A rare case of schwannoma of maxillary sinus

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ABSTRACT

Schwann cell tumor or schwannomas is the benign nerve sheath tumors, which arise mostly from head and neck region and rarely from the nasal and paranasal sinuses. Of these, very few arise solely from the maxillary sinus. This is a case report of a 25-year-old female patient presenting with painful swelling below the right eye turned out to be a rare case of schwannoma arising solely from the maxillary sinus. Details of clinical history, examination, histopathological finding, radiological finding, treatment, and follow-up have been presented.

Key words: *Antoni A and B, Maxillary sinus, Schwannoma, Verocay bodies*

Schwannomas are the tumors of the Schwann cell, which forms the nerve sheath of the peripheral nerve fibers. It is a well-encapsulated mass, clearly circumscribed, and well connected to neural tissue of origin. As reported in the literature, 25–45% cases of schwannoma occur in the head and neck region, of which only 2–4% affects the nasal cavity and the paranasal sinuses [1,2]. Only 2% of the cases of schwannomas have been reported to be malignant, and recurrence occurs in case the tumor is incompletely resected [3]. Schwannomas arising solely from maxillary sinus are extremely rare. As per the available reports, it seems only seven such cases have been reported [4]. We are reporting another case of a schwannoma arising solely from the maxillary sinus, which will be the eighth case to be reported so far.

CASE REPORT

A 25-year-old female presented to the plastic surgery outpatient department with complaints of a swelling below the right eye for 8 months, which was gradually increasing in size and pushing her lower eyelid upward. The swelling was slightly painful but was not associated with any history of spontaneous rupture, bleeding, trauma, nasal blockage, epistaxis, or fever. There was no history of any neurological deficit. The patient had a history of Caldwell-Luc operation done 4 years ago for a radicular cyst on the right cheek.

On clinical examination, a 3 cm × 3 cm, firm, well defined, mildly tender swelling, extending about 1 cm from medial canthus and 2 cm from lateral canthus was present over the right infraorbital region (Fig. 1a and b). The swelling was not fixed to the skin. Overlying surface of the skin was normal. There was no sensory deficit over the right side of the face. Visual acuity was normal, but field of vision was decreased inferiorly. We kept the differential diagnosis of odontogenic tumors like ameloblastoma, fibro-osseous lesions such as fibrous dysplasia, a soft tissue neoplasia, and any swelling which arises from that region like a radicular cyst. Neurofibroma was also considered as a differential during the histopathologic examination.

Magnetic resonance imaging of the face was done which showed an ill-defined mixed solid cystic lesion (measuring 2 cm × 5 cm × 2.8 cm) displaying signal intensity alteration with solid component showing heterogeneous post-contrast enhancement with non-enhancing necrotic area, filling the right maxillary sinus (Fig. 2). There was evidence of the mild expansion of the right maxillary sinus. The lesion was causing an erosion of the anterior wall of maxillary sinus with extension into the subcutaneous tissue up to the lower eyelid. Incisional biopsy was done. On histopathological examination, it was suggestive of schwannoma.

After diagnosis, *en bloc* removal of the tumor with the removal of an anterior wall of the maxilla with mucosal lining was performed by modified Weber Ferguson approach (Fig. 3a and b). Incision was given and deepened up to the bone. Subperiosteal dissection was done. The flap was elevated to expose the anterior wall of the right maxillary sinus. Intraoperative space-occupying lesion was found in the right maxilla. There was an erosion of anterior wall of the maxilla, with infiltration by the tumor. The tumor was excised along with an anterior wall of the maxilla and mucosal lining of the maxillary sinus. Sinus was packed with a saline bandage. The wound was then closed in layers. The reflected cheek flap was sutured to the normal anatomic site. Excised specimen was sent for histopathological examination in the department of pathology. Post-operative period was uneventful. The saline bandage was removed on post-operative day 2.

Final histopathological findings revealed compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Hypercellular areas had spindle-shaped cells with wavy nuclei with the tapered end and ill-defined cytoplasm. Verocay bodies were also seen in the hypercellular areas. Hypocellular areas were showing hyalinized blood vessels (Fig. 4).

DISCUSSION

Benign schwannoma, also known as neurilemomas or neurinomas, is the tumors arising from nerve sheath. Common sites for schwannoma are the head and neck region, especially the lateral cervical region and mouth. Furthermore, intraorally, the tongue is a frequent site of the tumor to grow. Nasal cavity and paranasal sinuses are not common sites for the occurrence of this tumor [5]. Intranasal nerves, ophthalmic, and maxillary division of the trigeminal nerve and branches of an autonomic nervous system are the probable site for developing schwannoma in the nose and paranasal sinuses. The nerve giving rise to schwannoma may not be always identifiable. There seems to be no sex predilection and the tumor can occur at any age. Although among the previously reported seven cases of schwannomas arising solely from the maxillary sinus, five were reported in females and our patient is also a female. Schwannomas usually remain asymptomatic until they attain enormous size. Symptoms related to the site of involvement and also whether malignant transformation has occurred or not [6].

Our case was a 25-year-old female, presenting with a painful swelling below the right eye for 8 months which gradually increased in size. Lesions of maxillary sinus give rise to pain, whereas tumor of the nose and ethmoid sinus usually presents with epistaxis. Nasal blockage, rhinorrhea, hyposmia, exophthalmos, neuronal deficit, and facial swelling may be the other symptoms [7,8].

The gold standard for diagnosis of any lesion still remains to be the histopathological examination, which in this patient revealed compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Previous literature has mentioned about the typical biphasic pattern seen in schwannoma, consisting of Antoni type A and B areas [9]. Verocay bodies were also seen in the hypercellular areas. Verocay bodies were first described by Verocay in 1910 and are considered to be the diagnostic of schwannoma [10]. Verocay body is comprised a stacked arrangement of two rows of elongated palisading nuclei, alternating with cellular zones which are made up of cytoplasmic processes of Schwann cells (Table 1).

Differentiation between schwannomas and other similar looking lesions, for example, leiomyosarcomas and fibrosarcomas can be done by immunohistochemical demonstration of laminin in the tumor. The immunohistochemical staining using marker S-100 protein was positive. This is a neural crest marker antigen and it confirms the diagnosis of schwannoma [11].

There is a rare chance of the tumor turning into malignant one. Complete surgical removal of the tumor mass remains the treatment of choice and the choice of surgical procedure depends on the site and size of the tumor. In this patient, *en bloc* removal of the tumor was done by modified Weber Ferguson approach. The post-operative period was uneventful, and the patient was discharged with antibiotics and analgesics. No complications were reported at 1, 3, and 6-month follow-up.

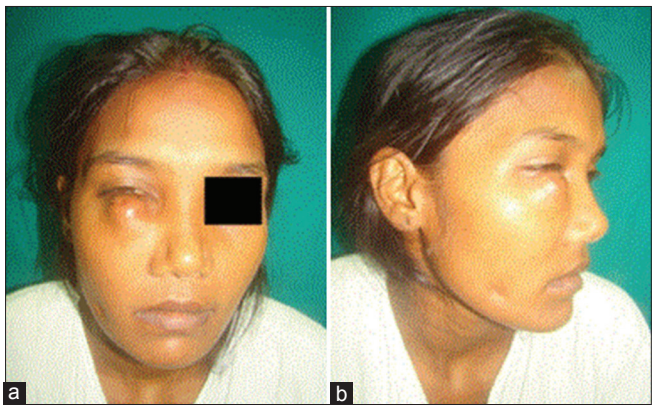

Figure 1: Swelling below right eye. (a) Front view, (b) lateral view

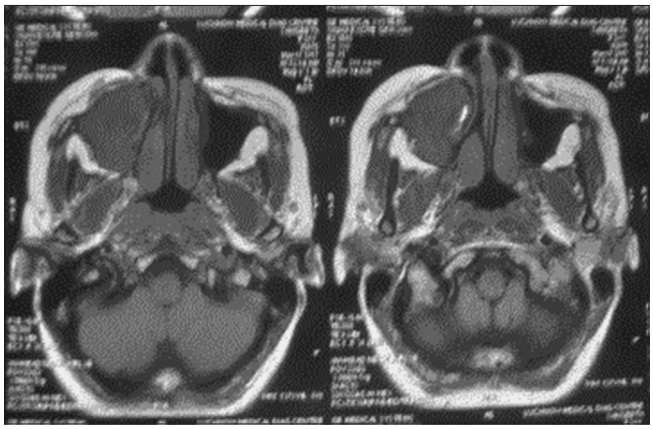

Figure 2: Magnetic resonance imaging showing solid lesion filling right maxillary sinus

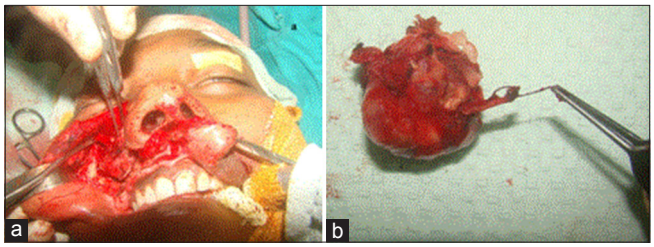

Figure 3: (a) Weber Ferguson approach, (b) excised specimen

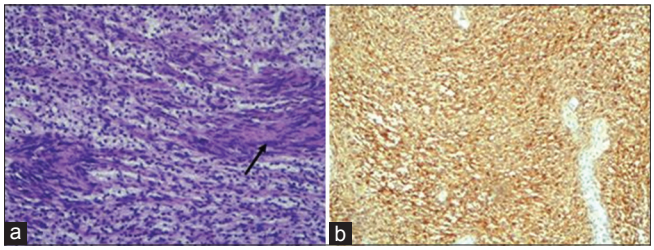

Figure 4: Schwannoma: (a) Showing both cellular areas with Verocay bodies (arrow) and myxoid hypocellular areas (H and E, ×100). (b) Diffuse positivity shown for S-100, ×100

Table 1: Review of previously reported cases[9]

Case	Age/sex	Symptoms	Lesion	Follow-up	Authors
1	60/F	Unavailable	Schwannoma (plexiform neurofibromatosis)	None	Stout (1935)
2	42/F	Pain	Malignant schwannoma	7 months	Millard and Bushar (1952)
3	24/F	Pain	Schwannoma	16 months	Debin *et al*. (1961)
4	38/F	Enlarging mass, exophthalmos	Schwannoma	12 months	Cross (1967)
5	45/M	Unavailable	Schwannoma (plexiform neurofibromatosis)	7 years	Robitale *et al*. (1975)
6	17/M	Proptosis	Schwannoma	6 months	Sanjay Khanna *et al*. (2003)
7	60/F	Swelling, fever	Schwannoma	3 months	Oshin Hegde (2016)
8	25/F	Pain, swelling	Schwannoma	6 months	Present authors

A successful *en bloc* removal of the tumor minimizes the chances of recurrence of the tumor. Since in our patient, the tumor was benign; therefore, there was no role of radiotherapy or chemotherapy [12]. Prognosis for benign tumor remains good, as in our patient, and she responded well to the treatment provided.

CONCLUSION

Schwannoma of the maxillary sinus is a rare clinical entity. Complete excision of the tumor is the treatment of choice for a benign schwannoma with good prognosis and with minimal chance of recurrence.

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