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RESEARCH ARTICLE

A RARE CASE OF BIPHENOTYPICSINONASAL CARCINOMA OPERATED IN OUR INSTITUTION

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Abstract

A newly identified cancer that exhibits dual differentiation with both myogenic and neural components is called biphenotypic sinonasal sarcoma. It is difficult to diagnose because of its histologic resemblance to other sinonasal cancers.

Introduction:-

Considerable histologic diversity of sinus cancers and their close proximity to essential organs such as cranial nerves, brain, as well as orbit make them difficult to diagnose and treat. Nonspecific symptoms that could be misdiagnosed as benign diseases often hamper early diagnosis. Furthermore, the histologic overlap between different sinonasal cancers is substantial, making biopsy diagnosis more difficult. According to most recent WHO edition of neck and head tumors, one of most recent sinonasal malignancies to be discovered is biphenotypic sinonasal sarcoma (BSNS)[1] . Distinctive tumour presence had been firstly inferred from prior research[2,3], followed by several publications that recently detailed its clinicopathological characteristics[4-10]. The defining feature of BSNS is coexistence of myogenic as well as neural differentiation. The tumor primarily affects the upper aerodigestive tract and progresses gradually. However, up to 50% of the afflicted patients may experience locally aggressive spread[4]. Most of the BSNS cases that have been reported are isolated incidents or small case series. All relevant information about BSNS is being compiled, with an emphasis on diagnostic techniques. We review existing literature on this novel discovered tumor and report individual's case who had been BSNS treated.

CASE:

A 52-year-old female Exhibited 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 expanding into nasopharynx.

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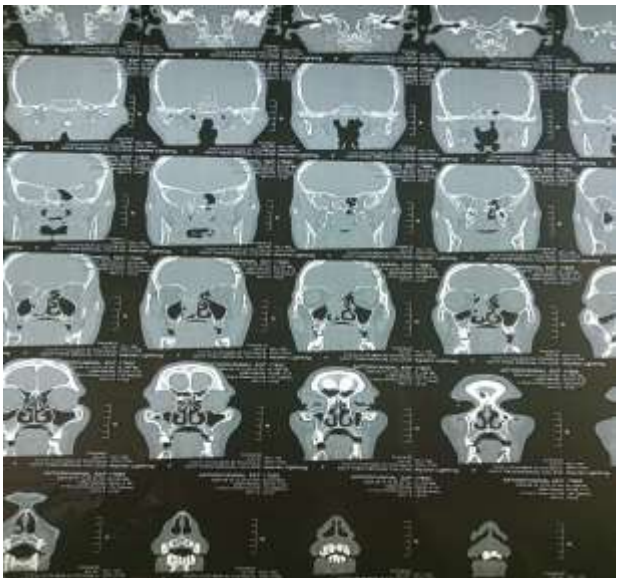
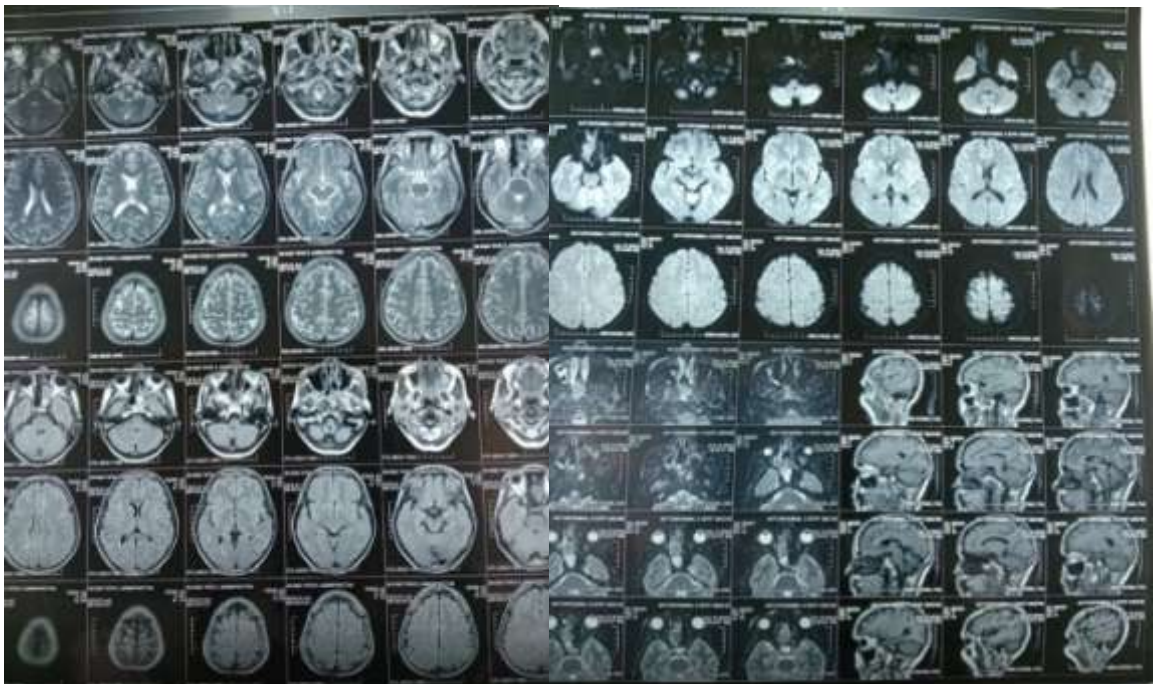
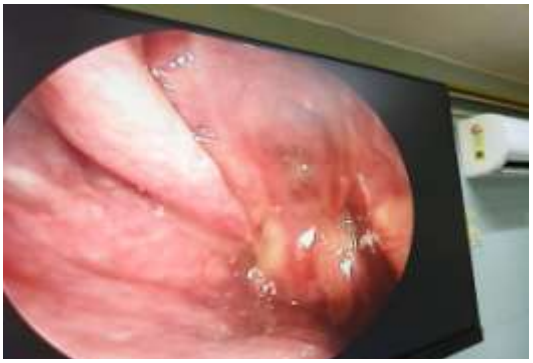


Preoperative CT Scan

Diagnostic nasal endoscopy and biopsy were taken under la and specimen was sent for HPE. According to the HPE report, the patient had “biphenotypic sinonasal sarcoma”, which is a low-grade condition which exhibits neuronal along with myogenic differentiation. On follow up the patient presented with 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.



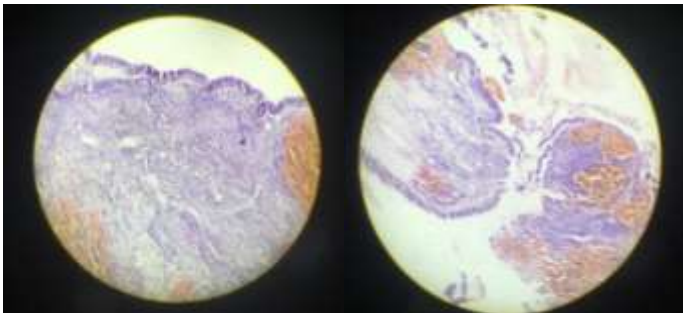
Preoperative Mri Scan



Pre operativeDNE 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 seems disease free moreover with no sinonasal symptoms.

Excised Mass



HPE Microscopic Picture

Discussion:

Neoplasms known as sinusonasal tract tumors mainly affect the sinuses, orbits, internal nasal cavities, and the base of the skull, though they can occasionally spread intracranially. Typical presenting symptoms include epistaxis, nasal obstruction, facial pain/pressure, smell impairment, alongside neurological/ophthalmic issues resulting from tumour extension. [11,12]

Biphenotypic sinonasal sarcomas had been initially discovered by Lewis et al in 2012. [13]
As one of recently identified sinonasal cavity tumors, BSNS was comprised in 2017 WHO revision of the head and neck tumor classification. [14-16]
Such tumors exhibit myogenic as well as dual neural differentiation, yet they have been histologically distinct from malignant sarcomas and other sinonasal cancerous tumours. Distinguishing features of such grp has been biphenotypic markers expression throughout immunohistochemical analysis, along with its distinctive identity that integrates morphologic, clinical, histologic, moreover genetic characteristics.
In every BSNS instance, imaging modalities alongside endoscopic examinations demonstrate increasing soft tissue mass exhibiting infiltrative growth, accompanied by hyperplastic /even bone infiltration which has been thus apparent that there are few specific features to assist ENT surgeon in identifying BSNS, because such entities resemble other nerve sheath tumors, mesenchymal neoplasms, along with various other sarcomas types[17].
BSNS diagnosis cannot be made solely through pathological characteristics because of possibility of pathological overlap. Consequently, immunophenotyping has been essential for diagnosis.
Immunophenotypic analysis demonstrates neural marker (S-100) alongside myogenic marker (SMA) have been positive consistently in BSNS, however, sox-10 (neural crest differentiation marker) has been negative consistently[18].
Molecular studies, particularly fish analysis, represent novel diagnostic modality addition employed for BSNS.
For certain instances, specific genetic aberration identification could validate BSNS diagnosis. The “Pax3-MAML3 fusion” is a classic fusion protein identified in 79 to 96% of cases[19,20].
Consequently, histological, immunochemical, and genetic analyses are necessary to confirm the diagnosis of BSNS.
All cases documented in the literature underwent surgical excision, either endoscopic or open, utilising lateral rhinotomy/craniotomy regarding access points, without/with adjunctive radiotherapy.
The local recurrence rate is deemed elevated; however, no distant metastasis has been reported in any BSNS case in literature.
It is prudent to assert radiotherapy must be tailored for individuals having spreading tumors alongside challenges in achieving complete endoscopic resections, moreover it must always stem from multidisciplinary team discussion that incorporates the patient's perspectives in decision-making process.

Conclusion:

BSNS has been unique sinonasal malignancy characterised by dual differentiation. Its clinical behaviour, pathological characteristics, immunophenotypic profile, standard treatment protocols, alongside prognostic results have been distinctly varying from both other non sarcomatous sinonasal malignancies and other neck as well as head sarcomas.
Clinical significance of such tumors has been encapsulated in their prevalent symptoms, coupled via non-particular radiological outcomes, and their elevated local recurrence rates, which render early diagnosis alongside comprehensive treatment imperative.
Radiotherapy treatment is tailored and guided by specific criteria, including tumor location, intraoperative surgical margins, histopathological characteristics, and the patient's general wellness status.
It is essential for the multidisciplinary team, comprising the ENT surgeon, radiologist, pathologist, and oncologist, to recognize such sinonasal entity for accurately diagnosing BSNS, prevent misdiagnosis, moreover ensure effective treatment.

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