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Case Report

Monophasic synovial sarcoma of the ethmoid sinus

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ABSTRACT

Sino-nasal synovial sarcomas are rare mesenchymal tumours, most arising primarily from the paranasal sinuses. Very few have been described with an extension to the nasal cavity. Due to a range of non-specific symptoms and local aggressiveness, diagnosis tends to be confusing. Here we describe a case of a primary sino-nasal monophasic synovial sarcoma, diagnosed by histopathology and treated by excision and adjuvant radiation therapy. On follow-up, the patient had minimal residual disease and was further offered revision surgery and chemotherapy. She declined surgery and received chemotherapy, and remains symptom-free to date. A wide range of diagnoses should be considered for nasal masses since all tissue types coexist in this region.

Keywords: Synovial sarcoma, Nasal cavity, Sarcomas, Endoscopic excision

INTRODUCTION

The nasal cavity and the paranasal sinus system have extreme heterogeneity in tissue structure. A wide variety of pathologies can arise owing to the multiple tissue types comprising the anatomy of this region.[1] Synovial sarcomas (SS) are known to comprise 5-10% of all soft tissue sarcomas. The head and neck regions are the second most common site of involvement.[2] SS of the paranasal sinuses is extremely rare. Here we present a case report of a monophasic synovial sarcoma of the ethmoids, initially treated with surgical excision, followed by adjuvant chemoradiotherapy.

CASE REPORT

A 63-year-old female presented with a complaint of left nasal obstruction for three months. It was associated with left purulent nasal discharge for two weeks. There was no history of breathing difficulty, visual complaints, pain, cheek numbness, blood-stained nasal discharge, or loosening of teeth. On examination, there was no apparent distortion of the external nasal framework. Facial symmetry was maintained. On anterior rhinoscopy, a red polypoidal mass was visualized, occupying the entire left nasal cavity, pushing the nasal septum to the right, and reaching the floor of the cavity. On probing, the mass did not bleed and appeared to be attached to the lateral wall of the nasal cavity. There was no extension to the nasopharynx. All cranial nerves seemed to be intact. There was no palpable cervical lymphadenopathy. Visual, dental, and systemic examinations were unremarkable.

Initial blood workup was normal, and a contrast-enhanced computed tomography scan (CECT) of the nose, paranasal sinuses, orbit, and neck depicted a well-defined 3×3×2 cm enhancing soft tissue density in the left nasal cavity and the anterior ethmoids, extending to the posterior ethmoids. The growth was abutting the medial wall of the left maxillary sinus and the orbit with no apparent bone erosion (Figures 1). A contrast-enhanced magnetic resonance imaging (MRI) showed a T2 intermediate, irregularly enhancing lesion in the left ethmoid sinuses with no intracranial extension (Figure 2). An endoscopy revealed the mass attached to the middle turbinate, with dilated vessels on the surface (Figure 3). An endoscopic biopsy

was undertaken, which showed spindle cell morphology with nuclear atypia, possibly a sarcomatous lesion.

Due to the limited nature of the disease, a primary surgery, followed by adjuvant chemoradiotherapy, was planned. The patient underwent endoscopic excision under general anaesthesia. The red fleshy mass was visualized arising from the ethmoids occupying the middle meatus on the left. Middle meatal antrostomy and anterior ethmoidectomy were done, sparing as much of the nasal mucosa as possible. Adequate haemostasis was achieved. The mass was removed and sent for histopathology. Routine postoperative care was administered. The histopathology showed spindle-shaped tumour cells arranged in fascicles with nuclear atypia and hyperchromatism (Figure 4). Immunohistochemistry was positive for nuclear TLE-1 and bcl-2 (Figure 5) and was negative for EMA and CD34 (Figures 6), ruling out melanoma and epithelial tumours. A diagnosis of monophasic synovial sarcoma was finalized. She received intensity-modulated radiotherapy of 66Gy in 33 fractions to the nasal cavity and the surrounding tissues, along with palliative two-agent chemotherapy with Doxorubicin and Ifosfamide throughout her treatment. She is symptom-free on a follow-up of two years.

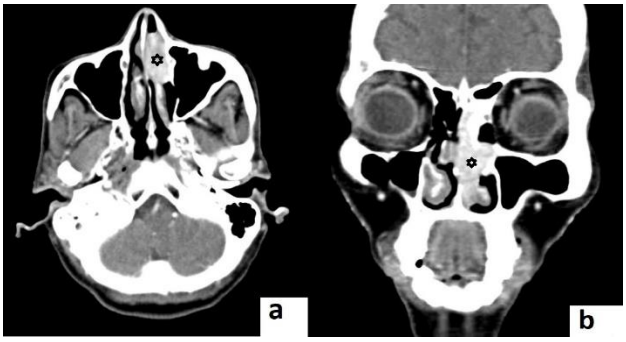

Figure 1: Pre-operative CECT (a) axial section; and (b) coronal section, depicting the heterogeneously enhancing mass within the nasal cavity and the ethmoid sinuses. The black star denotes the mass.

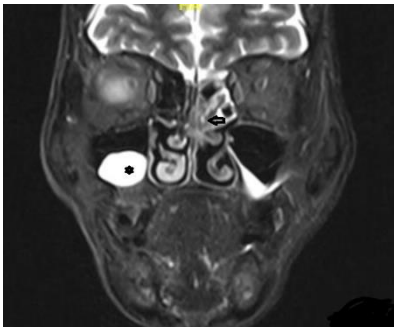

Figure 2: Pre-operative MRI T2 weighted image in coronal view depicting the tumour as an intense intermediate mass. The black arrow shows the mass. The black star represents an incidentally detected mucus retention cyst in the right maxillary sinus.

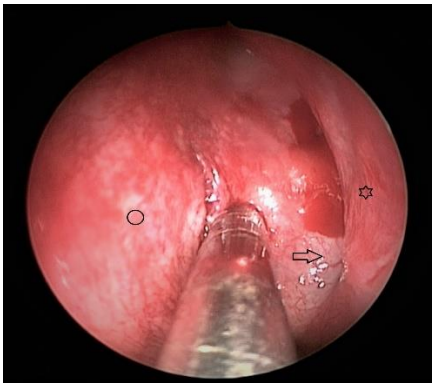

Figure 3: Pre-operative diagnostic nasal endoscopy image depicting the suction tip in contact with the nasal mass. Dilated veins over the mass surface are marked with the black arrow. The black star denotes the middle turbinate. The black circle marks the nasal septum medially.

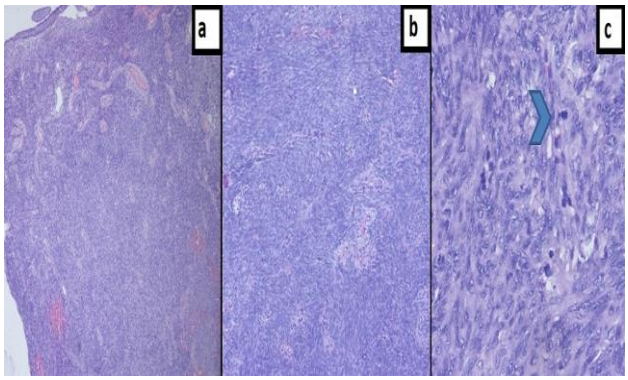

Figure 4: Photomicrographs stained with hematoxylin and eosin (a) depicts a highly cellular tumour with an overlying stratified epithelium (40X magnification); (b) shows tumour cells arranged in fascicles and sheets (100X magnification). The blue arrowhead in (c) indicates plump spindle-shaped cells with hyperchromatic nuclei (400X magnification).

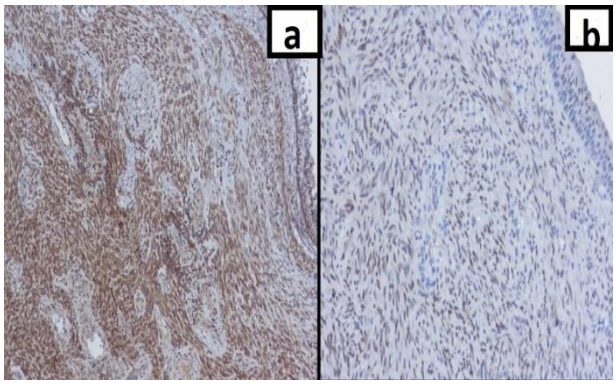

Figure 5: Photomicrographs of immunohistochemistry stained with diaminobenzidine (DAB) (a) depicts strong positivity with bcl-2 (DAB 100X magnification); and (b) shows strong nuclear positivity with TLE-1 (DAB 200X magnification).

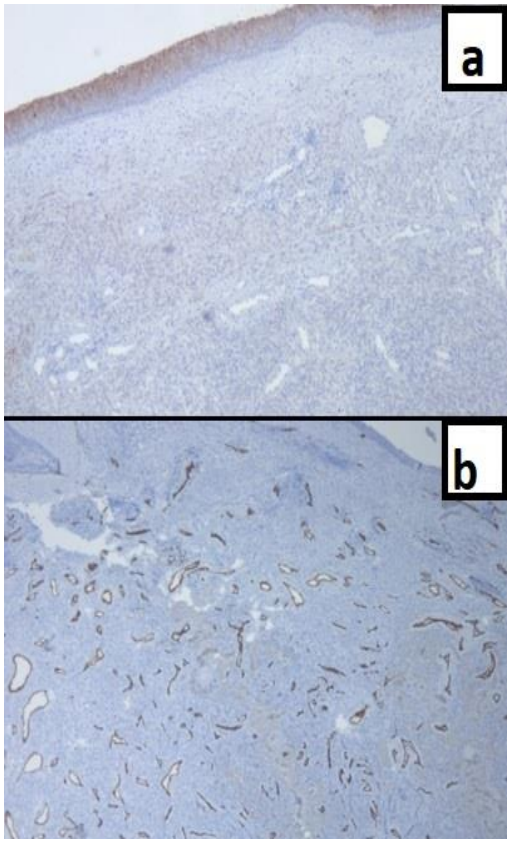

Figure 6: Photomicrographs of immunohistochemistry stained with DAB (a) shows tumour cells negative for EMA (DAB 100X magnification); and (b) depicts tumour cells staining negative for CD 34 (DAB 100X magnification).

DISCUSSION

SS of the nose and paranasal sinuses are misnomers because they are undifferentiated carcinosarcomas arising from mesenchymal tissues with no synovial membrane or joints.[3] They are named only due to the morphologic similarity with normal synovial membranes because of epithelial and stromal cells. These are considered high-grade malignant tumours. 5-12% of all SS have been reported in the head and neck region.[4] Isolated reports of sino-nasal SS exist. Comprising less than 0.1% of all soft tissue malignancies, SS of the nose and paranasal sinuses are rare.[5]

A wide range of non-specific clinical features is associated with this tumour. Nasal obstruction, pain, and epistaxis are similar to any other malignancy from the nasal cavity. Locally aggressive spread results in epistaxis, orbital invasion, and skull base erosion. A few published reports mention a gradually enlarging mass of the sinuses, which caused erosive effects on the surrounding bone. This causes a diagnostic dilemma due to similar presentations in primary epithelial malignancies. Synovial sarcomas present two types of tissues histologically: spindle cells and epithelial components. Monophasic fibrous (MFSS) is one of four subtypes of SS, the others being monophasic epithelial (MESS), biphasic, and poorly differentiated.[6] The classification is based on the relative proportions of the two tissues in the tumour. Immunohistochemistry of the excised mass plays a vital role in diagnosis. However, no specific markers exist for confirmation-cytogenetic analysis and molecular diagnosis help to differentiate the rarer monophasic fibrous type from other sarcomas. Synovial sarcomas are usually positive for epithelial membrane antigen (EMA), S-100, vimentin, cytokeratin, and bcl-2, showing a mixed immunoreactivity.

The histopathology depicted spindle-shaped tumour cells arranged in tight fascicles with minimal intervening stroma. Nuclear atypia with hyperchromatism and scanty cytoplasm suggests the fibrous type rather than the epithelial type. It is usually non-reactive to CD34 and CD31.[6] Synovial sarcomas, in general, arises due to the balanced chromosomal translocation t(X;18), producing SYT-SSX, responsible for the malignant proliferation. Diagnosis of this chimeric gene by fluorescent in-situ hybridization (FISH) or reverse transcriptase-polymerase chain reaction (RT-PCR) is the gold standard for diagnosing SS of any subsite.[7] For poorly differentiated SS, this helps confirm the diagnosis and routine histopathology. FISH/RT-PCR was not performed in our case.

Transducin-like-enhancer of split-1 (TLE-1) has also been identified as a consistent and reliable marker of synovial sarcomas. TLE-1 precisely differentiates SS from other spindle cell tumours, such as malignant peripheral nerve sheath tumours (MPNST), which can also arise in the region of the nose and sinuses.[7] Ultrastructural electron microscopic studies contribute to the diagnosis, depicting tightly bound spindle cells in a hypocellular stroma surrounded by collagen fibrils.[8] Strong nuclear positivity was obtained after epithelial and mesenchymal antigens were suggestive of SS in our case.

A prudent management plan usually consists of wide local excision of the mass with an adequate margin. This often proves difficult for primary tumours arising within the nasal cavity as the benefit of wide surgical clearance has to be weighed against functional and peri-operative complications. An endoscopic approach is preferred for oncological safety. Limited tumours can be resected with the addition of postoperative radiotherapy.[9]

Definitive proof of its efficacy in reducing recurrence or distant metastases is yet to be obtained. A more extended follow-up period is recommended for getting concrete evidence of the effectiveness of definitive therapy. Both neoadjuvant and adjuvant chemotherapy has shown benefit, causing a clinically significant reduction in locally aggressive tumours.[4] The approach of primary surgery was adopted in our case, keeping in mind the limited extent of the disease, with no breach of orbital or maxillary compartments. A conservative approach towards surgery can be considered in such cases, although further studies are warranted.

CONCLUSION

Sarcomas are challenging to treat, and defining operative margins within the nasal cavity is challenging. The case is significant as sino-nasal SS has rarely been described in the sixth decade, with most patients seen in the second to third decade. There is a need for further reporting and research of such rare tumours to formulate a definitive plan for a cure. There exists only empirical evidence of adjuvant chemoradiotherapy. Surgery could be considered the primary modality for treatment. Difficulty in obtaining margins for oncological safety could cause residual disease. Molecular cytogenetic analysis and immunostaining are the cornerstones for diagnosing such rare tumours. The generation of unfixed tissue samples should be prioritized during the surgical excision of suspected sarcomatous nasal masses.

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