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Maxillary Sinus Schwannoma—A Rare Tumor with Rarer Site of Occurrence

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Abstract

Schwannoma or neurilemmoma is a neurogenic tumor. Around 25% - 45% cases of schwannomas occur in the head and neck, of which less than 4% occurs in the nasal cavity and the paranasal sinuses. Isolated schwannomas of the maxillary sinus appear to be extremely rare. We present one such rare case of Right Maxillary Sinus schwannoma in a 48-year-old lady with swelling in the right cheek for 3 years. Infrastructure maxillectomy of the right side was done using Weber-Fergusson approach with Dieffenbach's modification. The patient made a good postoperative recovery. We report this case keeping in mind the rarity in occurrence of isolated maxillary schwannomas.

Keywords

Schwanoma, Maxillary Sinus, Maxillectomy

1. Introduction

Schwannomas are the benign tumors that arise from the Schwann cells, which form the nerve sheath of the peripheral nerve fibers. These are well-encapsulated masses, clearly circumscribed, and connected to neural tissue of origin. Around 25% - 45% cases of schwannomas occur in the head and neck region of which less than 4% affect the nasal cavity and the paranasal sinuses [1]. Schwannomas arising solely from maxillary sinus are extremely rare [2]. Schwannomas arising solely from maxillary sinus are extremely rare. To our knowledge, only eight cases of isolated maxillary schwannomas have been reported so far [3]. We present one such rare case encountered at our center with relevant literature.

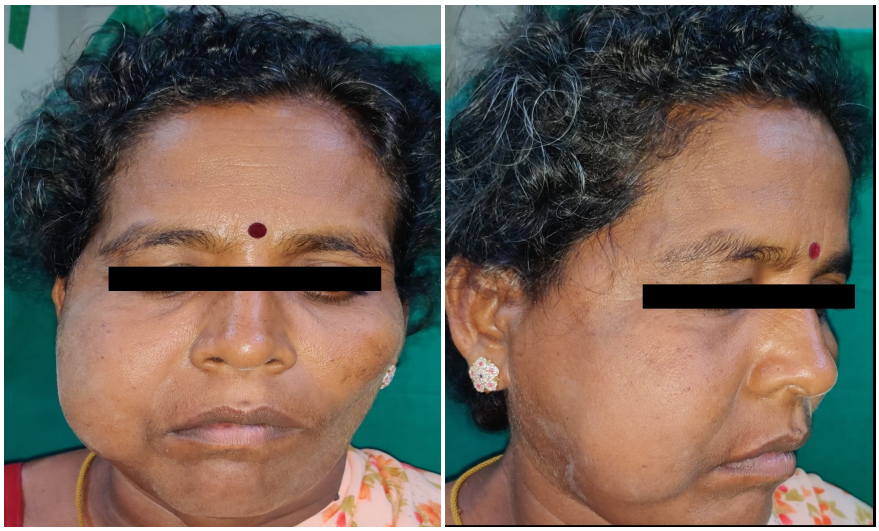
2. Case Report

A 48-year-old female presented to the surgical oncology department with com-

plaints of a swelling over the right cheek for 3 years, which was gradually increasing in size. The swelling was not associated with pain, epistaxis, fever, headache or visual disturbances (Figure 1). On clinical examination, a 7 × 8 cm firm, smooth, well defined, non-tender swelling, extending just above zygoma from tills the right sub-labial region with loss of nasolabial fold. No sensory deficit was seen over the right side of the face. The swelling was not fixed to the skin. Visual acuity was normal. There was no significant cervical lymphadenopathy. Intraoral examination demonstrated swelling of the right side extending posterior over hard palate, overlying mucosa was normal.

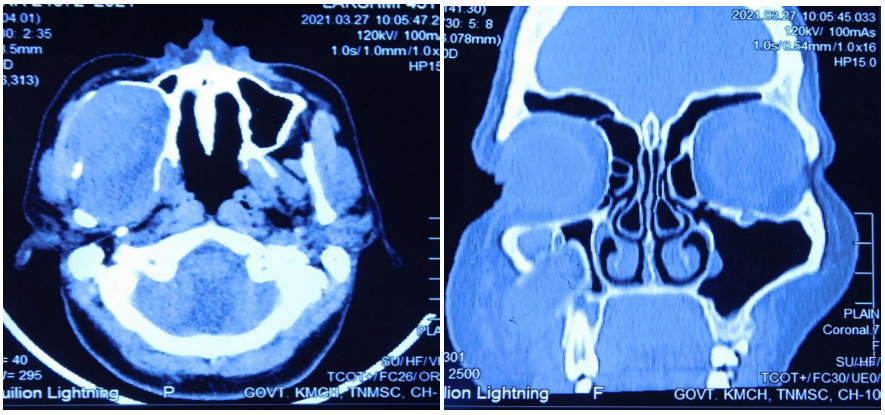
Computed tomography(CT) nose and paranasal sinus showed a soft tissue density lesion measuring 5.2 × 3.7 × 5.5 cm arising from infratemporal fossa on the right side, infiltrating the right pterygoid muscle, masseter and temporalis muscle and eroding the greater wing of right sphenoid (Figure 2).

MRI of paranasal sinus showed well defined expansile, smooth, marginated



(a) (b)

Figure 1. Swelling over the right cheek.



(a) (b)

Figure 2. Mass lesion was seen on computed tomography.

T1/T2 hetero intense lesion measuring 6.9 × 4.5 × 6.7 cm noted in Right maxillary sinus region. The lesion causes erosion of posterolateral wall of the right maxillar lesion involving the right superior orbital fissure. Medially, the lesion causes erosion of medial wall of the right maxillary sinus and infiltrates into right nasal cavity. Laterally, the lesion involves the right masticator space involving right masticator muscles and inferiorly the lesion involves left retromolar trigone. The lesion causes erosion of maxilla.

Under general anaesthesia, an endoscopic biopsy was performed and microscopic examination revealed spindle cell neoplasm of neural origin- probably schwannoma. Immunohistochemistry is suggestive of S-100 and CD 68 positive.

Subtotal maxillectomy under general anaesthesia was planned with Weber–Ferguson's incision with dieffenbach modification. Flaps were raised and entire maxilla was exposed. Osteotomy was performed and tumor was exposed and enucleation was done. Intraoral defect was packed with dental compound (Figure 3).

Postoperative period was uneventful. The intra-oral packs and dental fixate were removed on postoperative day 2. The patient was discharged on postoperative day 5 with antibiotics and analgesics. Dental obturator No complications were reported at 1 and 3-month follow-up (Figure 4).

Postoperative histopathology examination revealed encapsulated, well circumscribed solid mass (Figure 5). The presence of spindle cells was seen forming verocay bodies. Antoni A and B areas were identified. There was positive staining for S-100 protein (Figure 6). Hence, final histopathological report confirmed the diagnosis as schwannoma. Prosthetic functional rehabilitation was done with obturator.

3. Discussion

Schwannoma, also known as neurilemmoma, is a benign tumor of neuroectodermal

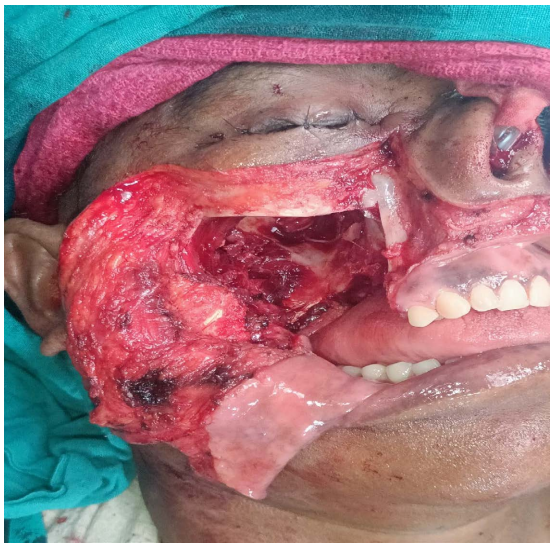


Figure 3. Intraoperative picture post excision of tumor.

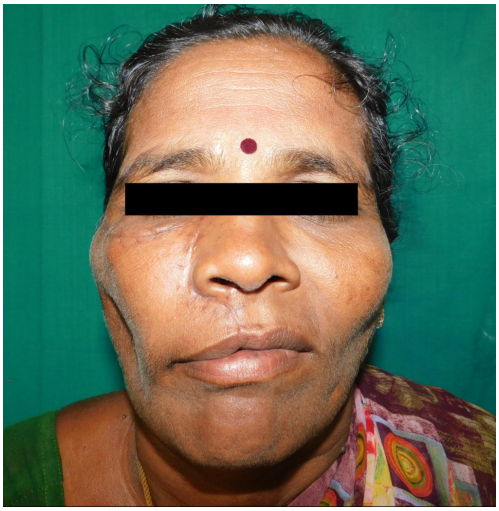


Figure 4. Postoperatively after one month.

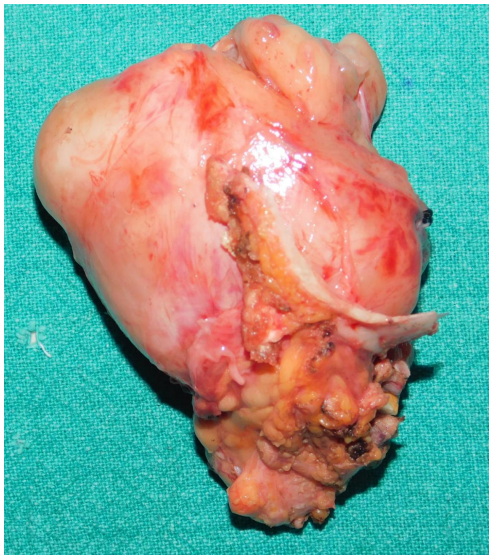
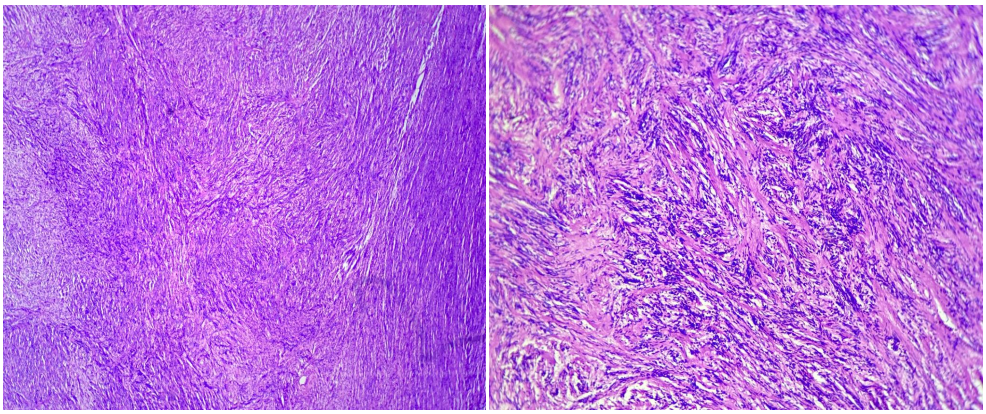


Figure 5. Whole specimen.



(a) (b)

Figure 6. Low power (100×) showing sheets and fascicles of spindle shaped cells (a); Cellular areas with verocay bodies (200×) (b).

origin which arises from Schwann cells of peripheral nervous system or cranial nerves. Schwannomas usually involves the head and neck region, but nasal cavity and para-nasal sinuses remain less common sites for the occurrence of this tumor [4]. Intranasal nerves, ophthalmic, and maxillary division of the trigeminal nerve and branches of the autonomic nervous system are the probable sites for developing schwannoma in the nose and para-nasal sinuses. The nerve giving rise to schwannoma may not be always identifiable. Schwannomas usually remain asymptomatic until they attain enormous size. Clinical features of schwannoma of paranasal air sinuses depend on site and affected nerve and compression of the surrounding neural structure. Pain and epistaxis may be seen due to the involvement of maxillary sinus and ethmoidal sinus, respectively [5]. Other clinical symptoms such as nasal blockage, rhinorrhea, hyposmia, exophthalmos, neuronal deficit, and facial swelling are also observed. The gold standard for diagnosis is histopathological examination which reveals compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas with hypercellular areas showing verocay bodies, as in our patient. These tumor cells typically show a diffuse positive immunoreactivity for S-100 protein. Virtually, all schwannomas are encapsulated. The treatment of choice is surgical enucleation or excision with regular follow-up [6]. Recurrence of tumor is rare if surgical enucleation is performed efficiently.

4. Conclusion

An isolated schwannoma of the maxillary sinus is a rare clinical entity. Treatment of choice of benign schwannoma is complete excision. The tumor has good prognosis with very minimal chance of recurrence.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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