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**Case Report**

# Navigating nasal swelling: the diagnostic journey to osteoblastic osteosarcoma

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## ABSTRACT

Nasal swelling is frequently encountered in ENT, but diagnosing osteosarcoma in the head and neck region is quite uncommon. Diagnosing and managing craniofacial osteosarcoma is challenging due to its rarity and nonspecific radiological features, which often lead to biopsy errors. Additionally, the proximity to vital structures complicates achieving complete resections. MRI is the gold standard for detailed assessment of osteosarcoma, offering crucial insights into tumor extent, soft tissue involvement, and proximity to surrounding structures for effective treatment planning and response monitoring. CT supports this by aiding in biopsy planning, staging, and detecting metastases, particularly in the chest. Craniofacial osteosarcoma treatment lacks consensus but generally prioritizes surgery, with negative margins crucial for better outcomes. Adjuvant chemotherapy can boost survival, while radiotherapy is used when complete surgery is not possible. In this case, we bring your attention to a 31-year-old gentleman who presented with progressive and painless nasal swelling for 6 months prior to visiting the ORL clinic, where a final diagnosis of osteoblastic osteosarcoma was made after history, examination, imaging, and histopathology results.

**Keywords:** Nasal swelling, Osteoblastic osteosarcoma, Craniofacial osteosarcoma, Preoperative planning

## INTRODUCTION

Osteosarcoma, also called osteogenic sarcoma, is a type of bone malignancy originating from primitive bone-forming (osteoid-producing) mesenchymal cells. It commonly occurs in the long bones of the extremities, such as the femur, tibia, and humerus.[1] The head and neck is a rare site for osteosarcoma, accounting for less than 10% of all osteosarcoma cases and less than 1% of all head and neck malignancies.[1,2] Due to the rarity of the disease, conducting rigorous studies to establish effective treatment is challenging. We report an intriguing case of a 31-year-old man with an aggressive high-grade sinonasal osteosarcoma of the osteoblastic subtype.

## CASE REPORT

A 31-year-old Indian gentleman presented with progressive and painless nasal swelling for 6 months prior to visiting the ORL clinic. Aside from worsening nasal blockage, hyposmia, and intermittent epistaxis, there were no other symptoms. Clinical examinations revealed a widened nasal bridge with a pinkish mass occupying the right nasal cavity, pushing the septum to the left and sparing the left inferior turbinate (Figure 1). On palpation, the mass had a smooth surface, was non-tender, and had a bony hard consistency. There were no cervical lymph nodes, and cranial nerves were intact. We were unable to pass a rigid nasal endoscope to further assess the nasal cavity.

**Figure 1: The clinical examination revealed a widened nasal bridge externally and a pinkish mass fully occupied the bilateral nasal cavity (right more than left).**

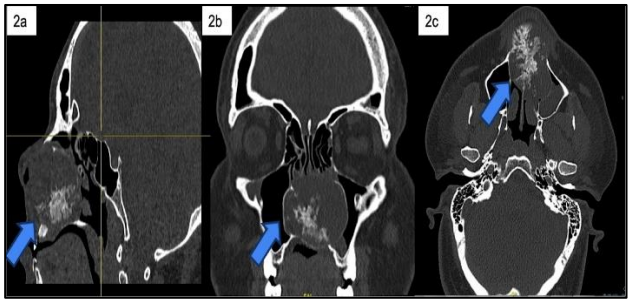


**Figure 2: CT BOS and PNS reported aggressive soft tissue mass with ring and arc calcification within anterior bilateral nasal cavity, causing erosion of the ethmoidal air cells, hard alate and anterior nasal septum. (a) Sagittal view (b) coronal view (c) axial view.**

We proceeded with an urgent CT of the base of the skull (BOS) and paranasal sinuses (PNS), which reported an aggressive soft tissue mass with ring and arc calcification within the anterior bilateral nasal cavity, causing erosion of the ethmoidal air cells, hard palate, and anterior nasal septum (Figure 2). A biopsy under general anesthesia was performed. The overall histological features favored a malignant neoplasm with predominant cartilaginous elements. However, strong clinical and radiological correlation is needed to support the diagnosis due to the presence of multiple overlapping features among a few of the close differential diagnoses of this tumor and the peculiarly low number of mitotic figures compared to the degree of nuclear atypia (Figure 3).

Thus, after thorough discussions with a multidisciplinary team, an MRI of the PNS was arranged to assess the presence of cystic and solid components. The MRI of the PNS reported a solid anterior-mid nasal mass demonstrating an iso-intense signal to skeletal muscle on T1W, heterogeneous hyperintense signal on T2W, and heterogeneous enhancement in post-gadolinium sequences (Figure 4). Based on history, examination, imaging, and histopathology, a diagnosis of chondroblastic osteosarcoma was made. Staging CT of the neck, thorax, abdomen, and pelvis showed no distant metastases.

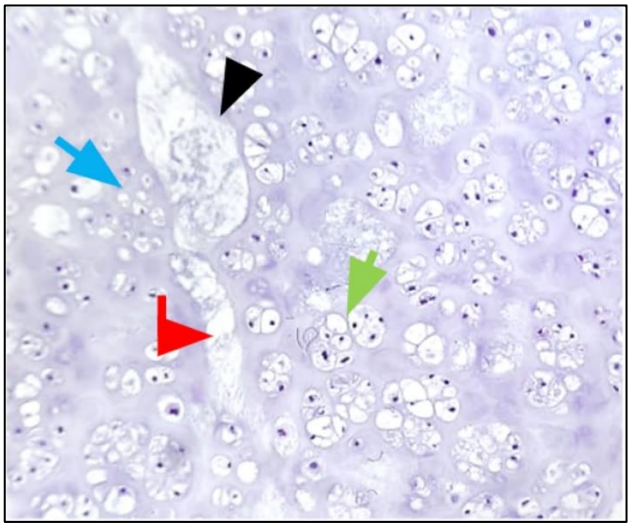

**Figure 3: The overall histological features favor malignant neoplasm with predominant cartilaginous elements. The chondrocytes are hypercellular (green arrow), with nuclear enlargement and binucleation (red arrow), associated with infarction (blue arrow) and myxoid change (black arrow), H&E, 20X.**

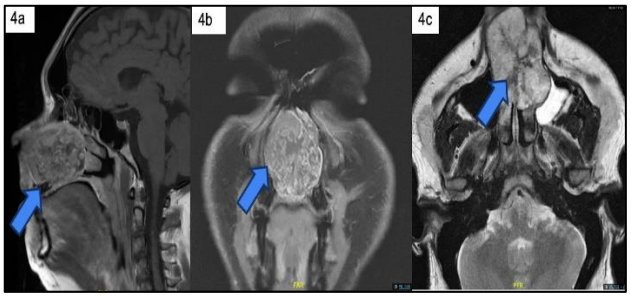


**Figure 4: MRI PNS showed the solid anterior-mid nasal mass demonstrating iso-intense signal to skeletal muscle on T1W (4a, 4b), and heterogeneous hyperintense signal on T2W (4c). The sunburst periosteal reaction at the centre of this mass appears hypointense on all sequences. a: sagittal view, T1W; 4b: coronal view, T1W; c: axial view, T2W.**

The patient was referred to a tertiary center and underwent partial rhinectomy and bilateral partial maxillectomy via a midfacial degloving approach. The tumor was excised en bloc, including the tumor, septum, nasal cartilages, hard palate (teeth 15 to 24), and the anterior part of the bilateral medial wall of the maxillary sinuses (Figure 5). Final histological analysis confirmed the diagnosis of osteoblastic osteosarcoma, histology Grade 2, moderately differentiated, high grade with AJCC staging T1. He is currently doing well post-

operation and is undergoing ongoing adjuvant chemotherapy.

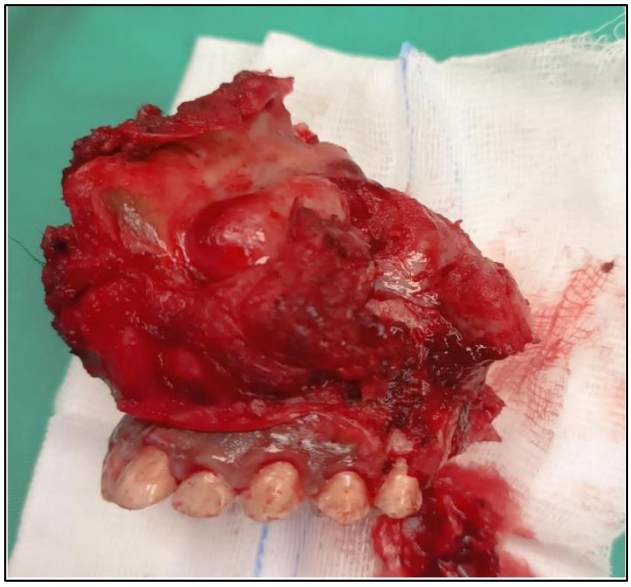

**Figure 5: Specimen removed en-bloc: tumour+ septum+nasal cartilages+hard plate (teeth 15 to 24) +anterior part of bilateral medial wall of maxillary sinuses.**

## DISCUSSION

Osteosarcoma has a bimodal age distribution with the first peak typically occurring in adolescents between 10 and 14 years old, coinciding with periods of rapid skeletal growth during puberty. The second peak is seen in older adults, particularly those over 65, where osteosarcoma may develop as a secondary malignancy.

In older adults, this second peak is often associated with pre-existing conditions such as Paget's disease of bone. The distinct age distribution reflects different underlying factors and mechanisms in these two age groups 3. Osteosarcoma incidence rates are highest among Black individuals, with the average ratio of males to females being 1.22:1.[4,5]

Head and neck osteosarcoma is a rare type of sarcoma, typically developing in the mandible or maxilla in the third and fourth decades of life.[6] The average age of sinonasal osteosarcoma is in the 30s, which is 10 years older compared to those with osteosarcoma in the long bones.[4,7] In sinonasal osteosarcoma, common paranasal effects are the maxillary sinus, ethmoid sinus, nasal cavity, sphenoid, and frontal sinus, with 63.8%, 52%, 46%, 28%, and 20%, respectively. The most common presenting symptoms were nasal obstruction (57%), epistaxis, V2 paresthesia, facial swelling, and facial pain (29%).[4]

Diagnosing and managing osteosarcoma becomes challenging due to the high incidence of errors in biopsy results secondary to its rarity and unspecific radiological features. The proximity of vital structures makes it difficult to achieve appropriate resections.[8]

Sinonasal osteosarcoma often lacks the "sunburst" pattern seen in long bone cases due to localized subperiosteal reactions causing calvarial thickening. MRI findings of enhancement with contrast and iso-intensity on T1-weighted images, as seen in our case, offer valuable diagnostic clues for osteosarcoma.[9] MRI remains the gold standard for detailed assessment, providing crucial information about tumor extent, soft tissue involvement, and proximity to surrounding structures, which is essential for treatment planning and monitoring response to therapy.[7] CT plays a supportive role, particularly in planning biopsies and staging the disease, with its primary utility being in the detection of metastases, especially in the chest. Thus, a comprehensive approach utilizing radiographs, MRI, and CT, as appropriate, ensures optimal diagnosis and management of osteosarcomas.[3]

Osteosarcomas are categorized into osteoblastic, chondroblastic, and fibroblastic types based on their predominant differentiated element. In the research conducted by Low CM et al, osteoblastic osteosarcoma was the most prevalent histologic subtype in sinonasal osteosarcoma, occurring in 76% of cases. This was followed by chondroblastic osteosarcoma at 32% and fibroblastic osteosarcoma at 20%.[4]

Histologically, spindle-shaped tumor cells are a key diagnostic feature of osteosarcoma, though distinguishing them from dense collagen or amorphous eosinophilic osteoid can be challenging.[10] Techniques such as cell blocks, immunocytochemistry, and electron microscopy are crucial for subtype differentiation. Typically, osteoblastic subtypes show positive vimentin staining and negative cytokeratin staining, with S100 protein usually negative unless a chondroblastic component is present .[9]

AJCC classification provides a structured approach to staging and grading, which is essential for determining prognosis and planning treatment. Low-grade tumors are classified as stage I, regardless of the extent of the primary tumor. High-grade tumors are categorized as Stage II. Metastatic tumors are classified as Stage III, irrespective of their grade 11. High-grade sinonasal osteosarcoma is more common in sinonasal osteosarcoma, affecting 63% of patients, compared to low-grade disease at 37%.[4] Additionally, high-grade tumors in sinonasal osteosarcoma tend to have fewer distant metastases than those in extremity osteosarcoma.[12]

In the treatment of sinonasal cancers, surgery is the gold standard when feasible.[13] It may be supplemented with adjuvant therapies as needed. Surgical approaches such as open, endoscopic, or combined approaches have been described. The choice depends on several factors: general criteria related to the oncological outcomes and morbidity

of each technique, specific criteria related to the tumor (such as tumor extension and pathology), as well as considerations regarding the patient and the surgeon's expertise.[13]

In our case, an open surgical approach of partial rhinectomy and bilateral maxillectomy via the midfacial degloving approach was opted for. The incision was made at the level of the mucosa of the bilateral upper labial, extending straight to the bone and superomedially to include the dorsal nasal flap. Exposure was achieved by raising the flap of the periosteum and the soft tissues, similar to the technique of lateral rhinotomy approach. This provides access to the mid-face skeleton without requiring a skin incision and avoids scarring over the face. Osteotomies were done with chisel and mallet anterolaterally between the canine and first premolar bilaterally; posteroinferiorly between the soft and hard palate junction, posterolaterally at the pterygoid plate; and superiorly at the nasal bone and vomer. The tumor was removed en bloc, including the tumor, septum, nasal cartilages, hard palate (teeth 15 to 24), and the anterior part of the bilateral medial wall of the maxillary sinuses.

Due to the limited number of sinonasal osteosarcoma cases, there is no consensus on the optimal treatment approach. However, it is generally agreed that surgery is the primary treatment and achieving negative surgical margins is essential.[4] Studies show a 75% survival rate for patients with negative resection margins versus only 32% for those with positive margins.[3] The 5-year overall survival (OS) rate is 40%, while the disease-specific survival (DSS) rate is 43.9%. Prognostic factors include increasing age and T4 staging, which are predictors of a worse DSS rate.[4]

Adjuvant chemotherapy improves overall survival. Common chemotherapeutic agents include doxorubicin, ifosfamide, cisplatin, and methotrexate, administered for 7 to 12 months, which has improved the 5-year survival rate from 20% to 60%.[14]

Radiotherapy is generally used as an adjunct treatment in cases where complete surgical resection is not possible, as well as in cases where intralesional resection has been performed or for palliation of symptomatic metastases.[15] In these cases, craniofacial intensity-modulated beam radiation therapy (CIBRT) appears to offer promising results, with superior overall survival rates, showing 92.9% at 1 and 2 years, and 83.6% at 3 and 5 years.[16]

## CONCLUSION

Osteosarcomas are highly malignant primary bone tumors that rarely occur in the head and neck region. This case serves as an important learning opportunity, highlighting the rare diagnosis of osteoblastic osteosarcoma in this area. When dealing with extensive nasal swelling, as in our case, prioritizing the exclusion of malignant conditions is crucial for practitioners. Thorough preoperative planning with an emphasis on radiographic imaging allowed us to identify crucial borders and structures to protect during the excision. This careful planning was essential to ensure the complete removal of the tumor, as incomplete excisions are linked to higher recurrence rates. Effective management requires prompt, multidisciplinary intervention to improve survival and outcomes for these patients.

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