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Case Report

An unusual case of maxillary sinus involvement by a malignant peripheral nerve sheath tumour

Devika Thayyezhuth*, Shashidhar Kallappa, Ravi Koppad, Sanjay R. Pawar, Dastayya Guttedar, Sheetal Ishwarappagol

Department of Surgical Oncology, Karnataka Medical College and Research Institute, Hubballi, Karnataka, India

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***Correspondence:**
Dr. Devika Thayyezhuth,
E-mail: deevooo@gmail.com

ABSTRACT

Malignant peripheral nerve sheath tumours (MPNSTs) are uncommon aggressive soft-tissue sarcomas, accounting for approximately 5–10% of all sarcomas. Their occurrence in the head and neck region is rare, and involvement of the paranasal sinuses, particularly the maxillary sinus is exceedingly uncommon. Here we report a case of a middle-aged patient who presented with pain and swelling over left cheek, hard palate and nasal obstruction since 3 months. Clinical examination revealed a spherical swelling of size 3×2 cm present over left cheek with indistinct borders, another spherical swelling of size 3×2 cm on left side of hard palate. Patient underwent multidisciplinary evaluation, leading to surgical resection. Histopathological examination proved as low grade malignant peripheral nerve sheath tumour with Immunohistochemistry positivity to Vimentin. Patient underwent adjuvant radiotherapy and post-treatment follow-up showed no evidence of recurrence. This case emphasizes the significance of a broad differential, thorough histopathologic assessment, and coordinated multidisciplinary care to optimize outcomes in such rare presentations. Limited cases in the literature make diagnosis and management particularly challenging.

Keywords: Malignant peripheral nerve sheath tumour, Maxillary sinus, Neurofibromatosis, Immunohistochemistry

INTRODUCTION

Malignant peripheral nerve sheath tumours (MPNSTs) are malignant tumours which is extremely rare sarcomas arising from the Schwann cells of peripheral nerves.[1,2] These tumours contribute overall incidence of 0.001% in the general population.[3]

In head and neck region these tumours are uncommon and occurs approximately 10% of all cases.[4,5] These are most aggressive tumours which have high recurrence rate and propensity for dissemination and metastasis if it is not timely intervened. Clinically, sinonasal MPNSTs present with non-specific symptoms including nasal obstruction, facial fullness, epistaxis, pain, or cranial nerve deficits, which often mimic more commonly as benign and malignant sinonasal diseases.[6]

Radiologic findings, although indicative of an aggressive lesion, they are typically non-pathognomonic and overlap with a wide spectrum of spindle-cell neoplasms.[7] Consequently, definitive diagnosis relies heavily on histopathologic examination and immunohistochemical analysis, both of which may be complicated by the tumour's variable morphologic patterns and inconsistent marker expression. Given the rarity of sinonasal presentations and the limited consensus on optimal treatment, each case adds valuable insight into improving recognition, diagnostic accuracy, and management strategies. Here we present a rare case of maxillary sinus MPNST which remains an exceptional entity with non-specific clinical and radiologic characteristics that can delay accurate diagnosis.

CASE REPORT

37-year-old man presented with 3-month history of pain and swelling over left cheek, hard palate and nasal obstruction which progressed in the last two months.

Clinical examination revealed a spherical swelling of size 3×2 cm present over left cheek with indistinct borders; no abnormality visualized over skin. No sensory deficit. A mass lesion was seen in anterior end of middle turbinate in left nasal cavity. Oral cavity examination showed another spherical swelling of size 3×2 cm on left side of hard palate extending horizontally from midline to premolar and 3rd molar vertically (Figure 1).

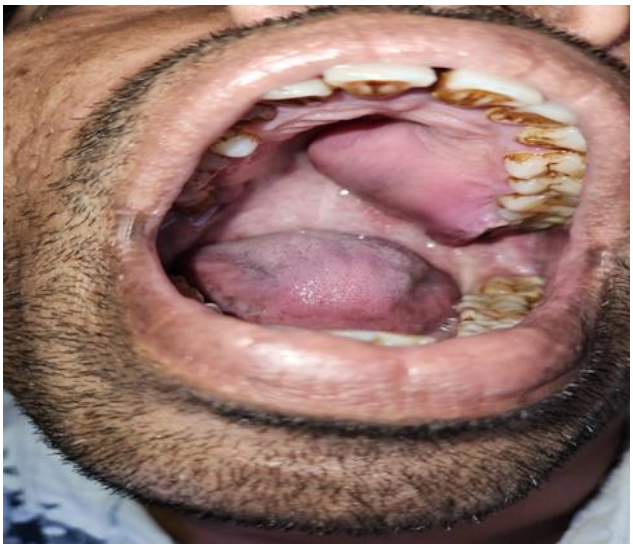

Figure 1: Swelling over hard palate.

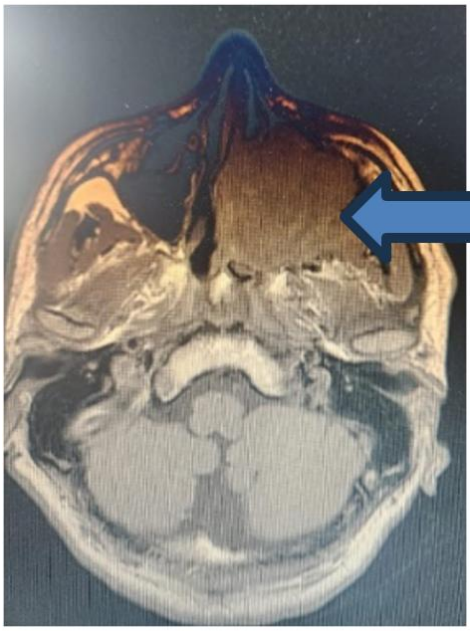

Figure 2: CT showing soft tissue lesion occupying left maxillary sinus with erosion of anterior, medial and inferior walls.

A contrast enhanced computed tomography (CECT) of PNS revealed a large homogeneous enhancing soft tissue lesion (4.7×4.3×5.3 cm) in left maxillary sinus with erosion and destruction of anterior, medial and inferior wall of left maxillary sinus (Figure 2).

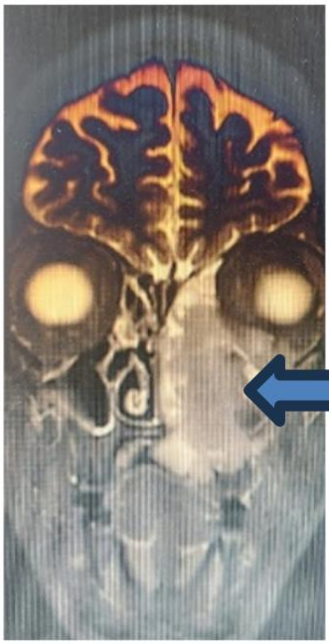

Figure 3: MRI showing lobulated hyperintense lesion in left maxillary sinus, eroding medial and anterior walls of maxillary sinus, extending medially into nasal cavity, with no intracranial extension.

Magnetic resonance imaging (MRI) was performed which showed ill-defined lobulated hyperintense lesion (4.5×4.8×3.7 cm) noted in left maxillary sinus, eroding medial and anterior walls of maxillary sinus, extending medially into nasal cavity, laterally into post antral region with no intracranial extension (Figure 3).

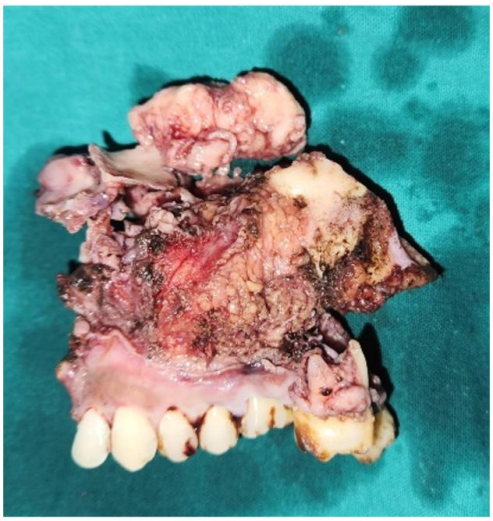

Figure 4: Specimen showing left total maxillectomy with tumour.

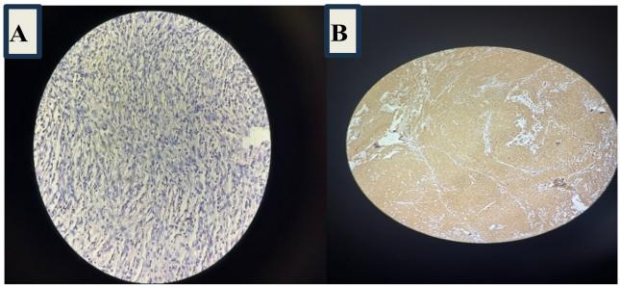

Figure 5: Specimen histology. (A) HES-stained cross section showing spindle cells arranged in fascicles within collagenous stroma and (B) immunohistochemical staining showing positive vimentin.

Total left maxillectomy was performed via Lateral rhinotomy approach (Figure 4). Histopathological examination proved as low grade MPNST of maxillary sinus (Figure 5A). Immunohistochemistry (IHC) showed strongly positive for Vimentin (Figure 5B), focally positive to S100. He received (60 Gy) 30 cycles of adjuvant radiotherapy. Assessment at 1 year after end of treatment found no tumour recurrence.

DISCUSSION

MPNSTs originates from components of the nerve sheath, including Schwann cells and perineural fibroblasts. Although uncommon, they represent a small but notable proportion of sarcomas affecting the head and neck region, accounting for 2-6% of cases.[8] Despite their rarity, MPNSTs are clinically significant because they tend to behave aggressively, recur frequently, and metastasize early. These tumours may develop spontaneously or occur as part of neurofibromatosis type 1 (NF1). Approximately half of all reported cases have been linked to NF1; however, our patient had no family history suggestive of the condition, raising the likelihood of a sporadic, de novo tumors.

MPNSTs most often arise in the extremities particularly the lower limbs and in the retroperitoneum. Less frequently, they occur in the trunk, upper extremities, and head and neck. Within the head and neck, reported sites include the nasopharynx, nasal cavity and paranasal sinuses, orbit, oral cavity, parapharyngeal space, neck, thyroid, and larynx.[9,10]

Establishing a diagnosis can be challenging because both clinical presentation and imaging findings may mimic benign nerve sheath tumours such as neurofibromas or schwannomas. Rapid enlargement, persistent pain, or neurologic impairment can raise suspicion, but these warning signs are not always present early in the disease course. MRI is the preferred modality, although no single radiologic feature is definitive. Prior studies have noted that irregular margins, surrounding oedema, and heterogeneous internal signal patterns may be more typical of MPNST, but they overlap with benign lesions which limits diagnosis certainty and reinforces the need for tissue sampling.[7]

Histologically, MPNSTs are composed of spindle-shaped tumour cells arranged in intersecting bundles and often display nuclear atypia, increased mitotic activity, and areas of necrosis. IHC remains valuable in supporting the diagnosis; markers of neural differentiation such as S100 or SOX10 may be present but are commonly weak or focal in high-grade tumours.[11] In our case, the tumour stained strongly for vimentin and showed focal S100 positivity, supporting the diagnosis of a low-grade MPNST.

Management of MPNST requires treatment in a multidisciplinary sarcoma centre where surgical oncology, specialized pathology, radiation oncology, medical oncology and rehabilitation services can coordinate care. Surgical removal with clear margins is generally viewed as the most effective treatment strategy for localized tumours and is the strongest predictor of local control and overall survival.[12] Adjuvant radiotherapy is recommended for high-grade tumours, tumours more than 5 cm, close or positive margins, or when re-excision for margin clearance is not feasible.[13] The role of chemotherapy in MPNST is less clearly defined but may be considered in selected high-grade or advanced cases, particularly in the neoadjuvant setting. Overall outcome remains guarded, with reported 5 year survival rates varying widely depending on tumour size, grade, NF1 association, and adequacy of surgical margins.[14] Early detection and prompt surgical management are essential for improving outcomes.

CONCLUSION

MPNST is a rare but highly aggressive sarcoma that requires early recognition, accurate histopathological diagnosis, and prompt multidisciplinary management. Wide surgical excision with negative margins remains the cornerstone of treatment, with adjuvant radiotherapy considered in high-risk or large tumours to improve local control. Future research may demarcate the role of targeted therapy for patients with MPNST. In view of rare presentation reporting such cases contributes valuable information to the existing literature and enhances awareness of this uncommon neoplasm in an unusual anatomical location.

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