Biphenotypic Sinonasal Sarcoma with Intracranial Extension – A Case Report with Review of Literature

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Abstract

Rationale: Biphenotypic sinonasal sarcoma is a rare malignant tumour exclusively involving the sinonasal cavity. These tumours have variable and atypical presentations. Early approach and correct treatment modalities are key factors in the management of such cases. **Patient Concern:** A 48-year-old male patient presented with left-sided nasal obstruction and intermittent nasal bleeding for one year. **Diagnosis:** Biphenotypic sinonasal sarcoma confirmed on histopathological examination and immunohistochemistry. **Treatment:** The patient underwent surgical excision with left lateral rhinotomy and bifrontal craniotomy with skull base repair. The patient also received postoperative radiotherapy. **Outcome:** The patient is on regular follow-up with no similar complaints. **Take-Away Lesson:** Treating team should keep the diagnosis of biphenotypic sinonasal sarcoma in mind while investigating a patient with nasal mass. Surgical management is the treatment of choice, due to its local aggressive nature and proximity to the brain and eyes. Postoperative radiotherapy is vital to prevent tumour recurrence.

Keywords: Biphenotypic, intracranial, sarcoma, sinonasal, soft tissue

Introduction

Biphenotypic sinonasal sarcoma is a rare malignant tumour. It has recently been added in the fourth edition of the World Health Organization classification of head-and-neck tumours.[1] It was first described by Lewis *et al*. as a rare low-grade sarcoma in 2012.[2] This tumour expresses dual markers for neural and myogenic differentiation, which led to its reclassification as a separate entity. Tumour occurs exclusively in the nasal cavity with ethmoid sinus being the most commonly affected paranasal sinus. Other sites of involvement include orbit, cribriform plate, brain, and oropharynx. Patients present with nonspecific symptoms such as nasal obstruction, congestion, and sinonasal pain. Histologically, biphenotypic sinonasal sarcoma can closely resemble other tumours such as monophasic synovial fibrosarcoma, solitary fibrous tumour, and peripheral nerve sheath tumour. However, it shows characteristic dual staining pattern differentiation for both neurogenic and myogenic markers and consists of highly cellular spindle cells arranged in irregular fashion with immunophenotyping showing S100 and smooth muscle actin (SMA) positive markers. This tumour is locally destructive without distant metastasis. We hereby report a case of biphenotypic sinonasal sarcoma with intracranial extension with only six cases reported in the literature [Table 1].

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Case Report

A 48-year-old male presented to us with left-sided nasal obstruction, hyposmia, and intermittent nasal bleeding for one year. There was no history of headache, vision loss, diplopia, or seizures in the past. The patient also revealed a history of excision of left nasal mass three years ago which recurred nine months postoperatively. On physical examination, there was mild proptosis in the left eye [Figure 1a] and anterior rhinoscopy showed a pinkish polypoidal mass occupying the entire left nasal cavity [Figure 1b]. The patient was further investigated with complete blood tests and viral markers. Contrast-enhanced magnetic resonance imaging of the brain, paranasal sinuses, and orbit was done showing large irregular T1-hypointense and T2-hyperintense homogeneously enhancing mass lesions involving the left nasal cavity,

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ethmoid, and frontal sinuses [Figure 2a]. A breach was also seen in the inner table in the frontal region with extension of soft tissue intracranially with mass effect on the frontal lobe on the left side [Figure 2b]. No distant metastatic deposits were seen in imaging studies. Diagnostic nasal endoscopy and biopsy resulted in profuse bleeding and required anterior nasal packing. Biopsy was suggestive of spindle cell lesion favouring malignancy. The patient was planned for surgical excision with neurosurgical team. The patient underwent a combined surgical approach through left lateral rhinotomy with bifrontal craniotomy and skull base repair [Figure 3]. A pericranial flap was raised with bicoronal incision. After craniotomy, moderately vascular friable tumour was removed from the frontal area and a small intradural component was also removed. After placing lateral rhinotomy incision, tumour was removed from the left nasal cavity and ethmoid sinus. The dural breach and skull base were repaired using abdominal fat, tissue glue, and pedicled pericranial flap. On histopathological examination (HPE), tumour cells showed extensive proliferation of spindle cells arranged in a herringbone pattern with loose collagen matrix and high nucleus-to-cytoplasm ratio. On immunohistochemistry (IHC), tumour cells were found to be positive for vimentin, SMA, and S-100 and showed negative for tumour markers glial fibrillary acidic protein, progesterone receptor, beta-catenin, calponin, and CD34 [Figure 4] favouring the diagnosis of biphenotypic sinonasal sarcoma. The patient was later referred for radiotherapy. Postoperatively, the patient had no complaints of diplopia or ophthalmoplegia; however, the patient had hyposmia and a small depression over the frontal region postsurgery. On follow-up period for 6 months, radiological investigations and endoscopy showed no recurrence [Figure 5] and the patient is on regular follow-up till now.

Discussion

Biphenotypic sinonasal sarcoma is a recently described rare malignant mesenchymal tumour that occurs in the nasal cavity and paranasal sinuses. The diagnosis of biphenotypic sinonasal sarcoma is based on focal arrangements of spindle cells in loose collagen matrix, which shows positive IHC with smooth muscle actin (SMA) and S-100 markers of myogenic and neuronal differentiation, respectively. Due to the low mitotic rate of spindle cells in the tumour, they were earlier described as low-grade sinonasal sarcoma. Biphenotypic sinonasal sarcoma affects adults with a male-to-female ratio of 1:2 with a mean age of 49–51 years.[7-9] Our case was a male with a mean age of 48 years without any comorbidities. Clinical features are variable and most commonly include symptoms such as nasal obstruction, nasal bleeding, and nasal discharge. Lewis *et al*., in their retrospective study, on 28 cases of biphenotypic sinonasal sarcoma also reported

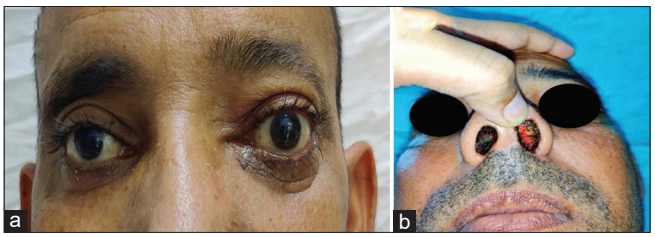

Figure 1: (a) Patient with left eye proptosis. (b) Anterior rhinoscopy – left nasal cavity mass

Table 1: Comparison of cases with intracranial extension and their treatment modalities

	Total cases	Number of cases with intracranial extension	Median age (years)	Clinical features	Sites involved	Management modalities
Lewis *et al*.[2]	28	1	48	Difficulty in breathing through the nose, facial pressure, and congestion. Facial pain and mild epiphora	Nasal cavity 15/28, ethmoid sinus 16/28, orbital involvement 7/28, cribriform plate 3/28, and cranial vault 1/28	Surgical excision, and craniofacial resection with/ without radiation
Cannon *et al*.[3]	3	2	67	Diplopia, facial discomfort, supraorbital mass, and nasal obstruction	Nasal cavity 2/3, frontal 3/3 and ethmoid sinuses 3/3, lamina papyracea 1/3, skull base 2/3	Purely endoscopic resection in 1st case, craniofacial resection of the anterior skull base in combination with neurosurgery in 2nd case, 3rd case could not be operated
Hockstein *et al*.[4]	1	1	79	Midline frontal mass	Left frontal, ethmoid and maxillary sinus, erosion of anterior and posterior cortex of frontal sinus	Combined endoscopic removal with bifrontal craniotomy
Lin *et al*.[5]	1	1	67	Nasal obstruction and right nasal cavity mass for 10 years	Right nasal cavity, nasopharyngeal cavity, right maxillary sinus, right frontal sinus, bilateral ethmoid sinuses, bilateral sphenoid sinuses, frontal lobe	Endoscopic surgery combined with craniofacial resection
Rooper *et al*.[6]	12	1 with recurrence	44	Not mentioned	Ethmoid sinus 4/12, frontal sinus 3/12, nasal cavity 3/12, both nasal cavity and ethmoid sinus 1/12	2/12 orbital exenteration, surgical excision

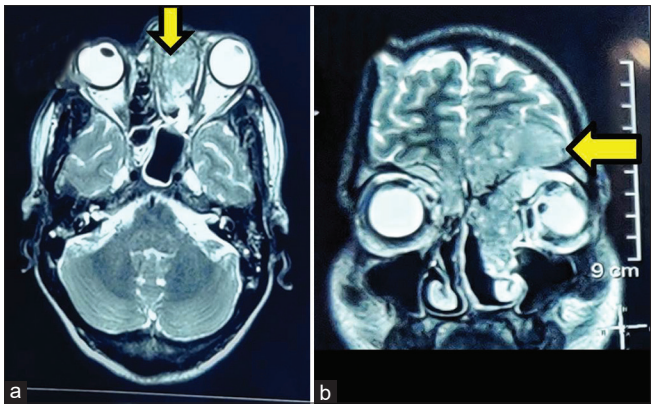

Figure 2: MRI PNS-orbit: (a) left nasal cavity mass, (b) intracranial extension. MRI = Magnetic resonance imaging, PNS = Paranasal sinus. Yellow arrows highlight and represents the tumour mass

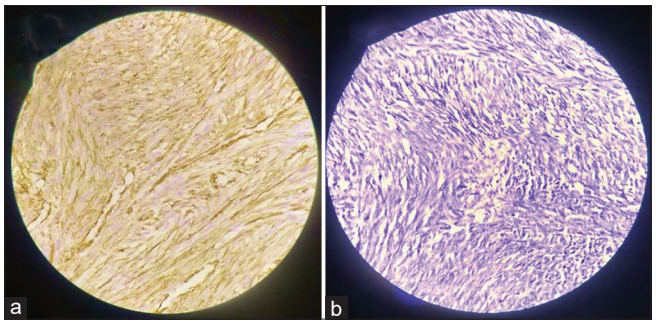

Figure 4: (a) IHC-positive for SMA, (b) HPE – showing spindle cells. IHC = Immunohistochemistry, HPE = Histopathological examination, SMA = Smooth Muscle Actin

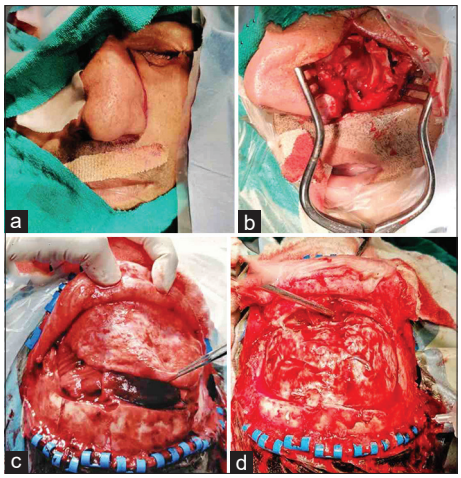

Figure 3: (a) Incision mark, (b) nasal cavity mass, (c) raised pericranial flap, (d) tumour in the frontal sinus

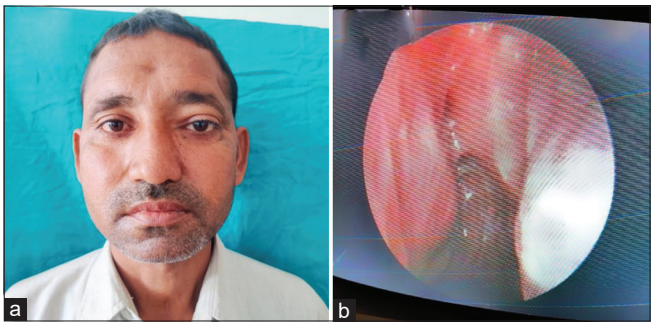

Figure 5: Postoperative image of (a) the patient and (b) endoscopy of the left nasal cavity

presenting symptoms as typically sinonasal, such as nasal congestion and facial pain. Less frequent presenting symptoms of biphenotypic sinonasal sarcoma include diplopia and blurred vision. Cannon *et al*. reported diplopia as presenting symptom of biphenotypic sinonasal sarcoma in one of their cases.[3] However, in some rare instances, these tumours can also present with atypical symptoms of midline frontal mass.[4] Biphenotypic sinonasal sarcoma usually involves multiple sites in the sinonasal region. Nose and paranasal sinuses involvement are common with ethmoid sinus being the most common, followed by frontal sinus and sphenoid sinus is less frequently involved. The brain is the least affected site with only a few cases reported in the past. Lin *et al*. reported a case of biphenotypic sinonasal sarcoma extending into the frontal lobe.[5] Cranial vault involvement was reported in one of the cases of biphenotypic sinonasal sarcoma by Lewis *et al*. Our case had the left nasal cavity involved with bilateral ethmoid and frontal sinus extending to the left frontal lobe. Lin *et al*. reported a case of biphenotypic sinonasal sarcoma with the involvement of bilateral sphenoid sinuses. Rooper *et al*. in their case report with 12 patients of biphenotypic sinonasal sarcoma, reported ethmoid sinuses being more commonly involved (33.33%) and one case had intracranial involvement due to recurrence.[6]

On HPE, there are cellular areas of spindle cells arranged in fascicles with prominent vasculature. Infiltrative biphenotypic sinonasal sarcoma found in ethmoids histologically resembles and mimics inverted papilloma of the nasal cavity due to the presence of foci of invaginated respiratory epithelium.[10] This close resemblance can lead to wrong diagnosis and treatment, as occurred in our case and one reported by Lin *et al*. Lewis *et al*. also reported four cases out of 28 who had operated for benign sinonasal mass in the past. Postoperative radiotherapy is often omitted due to the wrong diagnosis, resulting in recurrence with extensive complications. Biphenotypic sinonasal sarcoma on IHC showed bimodal pattern having neurogenic and myogenic differentiation. Diagnosis of these tumours is more consistent with strong positivity for both S100 and smooth muscle markers – smooth muscle actin (SMA). Since biphenotypic sinonasal sarcoma occurs in proximity with vital structures such as the brain, its locally aggressive nature can lead to the involvement of the skull base, as seen in two out of three cases reported by Cannon *et al*. Biphenotypic sinonasal sarcoma with intracranial involvement is an entity rarely described in the literature such that only six cases were found after thorough review. Hence, the treatment modalities can be challenging to define. Since tumour is

locally aggressive with no distant metastasis, early surgical intervention is required. Surgical excision with craniofacial resection has been adopted as the primary treatment in biphenotypic sinonasal sarcoma with intracranial extension in the past. As our case had involvement of the left nasal cavity with bilateral ethmoids and frontal sinuses, extending up to the left frontal lobe, the patient underwent left lateral rhinotomy with bifrontal craniotomy in a combined approach with the neurosurgeon team. Since it was a highly vascular tumour, the open approach improved visibility and provided better bleeding control. A few limitations of this approach were the external scar, delayed healing, and prolonged hospital stay. A similar approach was also used in one case reported by Hockstein *et al*. To prevent a recurrence, postoperative radiotherapy should be considered.

Conclusion

Biphenotypic sinonasal sarcoma of sinonasal origin is a relatively rare entity and its diagnosis is challenging due to atypical and variable symptoms. Due to its highly aggressive nature and location in proximity to the eyes and brain, surgical management is the treatment of choice. A higher recurrence rate is associated with biphenotypic sinonasal sarcoma; hence, postoperative radiotherapy should be included in the treatment modality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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