Ancient Schwanomma of Right Maxilla - a rare Case Report

Ayesha Niyaz[1], S. Padmashree[2], Rema J[3], Reshma D[1]

[1]Consultant, Oral Medicine & Radiology, Bangalore.

[2]Professor and Head, Department Oral Medicine and Radiology, Vydehi Institute of Dental Sciences, Bangalore.

[3]Professor and Head, Department Oral Medicine and Radiology, Al Azhar Dental College. Ker

Corresponding author

Dr. Ayesha Niyaz MDS, Consultant, Oral Medicine & Radiology, Bangalore, Karnataka, India.
Email: ayesha.n.94@gmail.com

Abstract

Intraoral Schwanomma, also known as neurilemmoma or neurinoma is a rare benign tumor of peripheral nerve sheath Schwann cells, of these intraosseous types are rare and rarest of all are maxillary schwannomas. They arise from the nerve sheath Schwann cells of the peripheral, cranial or autonomic nerves. Although it originates from nervous tissue, only in 50% of the cases a direct relation with a nerve is demonstrated. Frequently located in the soft tissues of head and neck region, but only 1 to 12% of them are located in the oral cavity. Frequent locations are mobile portion of the tongue, palate, cheek mucosa, lip and gingiva. Less than 1% is reported intraosseous in the jaws. Only 14 cases of maxillary schwanommas have been documented in the literature so far. The tumor is generally asymptomatic. Diagnosis is based on thorough detailed history, imaging and microscopy confirmed by Immunohistochemistry. Schwanommas are managed by complete surgical excision, but wide excision is not recommended because it rarely shows recurrence after surgery and malignant transformation. This paper highlights a rare case of Schwanomma of right maxilla in a 25 year old female patient who presented with a slowly enlarging asymptomatic swelling and explores the diagnosis and investigation by immunohistochemistry.

Key words: Schwanomma, Benign encapsulated neural tumor, Oral cavity, Immunohistochemistry.

Introduction

Schwannoma or Neurilemmoma is a benign tumor of the neural sheath derived from the Schwann cells. It was first described in the literature in the year 1910 by Verocay, who described the first peripheral nerve tumor. Nevertheless, it was not until 1932 that the term "Schwannoma" was introduced by Masson.[1] This neoplasm is composed primarily of Schwann cells in a poorly collagenized stroma. Etiology is unknown, but it is postulated that the lesion arises inside the perineurium by the proliferation of Schwann cells. The growth of this lesion usually causes the displacement and compression of the surrounding normal nerve tissue.[Vc]

Schwannomas most often occur in the fourth and fifth decade of life and have a 2:1 female predilection over males. Most of the intraoral cases are found on the upper and lower side of the tongue and vestibule, being rare on the palate. This tumor most commonly arise in the soft tissues but it can be also found in hard tissues.[2] Among the jaws, schwannomas are common in mandible but are reported rare in maxilla.[1,3]

Clinically, a circumscribed slow-growing, firm and solitary tumor is normally observed. Most of the cases are asymptomatic and have a long duration and large size because of their lack of symptoms and slow growth.3 In cases where intraosseous localization is found pain and paresthesia may usually be present, also in this type there is frequently expansion of the affected bone along with swelling.[2]

Ancient schwannoma is another rare variant of intraoral schwannomas. The ancient variant presents with clear areas of hypocellular tissues and the changes presented are usually attributed to long standing degenerative changes like cystic degeneration, calcification, hemorrhage, hyalinization and macrophages and hemosiderin deposition. Nuclear atypiamay also be one of the prominent feature of this type of variant.[3]

We hereby in this article report a rare case of intraosseous ancient schwannoma of the maxilla with clinical, diagnostic and histological characteristics of this pathology.

Case Report

A 25 year old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of gradually progressing swelling on right side of face since 2 years. The swelling was associated with pain on applying pressure since 3 months. There was no history of trauma to the region and the swelling was not associated with pus discharge or numbness or any visual disturbances. Patient was short statured for her age and had not attained puberty. The patient's medical, family and past dental history were non-contributory. There was mild proptosis of right eye but no visual disturbances or double vision was reported.

On extra oral examination, a swelling of approx. size 5x6cms was present on right upper and middle 3rd of face (Figure1) extending anteriorly from ala of the nose to posteriorly 2cm in front of tragus, superiorly from temple of head to inferiorly to corner of mouth. The swelling was bony hard in consistency and was tender on palpation.

Intra oral Inspection revealed (Figure2) a swelling in the right maxillary region extending anteriorly from mesial aspect of 15 posteriorly to maxillary tuberosity also the swelling extends on the palatal aspect crossing the intermaxillary suture. There is buccal and palatal cortical plate expansion with obliteration of buccal vestibule. No displacement or mobility of teeth was evident. Based on the clinical presentation, fibro osseous lesions were considered as working diagnosis and benign odontogenic and non odontogenic tumors were considered in the differential diagnosis.

Intraoral periapical radiograph (Figure 3) shows loss of lamina dura altered trabecular pattern with no resorption of roots in relation to 17,18. Occlusal radiograph (Figure 4) shows altered trabecular pattern. Orthopantamograph (Figure 5) showed a well defined radiolucent lesion of approx size 6x 6 cm occupying right maxilla obliterating the right maxillary sinus, displacing the lateral & posterior wall of sinus. Superiorly the lesion is

extending till lateral wall of orbit, inferiorly into the maxillary alveolus & tuberosity region medially encroaching the anterior & medial wall of maxillary sinus, posteriorly towards infratemporal fossa with few areas of focal calcifications within the lesion. The lesion is outlined by a sclerotic border. There is also inferior displacement of 17,18 by the lesion. On Computed Tomography (Figure 6) a well defined hypodense expansile lesion of size 6X6 cm occupying right maxilla is present with calcifications. There is obliteration of sinus with thinning of sinus wall. Posterior wall of right maxillary sinus appear thinned out in the right infratemporal fossa, optic canal. Also there is narrowing of nasopharyngeal airway secondary to medial extension of the lesion

All the routine blood investigations were within normal limits except for an increase in Alkaline phosphatase level (119 IU/L) and Phosphorus levels (5.1mg/dl).

The intraosseous lesion was subjected to surgical resection extra orally using Weber fergusson (Figure 7) approach and the specimen was subjected to histopathological examination. Microscopic examination of the specimen (H and E staining) revealed (Figure 8) Verocay bodies along with areas of dystrophic calcifications. Further, Immuno histochemistry was performed, the examination of the specimen showed thattumor cells were positive forS100 (Figure 9).Therefore the final diagnosis of Ancient schwanomma was arrived. The patient is still under follow upwith no other symptoms of recurrence (Figure 10).

Discussion

Intraosseous Schwannoma is a rare, benign, encapsulated nerve sheath tumor of unknown cause originating from the Schwann cells of the neural sheath of the peripheral nerves. It was first described by Jose Jaun Verocay in 1908, under the term 'Neurinoma'. Only 25 to 45% can be found on the head and neck region, with the parapharyngeal space being the most common site. Ancient schwannoma in the oral cavity, the first report was made by Eversole and Howell in 1971.1Lowpercentage of these lesions is present in the oral cavity and when this occurs, the tongue is the most frequently involved area. Most of the intraoral schwannomas are placed in the soft tissues. Intraosseous localization is rare in jaw bones and represents less than 1% of benign primary tumors.[2] Mandible is the most favoured site, the reason being attributed to the large caliber of the inferior alveolar nerve and its long course within the jaw. Maxillary schwannomas are extremely rare. Only 15 cases of intraosseous schwannomas affecting the maxilla have been reported.[3,4]

Ancient schwannomas occurs in all age groups (7-12), being the younger patient reported 19 years old. It is more common between the second and fifth decade of life and more often women, as it has been the case presented in this paper.

Clinically, it normally appears as a slow-growing, solitary exophytic lesion, non-indurated, well circumscribed, with a smooth surface. The size is variable to the time of evolution with slow growth of this lesion. Most of the cases are asymptomatic and many of them have a long duration and large size because of their lack of symptoms.[5,6] This clinical features coincide with presenting features of our case.

The clinical differential diagnosis could be with any other benign tumors such asfibroma, lipoma, neurofibroma, or salivary glands tumor.

Diagnosis must be based on radiographic investigations, thorough histologicevidence and further confirmed through Immuno histochemistry. Radiographic investigations demonstrated neoplastic bone destruction in the form of radiolucent lesions surrounded by thin, sclerotic margin. In few cases, margins were ill defined with associated alveolar bone resorption and root resorption. Further investigations include CT, which reveals tumor as a well defined, non enhancing, low density soft tissue mass without soft tissue infiltration. The radiographic imaging characteristic features were similar with our present case.

Investigations such as FNAC can be done

which can aid to exclude any cysts of odontogenic origin.[7,8] However, the histological differential diagnosis is made with other neural origin lesions, which could be neurofibroma or muscular or fibroblastic origin tumor.[9]

Histopathologically, the tumor tissue consists of Antoni A and B type areas. The Antoni A zone has parallel -formed thin reticulin fibres, fusiform-shaped cells and curled nucleus.

Among the sheets, there are acellular eosinophilic bodies or verocay bodies, formed by thin cytoplasmic fibers. Antoni B area is a hypocellular zone with loosely textured matrix, has myxoid consistency and tumor cells are haphazardly arranged. Degenerative changes such as the presence of mild pleomorphism bizarre nuclei, dilated blood vessels, calcifications and hemorrhagic features were recognized in several areas. These findings were similar in our case which also revealed verocay bodies along with dystropic calcifications.

The immunohistochemistry reveals the cells showing diffuse positivity for the protein S-100 and marked positivity for vimentin.[7,8]

Schwannomas are resistant to radiotherapy and as they are well encapsulated the treatment of choice is conservative surgical enucleation.[9] The encapsulated form is enucleated easily, whereas the non-encapsulated requires normal tissue margins to avoid relapse. If the nerve of origin is visualized, an attempt should be made to separate carefully so as to preserve function, although this is sometimes not possible. Periodic follow up has to be done. Prognosis is good since it does not usually recur, and malignant transformation is rare.[8,9] Our patient was subjected to surgical excision and is still under follow up without any symptoms of any recurrence.

Conclusion

Intraosseous ancient Schwannomas of the jaws are exceedingly rare and of these maxillary schwannomas are rarest and pose diagnostic difficulties. Whenever we oral diagnosticians encounter painless progressive maxillary swelling, rarities such as intraosseous schwannomas should also be considered in the differential diagnosis. A thorough microscopic evaluation is mandatory to arrive at the correct diagnosis further confirmed by immunohistochemistry.

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Fig 1.
Swelling on right side of face

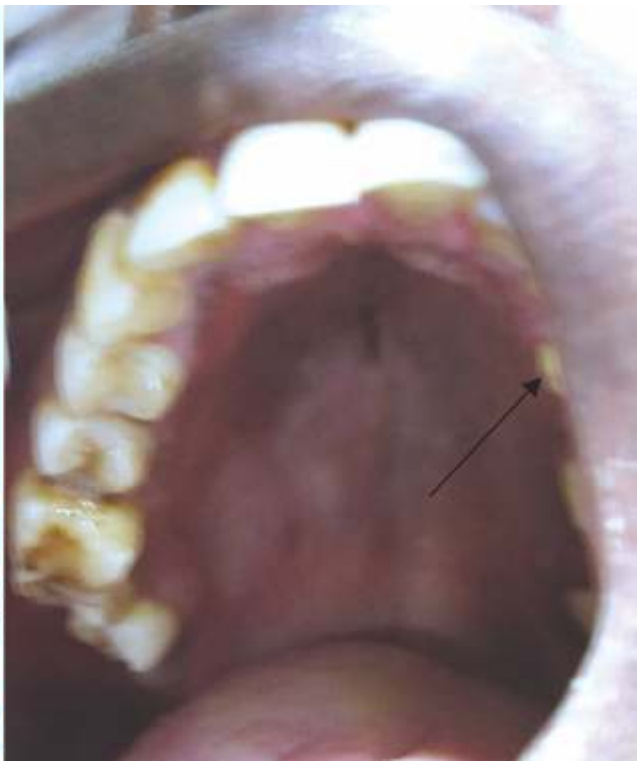

Fig 2. Intraoral swelling

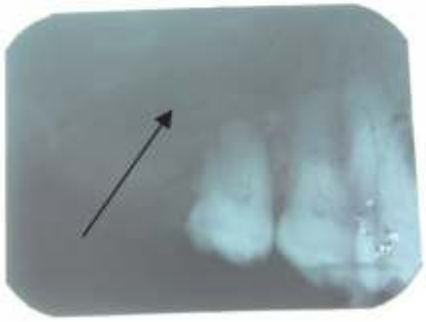

Fig 3.
Intraoral periapical radiograph shows loss of lamina dura altered trabecular pattern with no root resorption in relation to 17, 18.

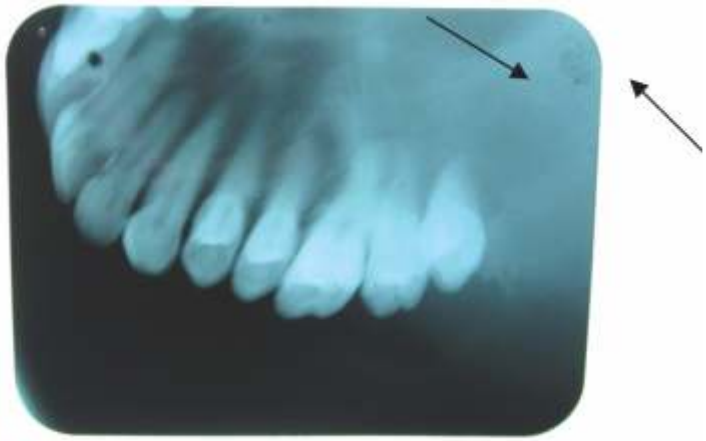

Fig 4. Occlusal radiograph shows altered trabecular pattern.

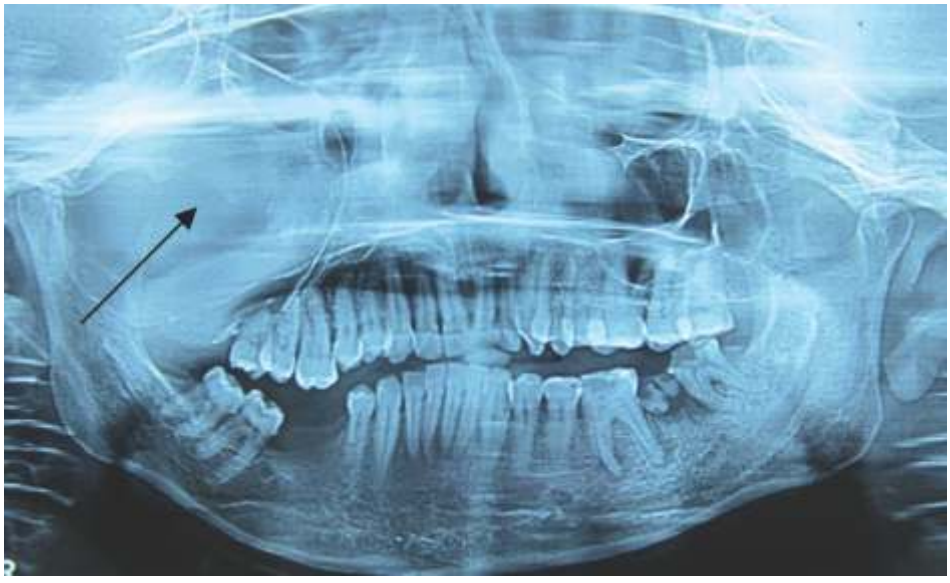

Fig. 5
Orthopantomograph shows unilocular well defined area of radiolucency present on right side of maxilla with focal areas of calcifications

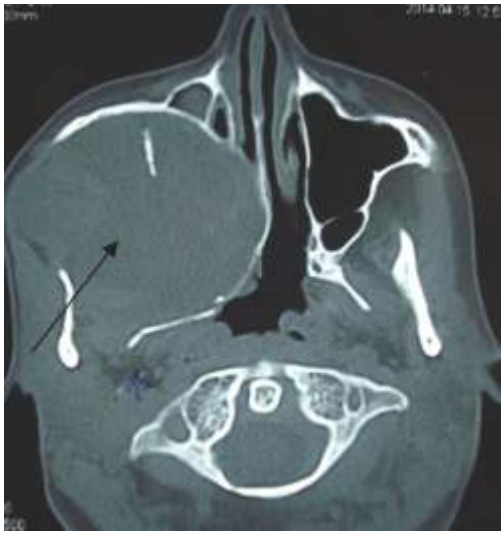

Figure 6
On computed tomography well defined hypodense lesion noted with calcifications in right maxilla obliterating the right maxillary sinus.

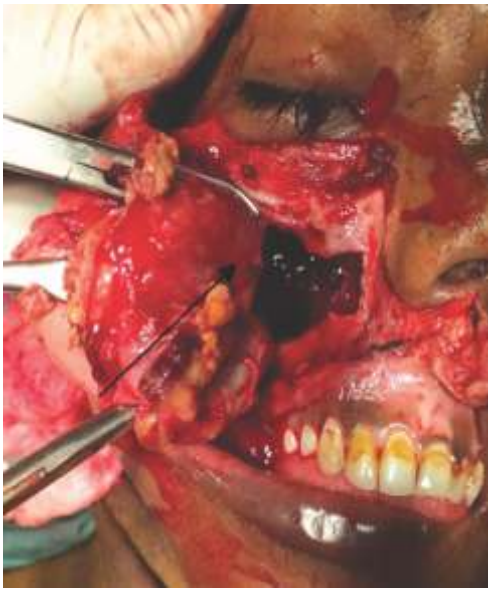

Figure 7. Intra Operative picture

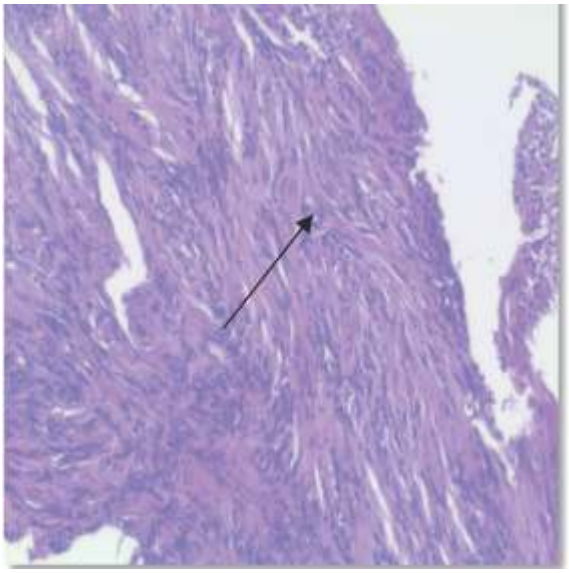

Figure 8.
On H&E staining
at 10x reveals verocay bodies

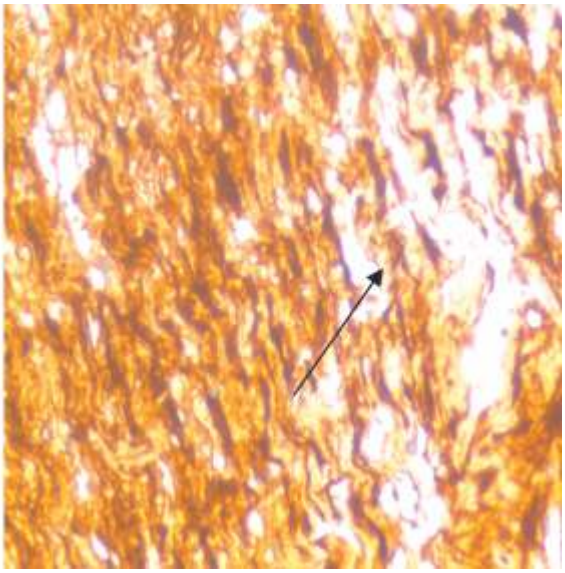

Figure 9.
On Immunohistochemistry
S-100 at 4x reveals tumor cells

Figure 10. Post Operative picture