

Schwannoma of the infratemporal fossa - A rare case report

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ABSTRACT

Schwannoma is a benign nerve sheath tumour that originates from schwann cells of the peripheral nerve. In spite of constituting 25-40% of head and neck tumours its intraoral presentation is rare which accounts for only 1%.

The purpose of this case report is to highlight the rarity of this lesion, to emphasize the significance of an accurate diagnosis and to include tumours of nerve sheath origin in the differential diagnosis of facial asymmetry.

Keywords

Schwannoma, nerve sheath, head and neck region.

INTRODUCTION

Schwann cells intimately ensheath all neurons of both myelinated and non-myelinated nerves in the peripheral nervous system.¹ Schwannomas occurring in the head and neck region constitute 25–45% of head and neck tumours.²

The present case of schwannoma is limited to the infratemporal fossa, which is a rare location, arising from the extradural division of the trigeminal nerve. Trigeminal schwannomas account for about 0.2% of all intracra-

nial, tumours and 0.8% and 8% of intracranial schwannomas.³ Surgically a zygomatico-temporal approach was planned to provide access to such a cryptic anatomical region whilst ensuring a satisfactory cosmetic result.

CASE REPORT

An 18-year-old female reported with a main complaint of a gradually increasing painless swelling on right side of the face of 4 months duration. It extended from right ala of nose to preauricular notch, anteroposteriorly, and from right maxillary buccal vestibule to right infraorbital region, superoinferiorly (Fig. 1).

Intraorally, the right maxillary buccal vestibule was obliterated. The swelling was firm on palpation and the patient presented with difficulty in opening of mouth. There were no neurological signs on the right side of the face.



Figure 1. Extra-oral swelling extended from right ala of nose to preauricular notch, anteroposteriorly, and superoinferiorly from right maxillary buccal vestibule to right infraorbital region.

T2 weighted MRI demonstrated a large well defined, heterogeneously enhanced mass measuring 5x4.4x4.2 cm in right infratemporal fossa. The posterior wall of the right maxillary antrum was bowed anteriorly and the floor of the orbit was attenuated by the mass (Fig. 2A). The lesion showed extension into the right cavernous sinus. Widening of foramen rotundum was noted.

Based on the clinical and radiological findings, a provisional diagnosis of a benign soft tissue neoplasm in the right infratemporal fossa was made. An intraoral incision biopsy was performed under local anaesthesia from right maxillary molar region. Histopathologically, connective tissue was shown to be composed of numerous spindle cells arranged in fascicles and in a palisading pattern depicting hypercellular Antoni A arrangement.

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Author contributions:

1. **Shivani Bansal:** Examined the patient; conceiving the idea for the case report, contributed to writing the case report, edited the case report and approved the final draft - 20%
2. **Dilipkumar Basavanhappa:** Treated the case, approved the final draft of the case report - 20%
3. **Pooja Prasad:** Lead the writing of the case report, approved the final draft - 20%
4. **Mugdha S Raut:** Lead the writing of the case report, approved the final draft - 20%
5. **Rajiv S Desai:** Examined the patient; conceiving the idea for the case report, approved the final draft - 20%

The spindle cells were characterized by flattened elongated and tapered nuclei. Presence of chronic inflammatory infiltrate was seen; neural bundles were also evident in a few areas of the stroma consistent with a diagnosis of spindle cell neoplasm (Fig. 2B). Immunohistochemical analysis showed S100 positivity (Fig. 2C). Based on the clinicopathological and immunohistological features, a final diagnosis of the spindle cell lesion Schwannoma was established.

The tumour resection was planned through zygomatico-temporal approach with osteotomy of zygomatic complex bony structure for access to infra temporal space and maxilla (Fig. 2D) for ensuring a satisfactory cosmetic result. The whole zygomatic complex bony structure was retracted laterally to expose the tumour in the infra temporal fossa. The tumour was enucleated as a single mass. After excision the tumour, a rigid fixation of osteotomised bony complex back in premarked position was done with miniplates. Healing was uneventful. A 5-year follow-up showed no sign of recurrence.

DISCUSSION

Schwannoma (neurilemmoma) is a rare, benign, encapsulated perineural tumour of neuroectodermal origin that is derived from the Schwann cells of the neural sheath. Earlier, Verocay⁴ referred to it as a "neuronoma." Later, Stout used the term, "neurilemmoma," believing that this tumour originated from the Schwann cells.⁴

Twenty-five to forty-eight percent of all cases of schwannomas occur in the head and neck region.⁵⁻⁷ It is more common between 30 and 50 years of life.⁸ William et al.⁹ showed that in 83% of the cases studied by them, schwannomas presented in males, while for Lucas, there was a greater predilection for females which is in accordance to our case and for Hatziotis⁹ and Asprides, Enzinger and Weiss,⁹ there was an equal distribution.^{4,9} Within the head and neck region, the tongue, palate, cervical and pharyngeal regions are frequently involved, but a lesion in the infratemporal fossa is relatively rare.^{8,10,11}

Clinically Schwannoma presents with symptoms related to the nerve involved maybe associated with pain along with other symptoms such as hearing loss, hoarseness and dysphagia, depending on the site of involvement. However, lack of symptoms is common when the lesion originates from the trigeminal nerve and is located in the infratemporal fossa which is in accordance to our case report.¹¹

Trigeminal schwannomas account for about 0.2% of all intra cranial tumors, and 0.8% and 8% of intracranial schwannomas.³ In the present case, the tumour showed evidence of originating from the second division of the fifth cranial nerve near the foramen rotundum. They can originate from any section of the fifth cranial nerve, from the root to the distal extracranial branches, but the majority develops at the Gasserian ganglion, usually growing in the middle cranium.³ In our case, though intracranial extension into the right cavernous sinus was seen, during surgery the duramater was found to be intact. It was concluded that the tumour primarily originated within infratemporal fossa.

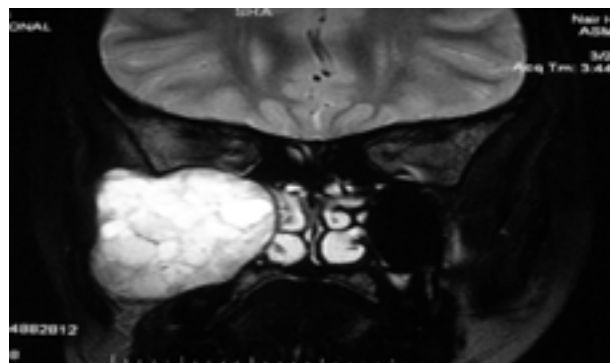


Figure 2A. T2 weighted MRI demonstrated a low-density mass in the right infratemporal fossa and bowing in of the right maxillary sinus.

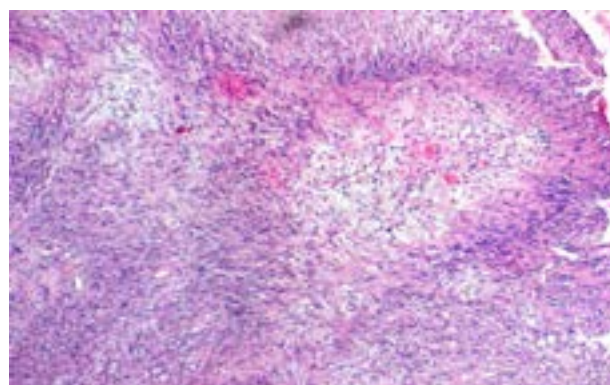


Figure 2B. Photomicrograph demonstrates cellular neoplastic connective tissue arranged in short fascicles, composed of cells with aligned long nuclei and few spindle cells randomly arranged within a loose stroma (H&E 100x).

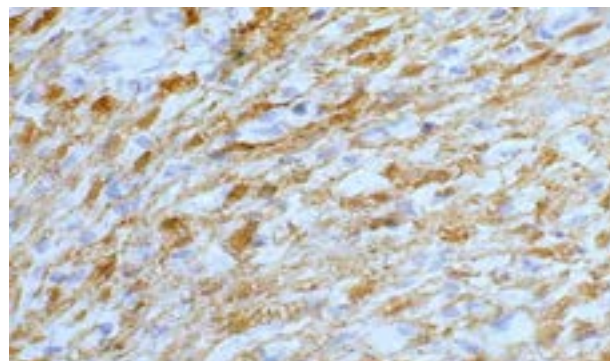


Figure 2C. The tumour cells showed positive immunohistochemical reactivity for S100 protein.



Figure 2D. Intra operative picture showing exposure of the tumour through temporal approach with osteotomy of zygomatic complex bony structure for access to infratemporal space and maxilla.

The infratemporal region is the irregular retromaxillary space, inaccessible by virtue of its relatively concealed location, and is remote for clinical examination.¹² Tumours in this clinically silent area always pose a problem in diagnosis and management. They cannot be seen clinically while they are still confined within the boundaries of the fossa, and localizing signs and symptoms usually do not become apparent until the tumour reaches a significant size.¹¹ In the present case, the patient was asymptomatic, radiographs showed erosion of the bony walls of infratemporal fossa, thinning of floor of orbit and extension into the sphenoid sinus.

CONCLUSION

Though rare, the treatment of infratemporal schwannomas provide a great surgical challenge. The low incidence of schwannomas often leads to the omission of the tumour in the differential diagnosis of soft tissue tumours of the oral cavity. We have presented a unique case of a schwannoma of the infratemporal fossa to stress its rarity and importance of ruling out neurogenic tumours in differential diagnosis of facial asymmetry without paraesthesia.

Declaration

The authors of this manuscript declare no conflict of interests.

Ethics statement

Ethics approval is obtained. All details of the patient have been anonymized.

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