

Maxillary Schwannoma – A Rare Case Report

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ABSTRACT

Background: Schwannoma or neurilemmoma are a type of nerve sheath tumors. Most of the literature, reports of schwannoma in the head and neck involve tongue. Intraosseous schwannomas account to about less than 1% with affliction to the mandible over maxilla. Very few cases of maxillary schwannomas have been reported till date.

Case presentation: We present one such rare case of schwannoma involving the maxilla extending till the nasal cavity in a 35 years old male. Wide excision of the lesion with hemi-maxillectomy of the right side was done using modified Weber–Fergusson approach (Dieffenbach's extension). The patient's postoperative recovery was uneventful without recurrence when followed up till 2 years.

Conclusion: Though schwannoma of the Head and neck region are rare, they should be considered as a differential diagnosis of unilateral slow growing mass in the head and neck region, particularly in an adult.

Keywords: Schwannoma, Neurilemmoma, Neurogenic tumors, Weber–Fergusson approach and Hemi-maxillectomy
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INTRODUCTION

Schwannomas or neurilemmomas are defined as rare well circumscribed and encapsulated neoplasm arising within the nerve sheath.¹ It was first described by Verocay in 1910, who named it as "Neurinoma". Later, Stout, termed it as "Neurilemmoma".¹ The etiology is unknown, but it has been postulated that the lesion arises due to the proliferation of Schwann cells of the perineurium. This neoplasm has predilection for the head and neck region, with tongue being the most common intraoral site. However, intraoral lesions of the maxilla and mandible are relatively rare.^{2,3} The tumor generally presents as a slow growing painless swelling, and extensive lesions may cause dysphasia, nasal obstruction, and hoarseness, based on the site of the lesion. The treatment includes complete surgical excision of the benign tumor followed by histopathological examination for conclusive diagnosis.² In the current case report, we present a case of Schwannoma in the maxilla in a 35-year-old male, which was treated hemi maxillectomy.

CASE REPORT

A 35 years old male reported presented to the department of oral and maxillofacial surgery with 2 years history of progressive swelling of the right side of the face, leading to gross facial asymmetry. General medical examination was normal. On local examination there was a swelling on the middle third of the face on the right side extending 3 cm away from the right infraorbital margin, to the corner of the mouth. Swelling was non tender with no pus discharge, paresthesia or surface ulcerations. The skin over the swelling skin appeared normal, with no surface rise in temperature and cervical lymphadenopathy. On intraoral examination, there was a well-defined swelling extending from 11 to

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16 measuring approximately 3cm × 4cm antero-posteriorly and from the depth of the vestibule to the alveolus, superior-inferiorly. There was a remarkable vestibular obliteration. On palpation the swelling was non-tender, firm in consistency, non-compressible, non-fluctuant and non-pulsatile. With no evidential dental pathologies in the same region [Figure. 1]. At the level of maxillary sinus a bony window was visible in axial section of computed tomography. Large isodense expansile lesion could be appreciated extending from the central incisors to the first molar area. Perforation of buccal cortical plate was seen with bone expansion on 3D reconstruction [Figure 2]. Based on the clinical examination and radiographic features, a differential diagnosis of benign neoplastic lesions of maxilla such as ameloblastoma or central giant cell granuloma was established.

Hemi maxillectomy under general anesthesia was planned by modified Weber–Ferguson’s incision (Dieffenbach’s extension). Initially, the patient’s eyelids were closed using Temporary tarsorrhaphy sutures. The outline of the approach was highlighted using a marker, starting from lower border of eye until 2mm from the medial canthus, extending downward along the lateral border of nose until midpoint of philtrum of the upper lip [Figure.3]. Layer by layer dissection and flap elevation was done. Incision was then given medially across the posterior end of the hard palate further turning 90 degrees anteriorly crossing the midline till canine of opposite side. Cheek flap was reflected to expose the lesion, tumor was exposed and bony cuts were marked [Figure 3]. Palatal bone was divided using a saw blade and bur after elevation of gingival and palatal mucosa. The basal bone was separated from the frontal of maxilla using an osteotome. The orbicularis oculi muscle was retracted superiorly and the bone was cut across the maxilla just below the inferior orbital rim. The entire specimen of hemi-maxillectomy [Figure 4] was removed. The remaining attachments were severed using a large curved scissor placing behind the maxilla. Excessive bleeding was controlled with electrocautery, all the sharp bony projections were trimmed off. Defect was packed by Gauze pack intraorally and closure was done. Postoperative recovery was uneventful and the patient was discharged on the 3rd postoperative day [Figure 5]. Histopathological examination revealed a well capsulated mass with a typical biphasic pattern showing both Antoni A and B cells. [Figure 6] Immunohistochemistry was also done using S-100 protein marker giving an intense diffuse immune staining thereby giving a clear-cut diagnosis of schwannoma. Hence, a confirmatory histopathological diagnosis of schwannoma was given. Patient was followed up for about 2 years and there was no recurrence.

Schwannomas are also known as neurilemmomas, neurinomas, neuromas and peripheral fibroblastomas. They are rare, benign tumors arising from the sheath of myelinated nerve fibers that may occur in any part of the body.¹ These lesions arise from proliferation of Schwann cells at a point inside the perineurium, that causes a displacement and compression of the surrounding normal nerve tissue and trauma and neurofibromatosis type 2 are well-recognized risk factors for these lesions.^{4,5} Two types of schwannomas have been identified: a) Peripheral: located in the soft tissues and b) Central (intraosseous): located within the bone.^[6] Being neural in origin, majority of the schwannomas are peripheral tumors; approximately 45% of them affecting the soft tissues of the head and neck.^{6,7} Schwannomas have a predilection for the head, neck and flexor surface of the upper and lower extremities (Hatziotis & Asprides 1967, Zachariades et al. 1987), Intraorally, tongue being the most common site, followed by buccal mucosa, floor of the mouth, palate and lip.^{8,9} Schwannomas are considered to be uncommon in head and neck region with less than 1% of occurrence. Mandible is the most favored site; the reason being attributed to the large caliber of the inferior alveolar nerve and its long course within the jaw.⁷ They are presumed to involve bone by one of the three mechanisms: a) arising centrally within bone b) arising within the nutrient canal and causing its enlargement c) arising from the periosteum and invading into the bone by secondary erosion.^{10,11}

Clinically schwannoma presents as a solitary circumscribed nodule, rarely being multi-nodular.¹² Most reported cases describe small, slow growing tumors that are asymptomatic, although pain and paresthesia may occur. Other symptoms include disturbance in mastication, phonation, dyspnea or dysphagia, depending on the location of tumor.¹³ Analysis of the symptomatology revealed

DISCUSSION



Fig. 1: Intra oral picture of the swelling from 11-16 region

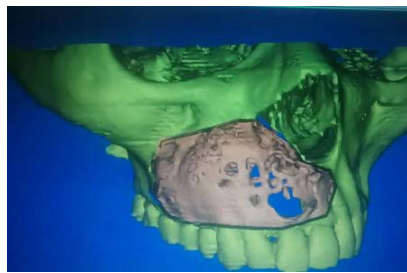


Fig. 2: 3 D reconstructed image of CT scan showing tumor mass.



Fig. 3: Surgical excision of the lesion.



Fig. 4: Gross Specimen of the excised lesion.



Fig. 5: Post-operative picture after 2 years follow-up.

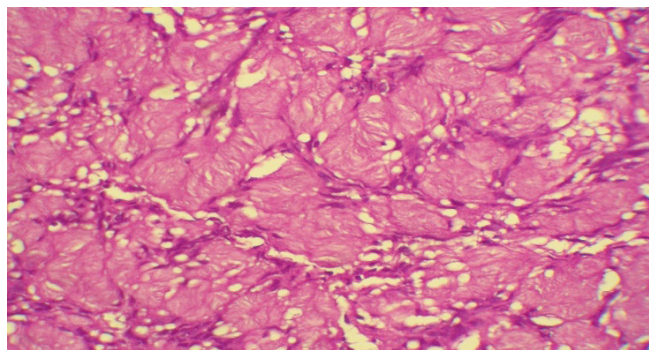


Fig. 6: Antoni A and B areas with ill-defined areas of nuclear palisading (H and E x200)

that swelling was the most common complaint of the patient during their initial visit. The duration of the swelling ranged from three months to 20 years. Other symptoms like dysphagia and garbled speech were reported by few patients.^{14,15} Schwannomas can be an incidental finding.¹⁶ Based on the review of the literature, maxillary schwannomas have equal predilection for anterior and posterior segments of the jaw unlike their mandibular counterparts that have greater affliction towards posterior part of the ramus of mandible.¹⁷ Maxillary schwannomas affect younger individuals; often in the second decade of life whereas mandibular tumors are common in older age group. Similar to mandibular schwannomas, maxillary ones have a definite female predilection with a ratio of 1.6:1.¹⁷ Regarding the site of affection, the tumors showed slightly increased predilection for the hard palate.¹⁷

Schwannomas present radiographically as radiolucent lesions with thin sclerotic borders suggesting neoplastic bone destruction.^{18,19} Mandibular schwannomas may be unilocular or multilocular lesions while maxillary schwannomas are unilocular in most of the cases.²⁰ Maxillary schwannomas are often in close proximity to root apices with likely root resorption, dystrophic calcifications and alveolar bone resorption occasionally.^{18,21} This expansive pattern of growth tends to disrupt involved nerves and teeth (Marzola et al. 1988, Llewelyn et al 1989, Takeda 1991, Villaneuva et al. 1995). In CT, the tumor presents as a well-defined, non-enhancing, low density soft tissue mass without soft tissue infiltration.^{14,22} It is difficult to make definitive diagnosis solely using radiographic interpretation, a biopsy is usually necessary.

Histologically, the schwannomas are encapsulated with a fibrous connective tissue capsule and present a distinctive feature which is presence of a biphasic pattern consisting of Antoni Type-A and Antoni Type-B cells. Antoni Type-A: spindle shaped cells with long, slender fibers forming a parallel arrangement of nuclei, known as palisading arrangement of the cells. Antoni Type-B: degenerative in nature with myxoid changes seen in connective tissue stroma, the cells and fibers run in haphazard manner, the entire tissue is loose in texture present due to cystic degeneration. The central schwannomas have a predominance of poorly organized cells with prominent, thickened blood vessels.²¹

Immunohistochemistry staining using marker S-100 protein, which is a neural crest marker antigen, gives an intense diffuse immune staining, it is expressed exclusively by Schwann cells thus a gold standard for diagnosis of schwannoma. Apart from S-100 protein, perineural cell derived tumor capsules which are immunoreactive to EMA (Epithelial Membrane Antigen), collagen type IV and laminin are also of diagnostic value. Capsular and Antoni-B cells are positive for CD34 and CD68 lysosome associated antigens.

Differential diagnosis of schwannoma can be given as, giants cell tumors, lipoma, fibroma, traumatic neuroma, and cysts.^{23,24} Investigations such as FNAC aids in excluding odontogenic cysts, fibromatous gingiva²⁵, odontogenic tumors such as ameloblastoma, can be differentiated with the absence of egg shell crackling or association with unerupted tooth.

Neurofibroma also originates from Schwann cells and must be considered as a differential diagnosis during histopathologic examination, since it possesses a risk of transforming into malignancy unlike schwannomas which are mostly benign.²⁶

Treatment of choice of benign schwannoma in the head and neck region has been surgical excision, with a very low reported rate of morbidity and recurrence.²⁷ The surgical approach depends on location, extent and the close proximity of tumor to surrounding

vital structures. Management of malignant schwannoma involves radical resection, post-operative radiotherapy and chemotherapy aren't of great value. Prognosis for malignant schwannoma is poor with a reported 5-year survival rate ranging between 30% and 65% contrasting to benign schwannoma which has excellent prognosis and low recurrence rate.

CONCLUSION

Though schwannoma of the Head and neck region are rare, they should be considered as a differential diagnosis of unilateral slow growing mass in the head and neck region, particularly in an adult. Intraosseous lesions located in the peripheral region with non-specific clinical and radiographic features often lead to diagnostic confusion. Most of the lesions located in the environs of the teeth are odontogenic in origin. Yet, other possibilities like intraosseous schwannomas must be considered.

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