A Rare Case of Biphenotypic Sinonasal Carcinoma operated in our Institution

ABSTRACT:

Biphenotypic sinonasal sarcoma is a recently described malignancy showing dual differentiation with both myogenic and neural elements. Due to its histologic similarities to other sinonasal malignancies, it is a diagnostic challenge.

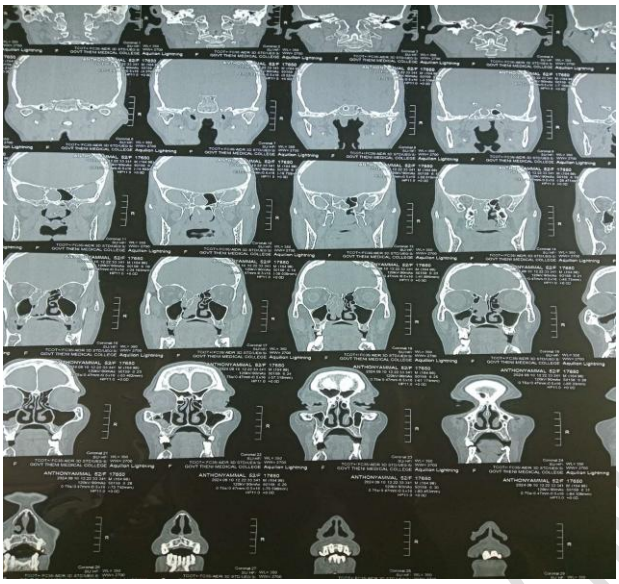
Keywords: biphenotypic sinonasal sarcoma neural and myogenic differentiation

INTRODUCTION:

Sinonasal malignancies are a diagnostic and therapeutic challenge due to the sheer histologic diversity and proximity to vital structures like the orbit, cranial nerves, and brain. Early diagnosis is often confounded by nonspecific symptoms which can be mistaken for benign disease. In addition, there exists a considerable degree of histologic overlap among distinct sinonasal malignancies, making diagnosis on biopsy challenging. One of the most recent sinonasal malignancies described in the latest who edition of head and neck tumors is biphenotypic sinonasal sarcoma (BSNS)[1]. The existence of this unique tumor was initially suspected based on earlier work[2,3] followed by a few publications detailing clinicopathological features only recently reported.[4–10] Perhaps, most characteristic of BSNS is the presence of both myogenic and neural differentiation. Clinically, the tumor is slowly progressive with a predilection for upper aerodigestive tract. However, locally aggressive spread may occur in up to half of the affected patients[4]. Most of the reported cases of BSNS have been isolated cases or small case series. Efforts are ongoing to consolidate all relevant data regarding BSNS with special emphasis on diagnostic modalities. Here, we present a case of a patient treated for BSNS and review the current literature concerning this newly identified tumor.

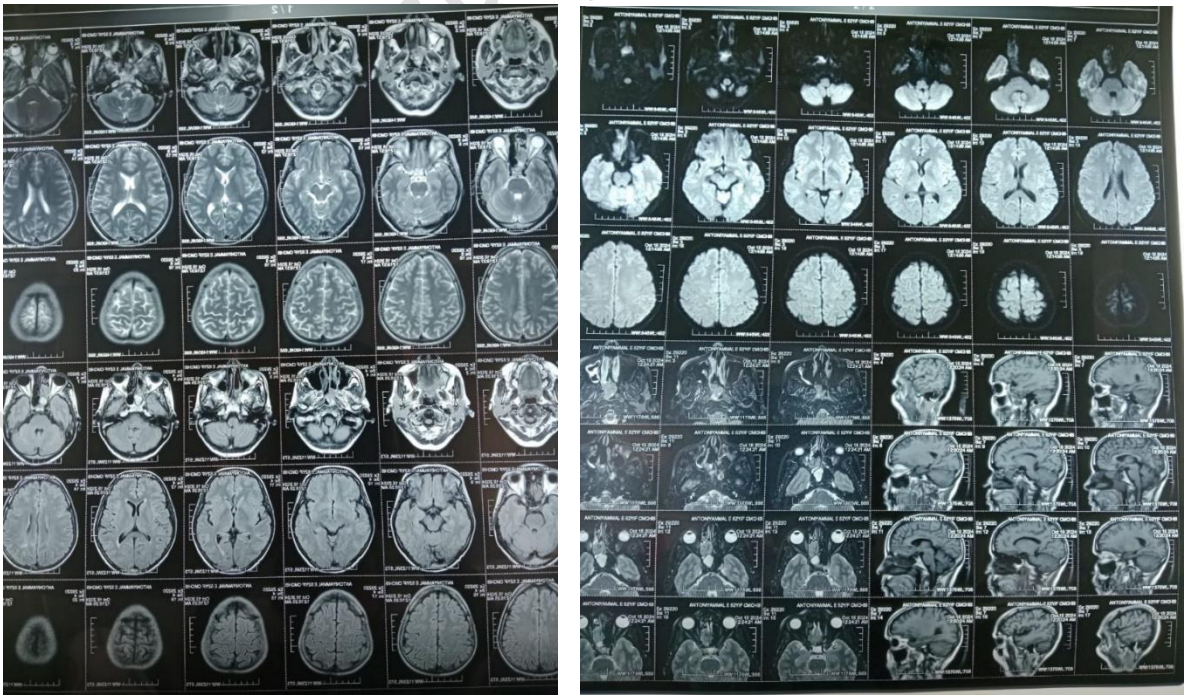
CASE:

A 52-year-old female presented with complaints of right sided nasal obstruction and anosmia for 6 months. Local examination revealed pale polypoidal mass in right middle meatus. She underwent CT scan of PNS that revealed polypoidal soft tissue density enhancing mass lesion of size 36x32x24mm seen in right nasal cavity, ethmoid sinus extending in to nasopharynx.

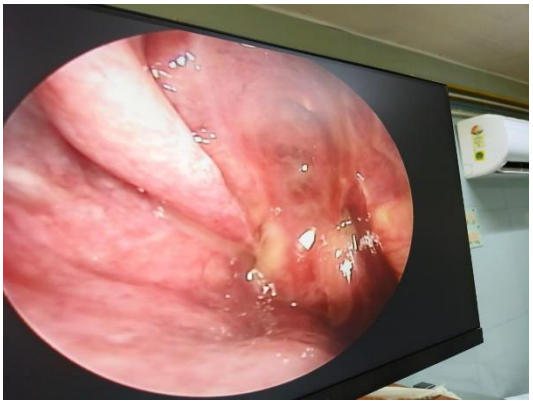


Preoperative CT Scan

Diagnostic nasal endoscopy and biopsy was taken under la and specimen was sent for HPE. HPE report came as biphenotypic sinonasal sarcoma (low grade sinonasal sarcoma with neural and myogenic differentiation). On follow up the patient presented with the complaints of right sided nasal obstruction. Examination revealed mass in right middle meatus which extends posteriorly upto skull base and posterior end of nasal septum. MRI scan revealed right mild to moderate enhancing posterior ethmoid/sphenoidal mass lesion it appears t2 intermediate to hyperintense;t1 intermediate mucocele with partial obliteration of the recess-3.9x2.1 cm.

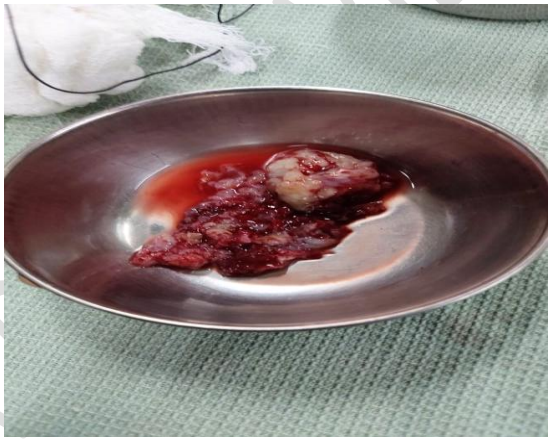


Preoperative Mri Scan

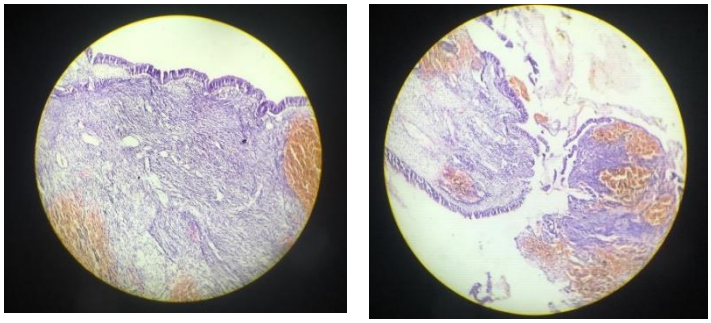


Pre operative DNE image

Endoscopic resection of tumour was planned. A friable mass was seen in right ethmoidal region which is attached to the nasal septum and the skull base was removed in toto and sent for HPE. Right total ethmoidectomy, right middle meatal antrostomy done. Right lamina papyracea was intact. Right sphenoidectomy done. Sphenoid sinus was normal. HPE report revealed biphenotypic sinonasal sarcoma. Patient was followed up till December 2024 and she appears to be disease free and has no sinonasal symptoms.



Excised Mass



HPE Microscopic Picture

DISCUSSION:

Sinonasal tract tumours are neoplasms that affect mostly the sinuses, internal nasal cavities, orbits, skull base and in some cases can have intracranial extension. Common presenting symptoms are nasal obstruction, epistaxis, facial pressure or pain, smell impairment, as well as neurological or ophthalmic complaints due to the tumour's extension. [11,12]

Biphenotypic sinonasal sarcomas were firstly discovered by lewis et al in 2012. [13]

Who announced addition of this entity in the reviewed 2017 who classifcation of head and neck tumours including BSNS as one of the newly discovered tumours of the sinonasal cavity. [14-16]

These tumours have double neural and myogenic differentiation but are histologically different from malignant sarcomas or other sinonasal cancerous masses. The primary different characteristic of this group is the biphenotypic marker expression during the immunohistochemical analysis as well as its unique identity combining clinical, morphologic, histologic and genetic features.

In all BSNS cases, imaging modalities and endoscopic investigations reveal an enhancing soft tissue mass with infltrative growth associated with hyperplastic bone or even bone infltration. It is therefore evident that minimal features exist to guide the ENT surgeon towards BSNS as these entities present similar to other nerve sheath tumours, mesenchymal neoplasms and other varieties of sarcomas[17].

Diagnosis of BSNS based on pathological features alone is not possible due to the potential for pathological overlap. Therefore, immunophenotyping is a prerequisite for diagnosis.

Immunophenotypical analysis reveals that s-100 (neural marker) and SMA (myogenic marker) are consistently positive in BSNS, while sox-10 (neural crest differentiation marker) is consistently negative[18].

Molecular studies, mainly the fish analysis, are a new addition to the list of diagnostic modalities used for BSNS.

In some cases, determination of a particular genetic aberration can confirm the diagnosis of BSNS. Pax3-maml3 fusion is a classical fusion protein found in 79 to 96% of cases[19,20].

It is therefore histological, immunochemical and genetic analysis which is required to confirm diagnosis of BSNS.

Regarding treatment modalities, all cases in the literature were treated with surgical excision either endoscopic or open using craniotomy or lateral rhinotomy as an access point with or without adjuvant radiotherapy.

Local recurrence rate is considered high but fortunately, no distant metastasis was observed in any case with BSNS in the literature.

It is therefore mindful to advocate, that radiotherapy should be individually selected in patients with spreading tumours and difficulties in complete endoscopic resections and should always be a result of multidisciplinary team discussion and involvement of patient views in the decision.

CONCLUSION:

BSNS is distinct sinonasal malignancy with dual differentiation. Its clinical behavior, pathological features, immunophenotypic presentation, standard of care, and prognostic outcomes are entirely different not only from other nonsarcomatous sinonasal malignancies but also from other head and neck sarcomas.

The clinical importance of these tumours is summarised to their common symptoms in association with the non-specific radiological findings but their high local recurrence rates that makes the early diagnosis and full treatment critical.

Treatment with radiotherapy is individualised and is supported by concrete criteria based on location of the tumour, intraoperative surgical margins, histopathological features and general condition of the patient.

It is therefore crucial for the multidisciplinary team that consists of the ENT surgeon, radiologist and primarily pathologist as well as oncologist, to be aware of this sinonasal entity to correctly diagnose BSNS, avoid misdiagnosis and treat effectively and successfully.

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