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Malignant Schwannoma of the Infratemporal Fossa with Perineural Extension in a Young Female: A Case Report

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ABSTRACT

Background : Malignant peripheral nerve sheath tumors (MPNSTs), historically referred to as malignant schwannomas, are rare soft tissue sarcomas originating from Schwann cells or peripheral nerve sheath elements. Head and neck involvement accounts for only a small proportion of cases, while localization within the infratemporal fossa is exceptionally uncommon. The deep anatomical location, proximity to critical neurovascular structures, and nonspecific clinical presentation often delay diagnosis.

Case Presentation : We report the case of a 20-year-old woman with no significant past medical history who presented with a nine-month history of progressive left cheek swelling. Clinical examination revealed a firm, fixed, painless jugal mass measuring approximately 5 cm without inflammatory signs. Cranial nerve examination demonstrated preserved trigeminal nerve (V1 and V2) sensation. Nasal endoscopy was unremarkable. Magnetic resonance imaging demonstrated a 32.4 × 42.7 × 50 mm parapharyngeal and infratemporal fossa mass exhibiting multidirectional extension along neural pathways toward the pterygopalatine ganglion. Complete surgical excision was achieved through a Le Fort I osteotomy approach. Histopathological and immunohistochemical analyses showed focal nuclear positivity for S100 protein and negativity for cytokeratin, smooth muscle actin, CD34, HMB45, and desmin, supporting the diagnosis of malignant schwannoma (FNCLCC grade II). Surgical margins were negative. The patient subsequently underwent adjuvant radiotherapy

Conclusion : MPNSTs of the infratemporal fossa remain exceedingly rare. Complete surgical resection with tumor-free margins remains the cornerstone of treatment, while adjuvant radiotherapy may improve local disease control. Early recognition and multidisciplinary management are essential to optimize outcomes.

KEYWORDS : Malignant schwannoma; MPNST; infratemporal fossa; Le Fort I osteotomy; perineural spread; head and neck sarcoma.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) represent approximately 5–10% of all soft tissue sarcomas and arise from Schwann cells, perineural cells, or fibroblasts associated with peripheral nerves [1,2]. Their annual incidence in the general population is estimated at 0.001%, making them among the rarest sarcomas encountered in clinical practice [3].

The head and neck region accounts for only 8–16% of all MPNSTs [4]. Within this anatomical territory, the infratemporal fossa represents an exceptionally rare site of occurrence because of its deep location and limited neural tissue volume. Consequently, most knowledge regarding infratemporal fossa MPNSTs is derived from isolated case reports and small case series [5].

Clinical manifestations are often nonspecific and depend on the anatomical structures involved. Facial swelling, pain, trismus, cranial nerve dysfunction, and orbital symptoms may occur as the tumor enlarges [6]. Radiological imaging, particularly magnetic resonance imaging (MRI), is essential for assessing tumor extension and identifying perineural spread, which constitutes an important hallmark of aggressive behavior [7].

Histopathological diagnosis remains challenging because MPNSTs may mimic other spindle cell neoplasms. Immunohistochemistry plays a crucial role, with S100 positivity observed in a subset of cases, whereas epithelial, muscular, melanocytic, and vascular markers are usually negative [8].

We report a rare case of a malignant schwannoma arising in the infratemporal fossa with suspected perineural extension in a young woman successfully treated by complete surgical excision through a Le Fort I osteotomy approach followed by adjuvant radiotherapy.

CASE PRESENTATION

A 20-year-old female with no significant past medical history presented to our department with a progressively enlarging left facial swelling evolving over nine months.

The patient reported gradual enlargement of the mass without associated pain. No rhinological symptoms, including nasal obstruction, epistaxis, rhinorrhea, or anosmia, were noted. Likewise, she denied visual disturbances, diplopia, proptosis, headaches, facial paresthesia, or other neurological complaints. General health remained preserved, with no weight loss, fever, or constitutional symptoms.

Physical examination revealed a left jugal swelling causing mild facial asymmetry. Palpation demonstrated a firm, fixed, painless mass measuring approximately 5 cm in greatest diameter. The overlying skin was normal, without erythema, warmth, or ulceration. Neurological examination showed preserved sensation within the ophthalmic (V1) and maxillary (V2) territories of the trigeminal nerve. No facial nerve deficit was detected. Oral cavity examination was normal.

Rigid nasal endoscopy revealed no endonasal mass or mucosal abnormality.

MRI demonstrated a well-defined deep facial mass located within the infratemporal and parapharyngeal spaces. The lesion measured 32.4 × 42.7 × 50 mm. It appeared hypointense on T1-weighted sequences, showed intermediate T2 signal intensity, and exhibited intense enhancement following gadolinium administration. Superior extension toward the infraorbital fissure and orbit was observed, with displacement of the inferior rectus muscle but no exophthalmos. Imaging suggested multidirectional extension along neural pathways toward the pterygopalatine ganglion, raising suspicion for perineural spread. No cervical lymphadenopathy was identified.

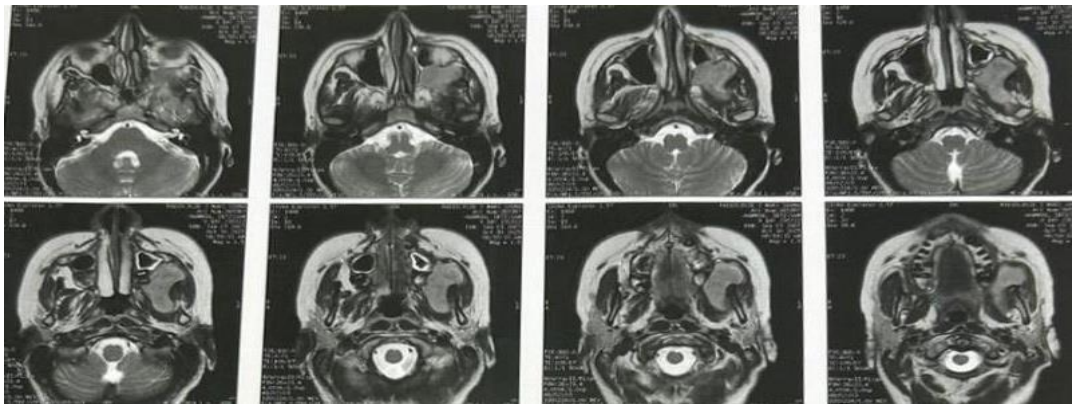

Figure 1: Preoperative axial MRI sequences demonstrating the infratemporal and parapharyngeal mass.

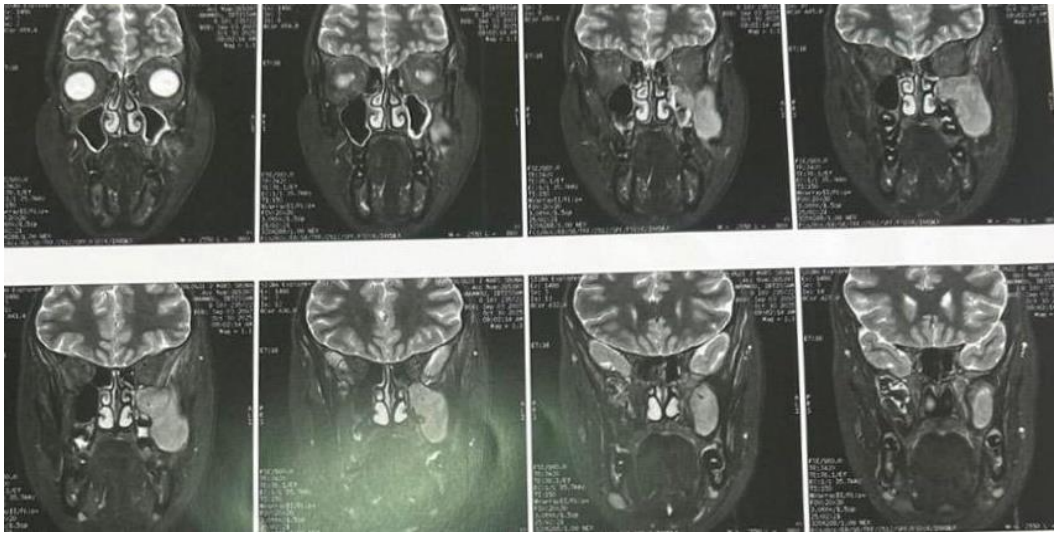

Figure 2: Gadolinium-enhanced T1-weighted MRI showing intense tumor enhancement with multidirectional extension along neural pathways toward the pterygopalatine ganglion.

Given the deep location and suspected malignant nature of the lesion, surgical management was undertaken.

A transmaxillary approach using a Le Fort I osteotomy was performed to provide adequate exposure of the infratemporal fossa. Complete macroscopic excision of the tumor was achieved. Intraoperatively, the lesion appeared encapsulated in certain areas but demonstrated close relationships with surrounding neurovascular structures.

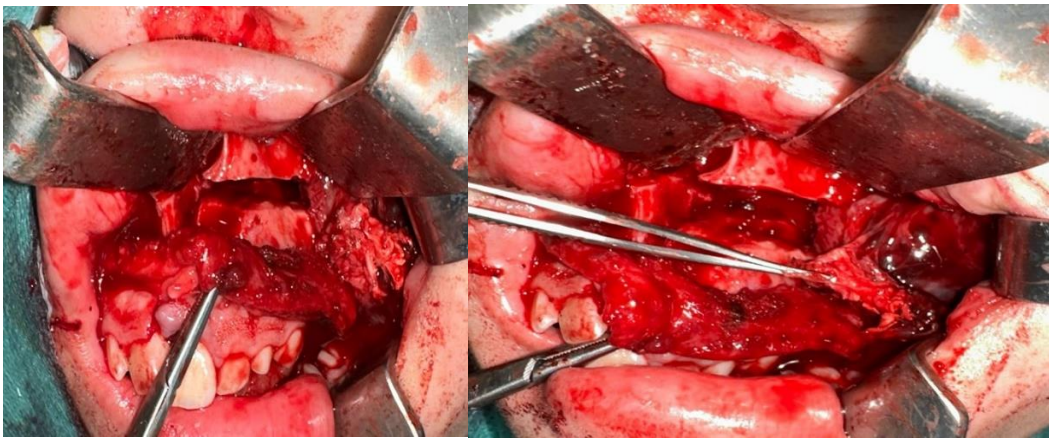

Figure 3 & 4: Intraoperative views after Le Fort I osteotomy showing surgical exposure of the infratemporal fossa and dissection of the tumor in close relation to surrounding neurovascular structures.

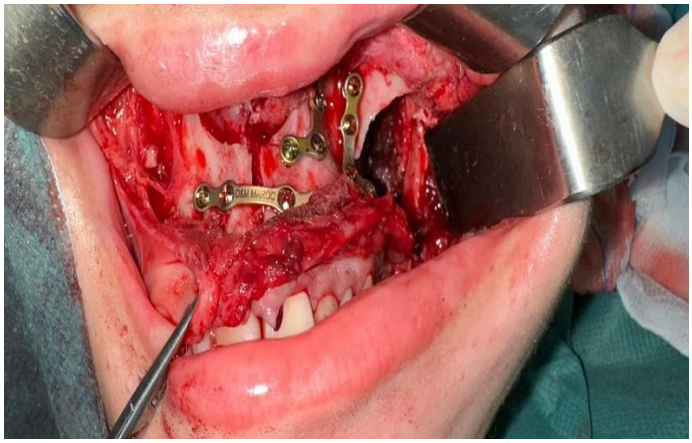

Figure 5: Intraoperative view after tumor excision showing plate osteosynthesis following Le Fort I repositioning.

Histopathological examination revealed a malignant mesenchymal spindle cell neoplasm. Immunohistochemical analysis demonstrated focal predominantly nuclear positivity for S100 protein. Cytokeratin, smooth muscle actin, CD34, HMB45, and desmin were all negative.

These findings supported the diagnosis of **malignant schwannoma (malignant peripheral nerve sheath tumor)**. According to the French Federation of Cancer Centers Sarcoma Group (FNCLCC) grading system, the tumor was classified as Grade II (score 3+1+0).

Importantly, surgical margins were free of tumor.

Following multidisciplinary tumor board discussion, adjuvant radiotherapy was administered because of the malignant nature of the lesion and the high risk of local recurrence associated with head and neck MPNSTs.

At follow-up, the patient remained clinically stable without evidence of local recurrence.

DISCUSSION

MPNSTs are highly aggressive sarcomas characterized by local invasiveness and a tendency for recurrence [1]. Their occurrence within the infratemporal fossa is exceedingly rare, making diagnosis and management particularly challenging.

The etiology of MPNST remains incompletely understood. Approximately 25–50% of cases occur in association with Neurofibromatosis Type 1 (NF1), while sporadic forms account for the remainder [9]. Our patient had no clinical history suggestive of NF1.

The age distribution of MPNST typically peaks between the third and sixth decades of life [10]. The occurrence in a 20-year-old woman therefore represents an unusual presentation.

Clinical manifestations depend largely on tumor size and anatomical extension. In contrast to previously reported cases presenting with pain, trismus, cranial neuropathies, or visual symptoms, our patient presented solely with progressive facial swelling despite significant tumor dimensions [11].

MRI remains the imaging modality of choice. Typical findings include iso- to hypointense T1 signal, heterogeneous T2 signal, and contrast enhancement [7]. In the present case, imaging additionally suggested perineural extension toward the pterygopalatine ganglion, a recognized route of spread for tumors arising from branches of the trigeminal nerve [12].

Histologically, MPNSTs are characterized by spindle-shaped cells arranged in fascicles with varying degrees of atypia and mitotic activity [13]. Immunohistochemistry is essential to exclude differential diagnoses such as synovial sarcoma, leiomyosarcoma, fibrosarcoma, spindle cell melanoma, and solitary fibrous tumor.

S100 positivity is observed in approximately 50–60% of MPNSTs and is usually focal rather than diffuse [14]. The immunophenotype observed in our case, namely focal S100 positivity with negativity for cytokeratin, desmin, SMA, HMB45, and CD34, strongly supported the diagnosis.

Complete surgical resection with negative margins remains the primary treatment modality [15]. However, achieving clear margins in the head and neck region is often difficult because of the complex anatomy and the proximity of vital neurovascular structures.

The Le Fort I osteotomy approach used in our patient provided excellent access to the infratemporal fossa while allowing complete tumor excision. This technique has been described as an effective route for deep-seated skull base and infratemporal lesions [16].

The role of radiotherapy in MPNST remains debated. Nevertheless, several studies have demonstrated improved local control following adjuvant radiotherapy, particularly for tumors larger than 5 cm, intermediate- or high-grade lesions, and tumors arising in anatomically constrained regions [17,18].

The prognostic factors most consistently associated with survival include tumor size, surgical margin status, histological grade, and local recurrence [19]. Negative margins achieved during surgery constitute one of the most favorable prognostic indicators in the present case.

Reported five-year overall survival rates range from 34% to 60%, emphasizing the aggressive nature of these tumors [20]. Long-term surveillance remains mandatory because recurrences may occur several years after treatment.

CONCLUSION

Malignant schwannoma of the infratemporal fossa is an exceptionally rare neoplasm. Diagnosis requires careful integration of clinical, radiological, histopathological, and immunohistochemical findings. This case highlights the importance of considering MPNST in the differential diagnosis of deep facial masses in young adults. Complete surgical excision through a Le Fort I osteotomy approach allowed achievement of tumor-free margins, while adjuvant radiotherapy was administered to reduce recurrence risk. Long-term follow-up remains essential because of the aggressive biological behavior of these tumors.

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