Case Report

Maxillary schwannoma – A case report of a rare tumor

ABSTRACT

Schwannoma or neurilemmoma is a neurogenic tumor. Intraoral schwannoma is rare and intraosseous schwannoma involving maxilla is even rare. We present one such rare case of maxillary sinus schwannoma extending to the nasal cavity, infratemporal fossa in a 22-year-old male. Wide excision of the lesion with subtotal maxillectomy of the right side was done using Weber–Fergusson approach. The patient made a good postoperative recovery. The relevant literature on the presentation, radiology, and management of schwannoma of maxillary sinus is discussed.

Keywords: Central neural tumor, maxillary sinus, neurilemmoma, schwannoma, Weber–Fergusson approach

INTRODUCTION

Benign nerve cell tumors have been known as spindle cell tumors, schwannoma, neurofibroma, neurilemmoma, and neurinoma. Extracranial head-and-neck schwannomas are commonly seen as solitary and well-defined lesions. Schwannomas are benign tumors arising from Schwann cells that encase the axons of cranial nerves as they abscond the central nervous system. The etiology is unknown.[1] The lesion presents as nasal obstruction, dysphasia, and hoarseness, depending on the site of the lesion. The treatment is complete surgical excision of the benign tumor followed by postoperative histopathological examination to establish the final diagnosis. These tumors can occur anywhere in the body but relatively common (25%–45%) in the head-and-neck region, but the involvement of the sinonasal region is rare (4%). They are more commonly seen in the ethmoid sinus, followed by the maxillary sinus, nasal cavities, and sphenoidal sinus.[2] We present one such rare case encountered at our center with relevant literature.

CASE REPORT

A 22-year-old male presented to the department of oral and maxillofacial surgery with 6-month history of progressive swelling of the right side of the face, nasal obstruction, and bulging of the right eye for last 2 weeks. General and systemic examination was normal. On local examination was significant for a large swelling of the right maxilla extending from the right infraorbital margin, temporal region, to the corner of the mouth. Swelling was noncompressible and nonfluctuant. There was a grayish white firm mass in the right nostril which was pushing nasal septum to the left side. Ophthalmic evaluation showed slight proptosis of the right eye, bilateral normal vision, normal visual field, and normal eye movements. There was no significant cervical lymphadenopathy [Figure 1].

Intraoral examination [Figure 2] demonstrated compressible swelling of the right maxillary buccogingival sulcus with expansion of alveolar ridge and hard palate. Overlying mucosa was smooth. All the three maxillary molars on the right side were Grade 3 mobile with intrusion on pressure. Computed

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tomography (CT) scan of nose and paranasal sinuses [Figure 3] showed destructive lesion centered within maxilla. There was erosion of anterior, lateral, and posterior maxillary sinus wall with extension into pterygoid musculature and nasal cavity. The orbital floor was thin but appeared intact without involvement of orbital cavity. Chest radiograph showed no evidence of metastasis. The final clinical and radiological staging of this tumor was $T_4N_0M_0$. An intraoral incisional biopsy by Caldwell–Luc approach was performed and microscopic examination revealed spindle cell carcinoma, a nerve cell tumor.

Subtotal maxillectomy under general anesthesia was planned by Weber–Ferguson's incision that includes lateral rhinotomy incision with horizontal infraorbital component and midline lip split. Incision was marked and temporary tarsorrhaphy on the right eye was performed for eye protection. Sublabial incision was performed after splitting upper lip in midline. This facilitates the elevation of flap from anterior wall of maxilla extending through the entire buccogingival sulcus up to maxillary tuberosity. Infraorbital horizontal component of Weber–Ferguson's incision was given 1 mm below infraorbital rim. Layer by layer dissection was performed and flap elevation done. Tumor was exposed and bony cuts were marked. Lateral incisor was removed. Bony cut was made using surgical handpiece. Cut was extended palatally and the remaining cut was made using 3-mm chisel and mallet [Figure 4]. Right maxillary bone was removed by preserving uninvolved zygomatic bone and infraorbital rim and encapsulated tumor was excised [Figure 5]. Defect was packed by Gauze pack intraorally and closure done [Figure 6]. Postoperative recovery was uneventful and the patient was discharged on the 3rd postoperative day. Histopathological examination reveals capsulated mass with biphasic pattern. Nodule was covered with mucosa. The presence of spindle cells was seen forming verocay bodies. Antoni A and B areas were clearly identified. There was positive staining for S-100 protein [Figure 7]. Hence, final histopathological report confirmed the diagnosis as schwannoma. Prosthetic functional rehabilitation was done with obturator [Figure 8].

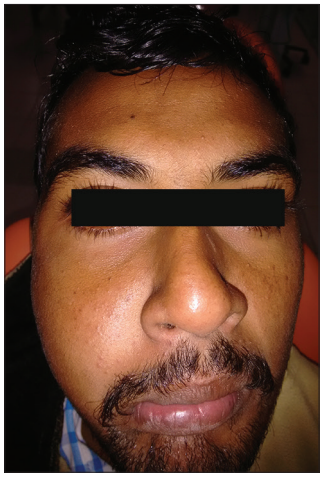

Figure 1: Extraoral swelling involving the right side of the face

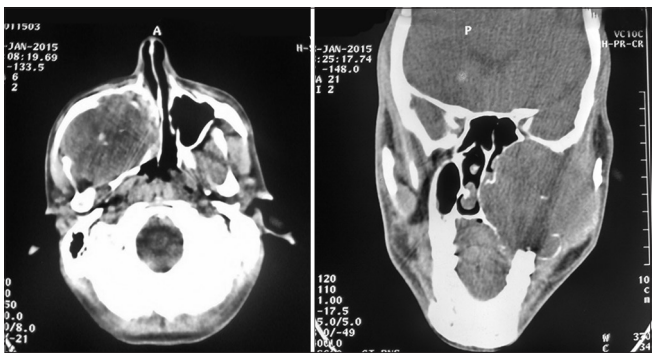

Figure 3: Axial and coronal computed tomography scan

DISCUSSION

Schwannoma also called as Schwann's cell tumor or

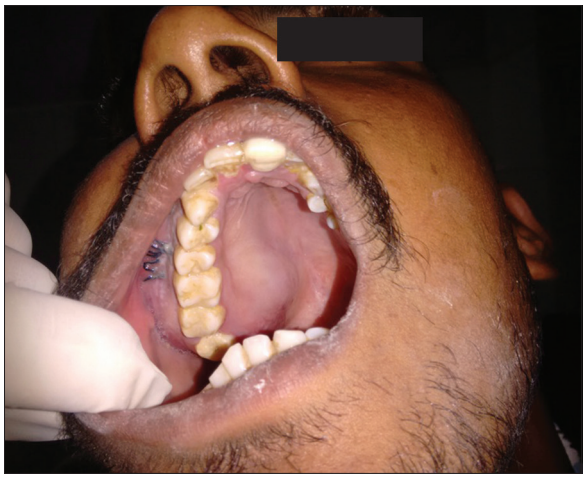

Figure 2: Intraoral noticeable swelling on the buccal vestibule and palate on the right side

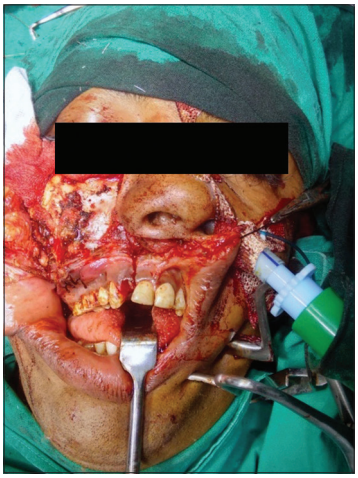

Figure 4: Intraoperative picture showing flap elevation after Weber–Ferguson incision and exposure of tumor

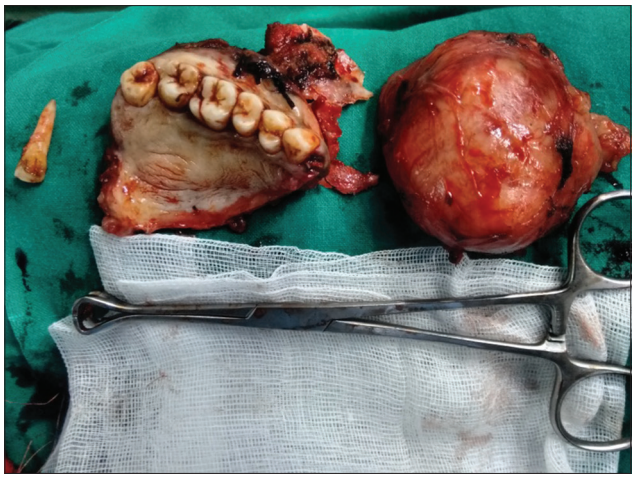

Figure 5: Whole specimen

Figure 6: Immediate postoperative

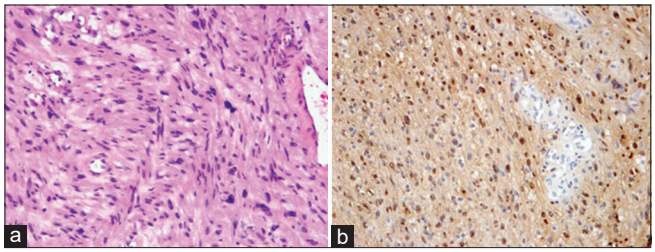

Figure 7: (a) Antoni A and B areas with ill-defined areas of nuclear palisading (H and E × 200 and (b) positive staining for S100 protein-S-100 protein immunoperoxidase avidin–biotin complex technique

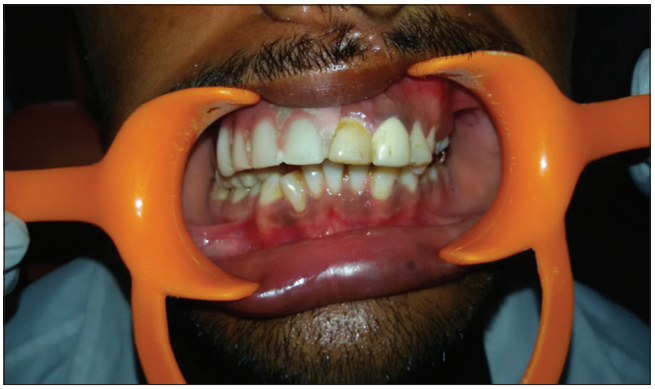

Figure 8: Maxillary obturator in functional position

neurilemmoma is a benign tumor of neuroectodermal origin arising from the Schwann cells of the neural sheath. It affects the peripheral nervous system, the cranial nerves (except the optic and the olfactory), the spinal nerves, and the autonomic nervous system, whereas Schwann cells enclose the neurons and their axons.[3] Schwannoma seldom occurs in the oral cavity. Wright and Jackson described 146 cases of soft-tissue intraoral schwannoma, of which 52% occurred in the tongue.[4] Intraosseous schwannomas are even atypical with most common site at posterior mandible and found in less than 0.2%. This may be due to the presence of large inferior alveolar nerve.[5,6] Chi *et al*. described 44 cases of intraosseous neurilemmoma, of which only 5 presented in maxilla.[7] The etiology is anonymous, but it is said that the lesion arises by the propagation of Schwann cells at one point within the perineurium. The growth of this lesion will cause the displacement of adjacent nerve tissue. Possible mechanisms through which schwannomas may involve a bone include a tumor arising centrally inside the bone, a tumor arising in a nutrient canal, or a soft tissue or periosteal tumor causing secondary erosion and incursion into the bone.[8] In our case, posterior superior alveolar nerve might be involved. The schwannoma is generally a single lesion, but when it is allied with neurofibromatosis, it can be multiple too. This tumor can occur at any age with prevalence in females more than males at a ratio of 2:1.[9] Clinical features of schwannoma of paranasal air sinuses depend on site and affected nerve and compression of the surrounding neural structure. Epistaxis and pain may be seen due to the involvement of ethmoidal sinus and maxillary sinus, respectively.[10] Other clinical symptoms such as swelling, numbness, nasal obstruction, exophthalmos, mucopurulent rhinorrhea, and hypo or anosmia are also observed. There is no classical radiological sign of this tumor. It is generally presented as unilocular radiolucency with well-defined sclerotic border on routine extraoral radiographs. On CT scan, the characteristic lesion is a well-defined mass associated with thinning of the surrounding bone by tumor. CT scan is useful in evaluating the size, location, and erosion of bony surfaces. Differential diagnosis of schwannoma of the paranasal sinuses should include polyp, ossifying fibroma, ameloblastoma, dentigerous cyst, solitary bone cyst, spindle cell carcinoma of any type, and other mesenchymal tumors.[11] The likelihood of malignant alteration of a schwannoma is unusual. There are only 12 reported cases of malignant schwannoma of the paranasal sinuses in literature.[12] Histologically tumor represents elongated (spindle like) cells with asymmetrical

nuclei lying amid bundles of collagen fibers. Two discrete patterns known as Antoni Type A and Type B may be observed. Antoni Type A is characterized by Schwann cells that are closely packed, forming bundles, or arranged in rows with elongated, palisading nuclei. Free bands of amorphous substance between the rows of nuclei constitute the Verocay bodies which appear to be composed of thin cytoplasmic processes with small amounts of collagen and basal laminar material. Antoni Type B is a mixture of Schwann cells, fibroblasts, and nerve fibers that are widely separated, dispersed in a loose and random fashion with a network of delicate reticulin fibers, and numerous microcystic spaces. The ground substance is myxoid. These tumor cells typically show a diffuse positive immunoreactivity for S-100 protein. Virtually, all schwannomas are encapsulated. Schwannoma are defiant to radiotherapy and the treatment of choice is surgical enucleation or excision with regular follow-up. Recurrence of tumor is also rare if surgical enucleation is performed efficiently.[13]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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