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Case Report

A Rare Case of Biphenotypic Sino-Nasal Sarcoma - Case Report and Review of Literature

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Abstract

Biphenotypic sinonasal sarcoma is a newly described tumour of nose and paranasal sinuses containing neural and muscle tissue together in the tumour tissue. There are almost 68 different types of tumours in the Sinonasal tract. This tumour is in addition to that. A 60 year old female with history of nose block of both sides of 8 years duration, and a visible mass in the right nasal cavity, since 2 years presented to us one month back. There was no obvious features of malignancy. CT PNS showed large heterogeneous calcified mass showing enhancement in the right nasal cavity eroding the nasal septum and reaching left side pushing it. Histopathology showed a neoplasm covered partly by partly by psuedostratified columnar epithelium and partly by squamous epithelium. Neoplasm was cellular and composed of spindle cells with spindle/oval/wavy nuclei arranged in in sheets It was seen invading the adjacent bone. Histopathology was suggestive of low grade sarcoma possibly Biphenotypic sinonasal sarcoma. Immunohistochemistry showed vimentin and S100 positivity, SMA was focally positive. Betacatenin showed cytoplasmic granular positivity, Ki 67 percentage of positive nuclei 2-3%, MyoD1 Negative and the final report was Biphenotypic sinonasal sarcoma. This neoplasm contained myogenic and nervous tissue together. Endoscopic excision of the tumour was done. She was advised Craniofacial resection for this low grade malignancy as it was dangerous to remove it endoscopically due to the close proximity with the cribriform plate.

Keywords: Biphenotypic Sinonasal Sarcoma; New Entities of Sinonasal Tract Tumours

Introduction

Sinonasal tract is not an uncommon site for malignancy. Apart from various types of carcinomas, lymphoma, even though rare, sarcoma do occur in this region. Biphenotypic sinonasal sarcoma is a newly described tumor of nose containing neural and muscle tissue together. This tumour was added to the already existing 68 types of tumours of this region. There are a few other tumours also in this new tumour list [1]. Recently we came across a 65 year old female patient with nasal block of both sides nose of 8 years duration and a visible mass in the right nasal cavity of 2 years duration. After all the investigations, it was diagnosed as a case of Biphenotypic sinonasal sarcoma (BSNS).

Biphenotypic sinonasal sarcoma is described along with other new neoplasms of sinonasal tract. The other rare tumours are NUT midline carcinoma, HPV related carcinoma with adenoid cystic like features, SMARCB1 (IN - 1) deficient sinonasal carcinoma., renal cell like adeno carcinoma. (WHO classification. 4th edition). Hence an update become necessary to include these tumours also. This update of WHO classification in 2017 describe tumours of nose, PNS, and skull base [2]. Here we report a case of Biphenotypic sinonasal sarcoma (BSNS) recently we came across.

Case Report

60 year old female with history of nose block of both sides of 8 years duration, and a visible mass in the right nasal cavity, since 2 years presented to us one month back. There was no head ache, or bleeding. No features of obvious malignancy (Figure 1).

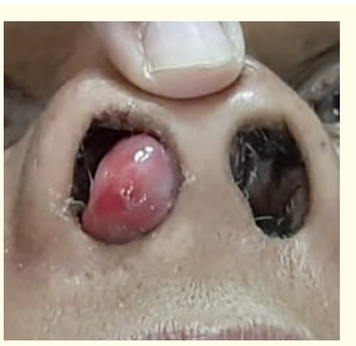

Figure 1

CT PNS - showed large calcified mass showing heterogeneous enhancement in the right nasal cavity eroding the nasal septum and reaching left side. It was seen extending to right frontal sinus, anteromedial aspect of the right orbit, right ethmoid, right Sphenoid through destruction of walls. Inward bowing of lateral wall of right maxillary sinus with sclerotic walls of the remaining region was also seen. It extended into right side of nasopharynx (Figure 2).

Histopathology findings - showed neoplasm covered partly by psuedostratified columnar epithelium and partly by squamous epithelium. Neoplasm was cellular and was composed of spindle

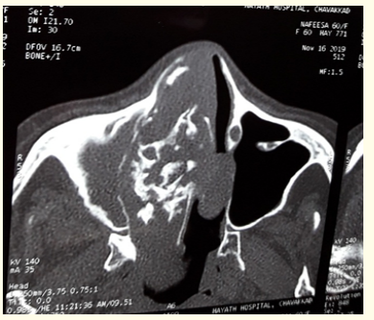

Figure 2

cells with spindle/oval/wavy nuclei arranged in sheets/bundles. Mitotic figures were sparse. Neoplasm is seen invading the adjacent bone. Surface epithelium showed marked proliferation with invagination of islands of epithelium into the tumor tissue. Many mast cells and scattered lymphocytes were present (Figure 3).

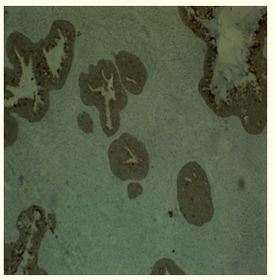

Figure 3

Immunohistochemistry and it's interpretation

Cytokeratin (AE1/AE3) Negative - Non epithelial in nature, vimentin positive - mesenchymal tissue.

S100 B positive - neurogenic tissue, SMA positive smooth muscle tissue present.

Desmin negative - not alveolar soft cell tissue sarcoma. MyoD1 - negative- not alveolar soft tissue sarcoma or rhabdomyosarcoma. Ki 67 - 2 - 3% means low grade malignancy.

Surgical findings

Reddish soft tissue mass in the anterior aspect of nose. Bleeding on firm touch from the superior aspect.

Endoscopic excision of the whole tumour was done on right side very carefully. There was not much of bleeding because of the hypotensive Anaesthesia. The mass was seen eroding the nasal septum Through the septal perforation mass was seen crossing to the other side. On the right side, it was pushing the lateral wall of nose and had destroyed the middle turbinate partially.

On the left side, it was a swelling mainly bony in nature. It also pushed the lateral wall to the side. It was dangerous to completely remove, as the cribriform plate may get damaged. Sufficient mass was removed to get adequate airway. Nose was packed and the pack was removed next day. Post operative period was uneventful.

After getting the biopsy report and understanding the nature of disease, she was advised a Craniofacial resection which will be the ideal treatment for this tumour of low grade malignancy in this patient.

Discussion and Review of Literature

Nose and paranasal sinuses, even though a small area, give rise to malignancy occasionally. Both Pathologist and ENT Surgeons who come across these tumours should be aware of the new entities for accurate diagnosis and management.

In 2012 a review article was published in American journal of surgical pathology, regarding low grade sinonasal sarcoma with neural and muscular features. This clincopathologiacal analysis of 28 cases of sinonasal tumours, was a retrospective study of specimens and case records of Mayo clinic. All cases showed characterised morphology immunophenotyoing and cytogenetic profile. On the basis of this they suggested the name low grade sinonasal sarcoma with neural and myogenic features [3].

Only 7 cases of BSNS have been so far reported as per an article published in 2017 [4]. Yuan Lib., *et al*. in that article reported a case of BSNS with diffuse infiltration and intracranial extension in a 67 year old female which was biopsied three times for a “nasal polyp” and finally undergone endoscopic surgery combined with craniofacial reaction. Chances for metastasis is less for this tumour. BSNS primarily affects adults mainly females and the superior nasal cavity and sinuses are usually involved [5]. Another case report by Hussein Al Zamel [6] says about a young female with BSNS. The tumor was arising from the right ethmoid sinus and was pushing the septum to the other side. Endoscopic excision was then done. The HPR was Angiofibroma. sin Angiofibroma being rare in women, the slides were reviewed at Harward Medical School and finally confirmed BSNS. Article by Vickie Y Jo., *et al*. [7] in 2018 reports that the total number of cases reported was less than 40. The expression of SMA, desmin, and S-100 protein is not always sufficient in distinguishing BSNS from all histologic mimics and diagnosis often requires confirmation of PAX3 gene rearrangement. This gene rearrangement can distinguish BSNS from its morphologic mimics with high sensitivity of 100%. An article published recently in November 2019, says the story of a 52 year old lady presented with 3 months history of recurrent bleeding from nose. The right side of nasal cavity showed a neoplasm inside. This was biopsied and histopathology report was spindle cell neoplasm with neural and myogenic differentiation.. Immunohistochemistry showed BSNS. She underwent complete excision and plastic reconstruction as she had alar involvement also.

The differential diagnosis includes schwannoma, malignant peripheral nerve sheath tumour (MPNST), synovial sarcoma, and

spindle cell rhabdomyosarcoma. The tumor is locally aggressive with recurrence rates up to fifty percent but no deaths from the disease and no metastasis [8]. Biphenotypic sinonasal sarcoma is more common in females [3-6,8].

Recent years have seen a growing number of publications on genetic aberrations in sinonasal cancer. Even a book published in 2020 on Molecular Pathology of Sinonasal Tumors gives a full chapter on this matter [9].

Conclusion

Immunohistochemistry and exact diagnosis goes a long way in the plan of treatment of patient with Biphenotypic sinonasal sarcoma containing neural and myogenic tissues together. Since it is low grade malignancy complete excision is curative and metastasis is rare. We should be watchful regarding this type of new entities of sinonasal tract tumours.

Conflicts of Interest

There are no conflicts of interest between the authors.

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