INTRA OSSEOUS SCHWANNOMA - A CASE REPORT

Gadiputi S.*, Sumalatha M.N.**

Schwannoma of the head and neck region is relatively uncommon, histologically distinctive, benign, encapsulated peripheral tumor of Schwann cell origin. Intraosseous schwannomas are rare and represent less than 1% of all benign primary bone tumors. They pose difficulty in preoperative diagnosis but most of the cases are confirmed by histopathology and immunohistochemistry.

Key words: Schwannoma, Antoni-A, Antoni-B, Petragus, Neurilemmoma.

Neurilemmoma was first described by verocay in 1910 and he called it as neurinoma. In 1935 the term Nurilemmoma was coined by Stout[1]. Schwannomas arise when proliferating shwann cells in the perineurium causing displacement and compression of peripheral, motor, sensory, sympathetic and cranial nerve sheath. Only 50% of them have a direct relation with a nerve[2]. They may occur at any age but the peak incidence is 3rd and 4th decades with no sex predeliction. 25% to 45% occur in the head and neck only 1% intraorally with rare intraosseous presentation[2]. Most of them are solitary and asymptomatic with fewer rates of recurrence and malignant transformation[3]. Histologically it is composed primarily of Schwann cells in a poorly collaginized stroma[1]. The treatment of choice is surgical excision with good prognosis and rare secondary complications[4]. A rare case of an intraosseous schwannoma in the right pretragus area in a 48 year old female is presented.

CASE REPORT

A 48 year old moderately built and nourished feamle reported with the chief complaint of a swelling on right side of the cheek for 3 years. History revealed gradually enlarging swelling with pain and paresthesia. No relevant medical history was revealed. Extraorally swelling was approximately 1x3cms extending from pretregus to innercanthus of the eye antero posteriorly with depressed notch and scar above the prominent zygomatic bone. Intraorally swelling was approximately 2x3cms with ulceration and erythema in the retromolar and faucial pillar region secondary to occlusal trauma. Upon palpation it was soft to firm in consistency with pain. A provisional diagnosis of salivary gland neoplasm was made and lipoma, schwannoma, fibroma were considered as an alternative diagnosis. Radiographically Ortho Pantamo Gram revealed a well defined unilocular radiolucency with sclerotic margins involviving right ramus of the mandible, extending superiorly upto zygomatic bone causing resorption and fracture of the zygomatic bone and lateral wall of the maxillary sinus with complete obliteration of the pterygoid plates. Coronal and axial CT scan revealed a partially enhancing space occupying lesion of about 3.58x4.84cms in size in the right masticatory space. The incision biopsy was performed under local anesthesia. Grossly, cut surface of the specimen was soft greywhite with light brown and few yellowish foci were seen. The hematoxylin and eosin stained section showed fragments of tissue composed of spindle cells with hypercellular and edematous hypocellular areas. Hypercellular areas showed palisading and regimentation of nuclei with abundant fibrillary matrix. Hypocellualr areas showed admixed chronic inflammation, foam cells and hemosiderin deposits. No atypia or increased mitotic activity is seen From histopathologic features a diagnosis of ***intra osseous schwannoma was made.***

DISCUSSION

Peripheral nerve tumors of the oral cavity include schwannoma, neurofibroma, traumatic neuroma, nerve sheath myxoma, palisaded encapsulated neurinoma

**Professor*

***Senior lecturer, Department of oral pathology and microbiology, Kothiwal Dental College and Research Centre, Kanth Road, Moramustaqueem, Moradabad-244001, Uttar Pradesh*

and granular cell tumor[3,5]. Embryologically, Schwann cells arise during the 4th week of IUL from a specialized population of ectomesenchymal cells of the neural crest, which then detach from the neural tube and migrate into the embryo[2]. Schwann cells form a thin barrier around each extracranial nerve fiber and wrap larger fibers with an insulating membrane, myelin sheath to enhance nerve conductance[2]. As nerve exit the brain and spinal cord there is a change between myelination by oligodendrocytes to myelination by Schwann cells[2].

The Schwannoma is a rare benign neural tumor arising from the neural sheath Schwann cells of the peripheral, cranial or autonomic nerves[1]. It does not arise from cranial nerves I and II because they lack Schwann cells[1]. It is believed to originate from a proliferation of Schwann cells in the perineurium causing displacement and compression of the adjacent nerve[1].

It can occur at any age but more common between the ages of 20 and 50years. Tumors in the oral cavity are often seen in the 2nd and 3rd decades of life but are very rare in those aged 10years younger[3,6]. Chi et al has reported schwannoma of the jaw occurred in the age range of 8-72 years, with the average age of 64 years with definite female predilection[1]. The present case was of female aged 48 years which corelate with above findings.

Extracranially 25% of the reported cases are from the head and neck region, only 1% intraorally being found principally in the tongue, more frequently in the anterior portion[3]. Among the jaws, the mandible is a favoured site compared to the maxilla. Mandibular Schwannomas are mostly localized to the posterior body and the ascending ramus and can involve symphysis region[1]. ***However the location in the pretragus region involving zygimatic bone, ramus and lateral wall of the maxillary sinus is a rare finding could not be linked to any such case mentioned in the English literature.*** Schwannomas involve bone by any of the three mechanisms a tumor may arise centrally within the bone, within a nutrient canal and produce canal enlargements, or soft tissue\ periosteal tumor may cause secondary erosion and penetration into a bone[2]. Clinically it presents as a solitary, smooth surface, asymptomatic growth; although symptoms depends on nerve of origin[1]. The present case was associated with pain, paresthesia and ulceration. Cliniclally Oral Schwannomas are of two types, the common type is the submucosal nodule, which is encapsulated, well defined, firm in consistency, thus resembling a cyst. The second type is the non-encapsulated where the tumor is found below the basal layer of mucous membrane[7].

The preoperative diagnosis is quite difficult because this is an infrequent tumor and is not usually suspected in the oral cavity[3]. Diagnostic investigations include ultrasound scanning, computed tomography, magnetic resonance imaging and fine needle aspiration cytology.

Radiographically, Schwannomas of either jaw are well-demarcated, unilocular radiolucencies with a thin sclerotic border with additional features such as external root resorption, cortical thinning, cortical thickening and expansion[1]. The findings of present case are coroborating with above. The schwannoma typically a well –covered and well defined fusiform tumor on CT scans[9]. The present case revealed a partially enhancing space occupying lesion of about 3.58x4.84cms in size in the right masticatory space. However, MRI is best choice in showing the extent of tumors and correlates well with operative findings[4].Generally, ultrasonic images of Schwannoma are charecterized by a round or elliptical cross section with a clear border, cystic changes may be seen in some parts and the tumor which tends to be hypoechoic. If the diameter of the nerve origin is large, it can be observed that the mass becomes fusiform and is connected to the nerve[9].Yamazaki et al reported that ultrasound and magnetic resonance imaging were effective in preoperative diagnosis of schwannoma originating in the mental nerve. On MRI, a tumor can appear as solid, cystic \mixed, based on its pathologic charcteristics[2,9].

The definitive diagnosis is based on histopathology with the characteristic fibrous

encapsulation cosisting of epineurium and residual nerve fibers. The hallmark of Schwannoma is the presence of alternating patterns of **Antoni -A** and **Antoni -B** areas. The **Antoni-A** areas are composed of compact spindle cells that usually have twisted nuclei, indistinct cytoplasmic borders and occasionally clear intranuclear vacuoles. They are arranged in short bundles or interlacing fascicles. **Antoni –B** areas are far less orderly and less cellular. The spindle or oval cells are arranged hapazardly in the loosely textured matrix, which is punctuated by microcystic change, inflammatory cells, and delicate collagen fibers[1].

The immunohistochemical tests reveal the Schwannoma cells to be positive for the protein s-100, a marker for the nervous system[3,4]. For the differential diagnosis, neurofibroma, granular cell tumors, liopma, fibroma, leiomyoma, rhabdomyoma, adenoma, neuroma should be considered[2]. Schwannomas are treated by complete surgical excision but wide excision is not recommended because schwannomas show few recurrences after surgery[4]. The prognosis is very good as it does not usually recur. Maliganant transformation is exceedingly rare and no such transformation has been reported for intra-osseous neurilemmoma[1,7].

CONCLUSION

The nerve sheath neoplasms are more common that pose problem in preoperative diagnosis and the Schwannomas must be included in the differential diagnosis of asymptomatic massacs in the head and neck. Prognosis is excellent as the tumor is benign and recurrence is nearly unknown, so it is possible and indeed recommended to preserve nerve integrity with careful dissection.

Refernces

1. Parikh RN, Desai N. Intra-oral schwannoma (neurilemmoma): An unusual anterior palatal swelling - A case report. J Int oral health 2010; vol. 2; 87-91.
2. Pfeifle R, Baur AD, Paulino A et al. Schwannoma of the tongue: report of two cases. J oral Maxillofac surg 2001; 59: 802-804.
3. Carriches CL, Bryan RP, Montero SM: Schwannoma located in the palate: clinical case and literature review. Med oral pathol oral cir Bucal 2009; 9:e465-8.
4. Naidu SG, Sinha MS: Schwannoma of the tongue: An unusual presentation in a child. Indian J Dent Res 2010; 21(3): 457-59.
5. Baranovic M, Macan D, Begovic EA et al: Schwannoma with secondary erosion of mandible: Case report with a review of literature. Dentomaxillofac Radiol 2006; 35: 456-460.
6. Kowatsch E, Feichtinger, Zemann W et al. Intra-osseous schwannoma of the mental nerve clinically stimulating intraosseous. J oral pathol med 2006; 35:517 9.
7. Nakasato T, Kamada Y, Ehara S et al. Multilocular neurilemmoma of the tongue in a child. Am J Neuroradiol 2005; 26: 421-423.
8. Subhashraj K, Balanand S, Pajaniammalle S. Ancient schwannoma arising from the mental nerve. A case report and review. Med oral patol oral bucal 2009; 1: e12-4.
9. Yamazaki H, Kaneko A, Ota Y, Tsukinoki K. Schwannoma of the mental nerve: usefulness of preoperative imaging. A case report. Oral surg oral med oral pathol oral radiol Endod 2004; 97:122-6

Discourage
&
Condemn
Plagiarism



Fig.1: Extra photograph showing ill defined swelling on zygomatic area with scar.

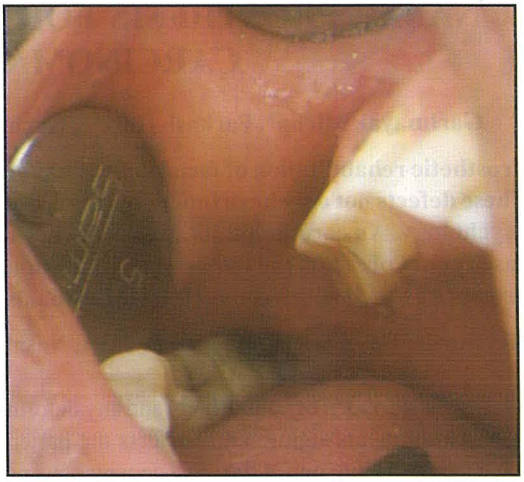


Fig. 2: Intra oral photograph showing swelling in retromloar area with ulceration.

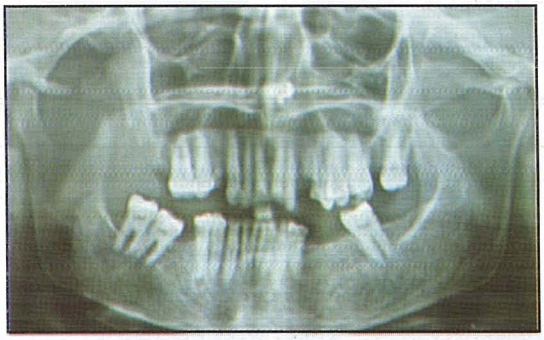


Fig. 3: Orthopantomograph showing radiolucent area in right ramus of mandible.



Fig. 4: Computed tomograph showing welldefined, hyperdense area in masticator space.

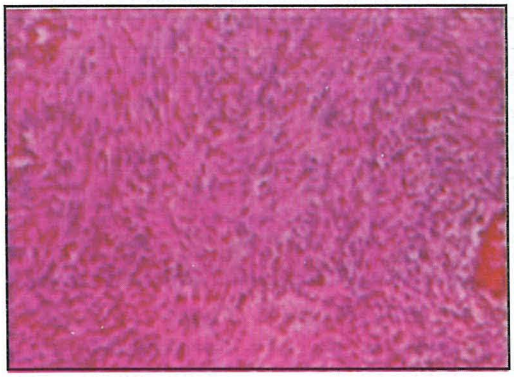


Fig. 5: Showing Antoni type-A and Antoni type-B tissue with palisaded arrangement of nucleus.