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Case Report

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Isolated schwannoma of maxillary sinus: a rare occurrence

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ABSTRACT

Schwannomas are benign tumors originating from the neural crests (schwann cells), which are cells that form the nerve sheath of peripheral nerve fibers. 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 report a case of an isolated maxillary schwannoma in a 45 years old lady who presented with swelling in the right cheek for 1 year and right sided nasal obstruction for 4 months. The swelling was not associated with epistaxis, fever, headache or visual disturbances. We report this case keeping in mind the rarity in occurrence of isolated maxillary schwannomas

Keywords: Schwannoma, Maxillary schwannoma, Isolated schwannoma

INTRODUCTION

Schwannomas are the tumors of the schwann cells, which form the nerve sheath of the peripheral nerve fibers and 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. Only 2% of the cases of schwannomas have been reported to be malignant, and recurrence occurs in cases of incomplete resection.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 Presented an isolated would be the ninth report of maxillary schwannoma.

CASE REPORT

A 45 year old female presented to the outpatient department of our institution with complaints of a swelling over the right cheek for 1 year, which was

gradually increasing in size and occasionally associated with pain. She also had gradually increasing nasal obstruction on the right for 4 months. The swelling was not associated with epistaxis, fever, headache or visual disturbances.

Clinical examination revealed a 4×3 cm smooth, firm, well defined, nontender swelling, extending from the right infraorbital region till the right sublabial region with loss of nasolabial fold. The swelling was not fixed to the skin and the overlying skin was normal. There was no sensory deficit over the right side of the face. Visual acuity was normal.

We considered a differential diagnosis of odontogenic tumors like ameloblastoma, fibro-osseous lesions such as fibrous dysplasia, soft tissue neoplasia and radicular cyst.

CT PNS showed a well defined, thin walled, expansile, non-enhancing lytic lesion measuring 29x27x22 mm arising superior to the roots of the first and second premolar teeth thinning the anterior and medial wall of

right maxillary sinus leaving the posterior maxillary antrum free.

With CT evidence of a benign lesion in the right maxillary sinus, in toto removal of the tumor was performed by Caldwell Luc approach. Sublabial incision was given from lateral incisor to second molar tooth and deepened up to the bone. Subperiosteal dissection was done. Intraoperatively, a smooth surfaced fleshy mass was found in the anterior wall of right maxilla extending to pyriform aperture and entering the maxillary antrum causing a bulge in the lateral wall of nasal cavity. There was erosion and thinning of anterior wall of the right maxilla. The tumor was excised along with a portion of the anterior wall of the right maxilla and mucosal lining of the maxillary sinus. Inferior medial maxillectomy was The excised specimen was sent for histopathological examination. Postoperative period was uneventful. The antral and nasal packs were removed on postoperative day 2. The patient was discharged with antibiotics and analgesics. No complications were reported at 1 and 3 month follow-up.

Histopathology revealed compact hypercellular antoni A areas and myxoid hypocellular antoni B areas. Hypercellular areas had spindle-shaped cells with wavy nuclei with a tapered end and ill-defined cytoplasm. Verocay bodies were also seen in the hypercellular areas. Hypocellular areas were showing hyalinized blood vessels.



Figure 1: Swelling over right cheek.



Figure 2: Mass seen in coronal section of CT PNS.

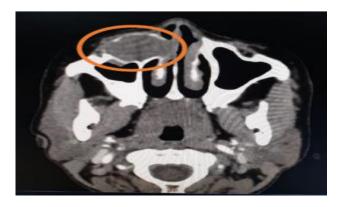


Figure 3: Mass seen in axial section of CT PNS.



Figure 4: Exposure of mass via sublabial incision.

DISCUSSION

Benign schwannoma, also known as neurilemmoma, is a tumor arising from schwann cells of peripheral nervous system or cranial nerves. Schwannomas usually involve the head and neck region, especially the lateral cervical region and mouth. Nasal cavity and paranasal sinuses are not 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 paranasal sinuses. The nerve giving rise to schwannoma may not be always identifiable. Schwannomas usually remain asymptomatic until they attain enormous size. Symptoms are related to the site of involvement and also depend on whether malignant transformation has occurred or not.⁵ Lesions of maxillary sinus give rise to pain and swelling, whereas tumors of the nose and ethmoid sinus usually present with epistaxis. Nasal blockage, rhinorrhea, hyposmia, exophthalmos, neuronal deficit, and facial swelling may be the other symptoms.⁶

The gold standard for diagnosis is histopathological examination which reveals compact hypercellular antoni A areas and myxoid hypocellular antoni B areas (Figure 5) with hypercellular areas showing verocay bodies, as in our patient (Figure 6). Previous literature has also mentioned the typical biphasic pattern consisting of antoni type A and B areas, seen in schwannoma.⁷

Verocay body is comprised of a stacked arrangement of two rows of elongated palisading nuclei, alternating with cellular zones which are made up of cytoplasmic processes of Schwann cells.⁸

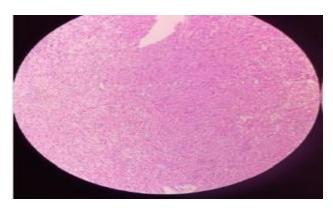


Figure 5: Compact hypercellular antoni A and myxoid hypocellular B areas (H and E stain).

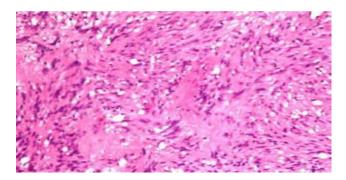


Figure 6: Verocay bodies seen in hypercellular area.



Figure 7: Postoperatively after 1 month.

Differentiation between schwannomas and other lesions like leiomyosarcomas and fibrosarcomas can be done by immunohistochemical staining using marker S-100 protein which is positive in a schwannoma. This is a neural crest marker antigen and it confirms the diagnosis of schwannoma. There is a rare chance of the tumor turning malignant. Complete surgical removal of the tumor mass remains the treatment of choice. The surgical procedure depends on the site and size of the tumor. In

this patient, in toto removal of the tumor was done by Caldwell-Luc approach. A successful removal of the entire tumor minimizes the chances of recurrence of the tumor. Since in our patient, the tumor was benign there was no role of radiotherapy or chemotherapy. ¹⁰ Prognosis for benign tumor remains good, as in our patient, and she responded well to the treatment provided.

CONCLUSION

An isolated 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 very minimal chance of recurrence.

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