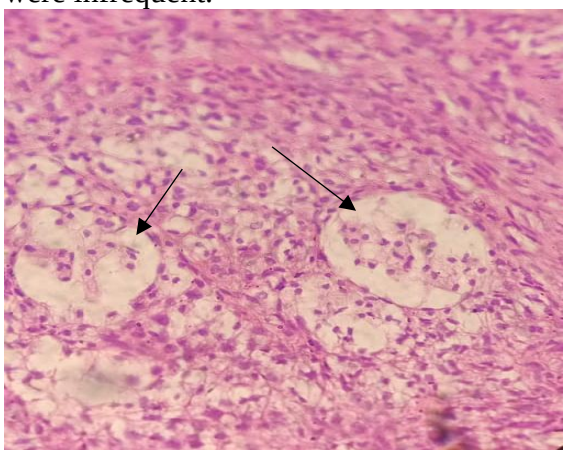

Fig 1: Photomicrograph shows cellular neoplasm with epithelial proliferation in small cystic spaces.

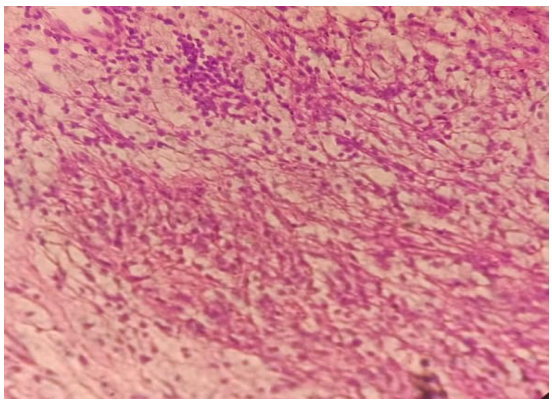

Fig 2: Photomicrograph shows cellular fascicles with intervening delicate collagen fibers.

On immunohistochemical studies, the tumor cells showed diffuse positivity for S-100 protein and only focal positivity for Smooth muscle actin (SMA). Whereas, expression of Desmin, Myogenin, Cytokeratin (CK) and HMB45 were found to be negative in the tumor. After a panel of immunohistochemical stains, a final diagnosis of Biphenotypic sinonasal sarcoma was made. The postoperative course was uneventful. The patient was referred to an oncologist for further treatment and follow-up.

DISCUSSION:

Biphenotypic sinonasal sarcoma is a low-grade, uncommon sarcoma that was first presented by Lewis et al [2] in 2012 as 28 cases of low-grade sarcoma with myogenic and neural differentiation which were negative for Synovial Sarcoma Translocated to X chromosome protein (SYT–SSX) chimeric transcript of synovial sarcoma.

The reported cases of BSNS show that it primarily affects women in the adult age group (24-78yrs) like our case.[2,5,6,7,8] Studies showed that patients present with nonspecific symptoms of nasal obstruction, like problems in nasal breathing, bouts of bleeding, pain and congestion in sinonasal areas the same as in our case. BSNS presents as a locally destructive tumor that involves multiple sinonasal sites, with the most commonly involved sites are superior nasal cavity and ethmoid sinus, and then by the sphenoid sinus. The invasion may occur beyond the sinonasal area, commonly into the orbit region (25% of cases) and via the cribriform plate (10% of cases). The CT scan findings of our patients were consistent with the previous literature and luckily with no evidence of intracranial involvement.[2]

Histopathologically, BSNS consists of an infiltrative, extremely cellular low-grade spindle cell lesion that has the long and slender tapered proliferation of uniform spindle cells, with syncytial borders, and nuclei with vesicular chromatin like in our case in which spindle cells are arranged in fascicles and sheets. Mitotic activity is low whereas necrosis is not a feature of BSNS. Commonly, there are entrapped respiratory epithelium as in our case epithelial proliferation in the form of glands and cystic spaces was seen.[2,7,9]

The immunohistochemical panel proposed by Rooper et al[9] is based on initial cases reported,[3,6,9] includes S100, SMA, β-catenin, desmin, SOX10, calponin, myogenin, factor XIIIa, and CK. BSNS cases mainly show at

least focal S100 expression[7,9] as well as of SMA and calponin. There could be a variable expression of factor XIIIa, desmin, myogenin, and negative expression for CK and SOX10. Immunohistochemical findings in this case were consistent with the previous data {positive for S-100, focal positivity for SMA, while negative expression of Desmin, Myogenin, Cytokeratin (CK) and Human melanoma black 45 (HMB45) were found in tumor cells}.

CONCLUSION:

BSNS was initially reported as a low-grade fibro-sarcoma or low-grade peripheral nerve sheath tumor prior to its description by Lewis et al.[2,10] However, cytogenetic studies augmented by immunohistochemistry (IHC), the BSNS has been finally recognized and designated as a separate entity. So this study highlights the absolute need for IHC and cytogenetic studies in such ambiguous cases for early detection and timely therapeutic management.

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