Intraosseous Schwannoma of the Mandible

V Manjunath, Vijeev Vasudevan, Nandakumar, Srinath, Radhika Manoj Bavle

1Senior Lecturer, Department of Oral Medicine and Radiodiagnosis, Krishnadevaraya College of Dental Sciences and Hospital, Bengaluru, Karnataka, India
2Professor and Head, Department of Oral Medicine and Radiodiagnosis, Krishnadevaraya College of Dental Sciences and Hospital, Bengaluru, Karnataka, India
3Professor and Head, Department of Oral and Maxillofacial Surgery, Krishnadevaraya College of Dental Sciences and Hospital, Bengaluru, Karnataka, India
4Professor, Department of Oral and Maxillofacial Surgery, Krishnadevaraya College of Dental Sciences and Hospital, Bengaluru, Karnataka, India
5Professor and Head, Department of Oral Pathology and Microbiology, Krishnadevaraya College of Dental Sciences and Hospital, Bengaluru, Karnataka, India

Correspondence: Vijeev Vasudevan, Professor and Head, Department of Oral Medicine and Radiodiagnosis, Krishnadevaraya College of Dental Sciences and Hospital, Hunasamaranhalli, Via Yelahanka, Bengaluru-562157, Karnataka, India
e-mail: vijeev29@rediffmail.com, manjuv79@gmail.com

ABSTRACT
Schwannoma (neurilemmoma) is a benign neoplasm originated from the neural sheath and occurring most frequently in maxillofacial region. Intraosseous schwannomas are rare. The mandible is the most common site of occurrence for these lesions. We report a rare case of Intraosseous neurilemmoma of the mandible, with an emphasis on clinical, radiographic and pathological findings. The tumor, located mainly in the ramus region, presented as a soft tissue mass in the oral cavity, Multilocular, well-defined, and radiolucent lesion on plain radiography. Dilatation of the mandibular canal was identified. Plain radiography partly helped to identify the solid nature of the tumor. A biopsy was necessary to make the final diagnosis because of the relatively nonspecific nature of the lesion. The lesion was surgically removed and no evidence of recurrence was observed after 6 months.

Keywords: Intraosseous schwannoma, Mandible, Multilocular, Neurilemmoma, Plexiform.

INTRODUCTION
Schwannoma is a benign neoplasm of neuroectodermal derivative, originating from the Schwann cells (peripheral nerve sheath). This painless slow-growing lesion may develop at any age and is most frequently located in the soft tissues of the head and neck. Intraoral schwannomas are rare, representing less than 1% of the benign primary bone tumors, intraosseous schwannoma is exceptionally rare. The most common site of occurrence is the mandible compared to both jaws.

This article reports the case of an intraosseous schwannoma located in the mandibular ramus as a lobulated soft tissue mass in a 44 years old male. The clinical nature, radiographic appearance, and histopathologic variant are described and findings in the literature are discussed.

CASE REPORT
A 44-year-old man complained of swelling in the left mandibular region with 2 years of evolution, following extraction of a tooth. The patient was free of any symptom and reported a history of pain on act of mastication in relation to swelling intraorally and no paresthesia. On clinical examination that area was swollen extraorally in the left angle region, with palpable and tender left submandibular lymph nodes (Fig. 1). Intraoral examination revealed well-defined soft tissue swelling in the left buccal vestibule distal to 37 measuring around 1.5 × 1.5 cm in diameter the surface appeared smooth and on palpation, cortical expansion was evident with an area of decortications was evident at the region buccodistal to second molar, swelling was soft and pain on percussion, and mobility of the molar teeth (37) was noted (Fig. 2). His past medical history was essentially noncontributory. The clinical diagnosis of infected residual cyst, irritational fibroma, was made based on history and clinical examination. The patient under went routine dental radiography, intraoral and panoramic radiography identified a well-defined, osteolytic lesion with septation in the left posterior mandible. The lesion was located in
Intraosseous Schwannoma of the Mandible

Fig. 2: Soft tissue swelling in the left buccal vestibule

Fig. 3: Periapical radiograph showing discrete resorption of the roots of the 37 in contact with lesion and multilocular radiolucent lesion

Fig. 4: Panoramic radiograph showing well-defined multilocular radiolucent area involving the alveolar bone between the roots of the left second molar and extending into ramus

Fig. 5: Photomicrograph of resected specimen (H and E stain 40X) shows Antoni type-A tissue and Antoni type-B tissue zones, arranged in palisading pattern interspersed with verocay bodies

the angle squamous region, between the roots of 36, 37, extending into ramus area. There was apparent resorption of roots of the second molar teeth, and oblique orientation of the roots suggested a long-standing lesion. A well-delineated, expansile lesion with peripheral scalloping and erosion of the buccal cortex, in the molar region was noted (Figs 3 and 4). Based on these findings a differential diagnosis of odontogenic keratocyst, ameloblastoma, central giant cell granuloma, intra-alveolar carcinoma was made. A biopsy was obtained under local anesthesia, and a preliminary diagnosis of schwannoma was made. Subsequently, under general anesthesia, the patient underwent total removal of the tumor. The tumor was easily removed, leaving the inferior alveolar, neurovascular bundle intact. Microscopic examination of the hematoxylin-eosin–stained section of the biopsy specimen revealed foci of antoni type-A tissue, including spindle cells arranged with palisading, ovoid, basophilic nuclei and acidophilic cytoplasm, and antoni type-B tissue. Acellular, eosinophilic areas described as verocay bodies, were also noted. With histopathological variant of plexiform type (Fig. 5). Immunohistochemical examination revealed positive anti-S-100 protein. The histopathological diagnosis of benign intraosseous schwannoma was confirmed by evaluation of the resected specimen.

DISCUSSION
Schwannoma was first described by verocay in 1910. He called it “neurinoma”. In 1935, the term “neurilemmoma” was coined by Stout. Neurilemmoma is a benign neoplasm that originates from the Schwann cells that cover the peripheral nerves. This tumor presents as a painless, slow-growing mass and it may develop at any age ranging from 2 to 72 years, with the peak prevalence in the second and third decades of life with more female predilection with a ratio of 2:1 over males. The present case is some what less common because of tumor in a male patient. Neurilemmomas are most frequently located in the soft tissues of the head and neck, but can also occur in upper and lower extremities.
Intraosseous neurilemmoma is extremely rare and fewer than 45 examples of this entity have been described in the literature pertaining to maxilla and mandible, of which mandible is the most preferred site.\textsuperscript{5-11} It accounts for less than 1% of all primary bone tumors.

In the mandible, the body and ramus are the most frequent sites of occurrence because of the intraosseous path of the inferior alveolar nerve canal.\textsuperscript{12,13} The tumor presents as a painless, slow-growing mass associated with parasthesia, when it encroaches the adjacent nerve. Where as parasthesia was absent in this case. The clinical presentation of this case was a painless soft tissue swelling tumor in the region of mandibular ramus and squamous region in a man of the fourth decade of life. There are three mechanisms by which schwannoma may involve bone. (1) the tumor may arise with in the bone, (2) the tumor may arise within a nutrient canal and produce canal enlargement or (3) soft tissue or periosteal tumor may cause secondary erosion and penetration into bone.\textsuperscript{14} In present case the tumor was purely intraosseous and no evidence of any involvement of soft tissue periosteal region. Intraosseous schwannomas radiographically present as unilocular or multilocular with sclerotic border, which may resemble many benign processes such as odontogenic keratocysts, periodontal cysts or ameloblastoma. In the present case, the possibility of neurilemmoma was not considered during the first radiologic study because of its multilocular nature, which is less common. In some instances, the sclerotic rim is absent focally, especially towards the ridge or in areas of cortical thinning or perforation. The inferior cortex as well as the posterior and anterior cortices may be eroded. The well-defined margins are often irregular, and this irregularity may involve the entire margin or may be limited to part of the margin within a given lesion giving a scalloped appearance of the lesion, and this has been termed as multilocular by some authors in literature. Additional features such as external root resorption, displacement of teeth; spotty calcification, cortical expansion, and peripheral scalloping can be evident and may rarely associated with pathological fracture.\textsuperscript{14-16} The present lesion had multilocularity, scalloping pattern, and root resorption which are consistent with the findings in literature.

Histopathologically, neurilemmoma provides a characteristic alteration of two types of tissue arrangement, Antoni A and Antoni B. The Antoni A areas are relatively cellular, and when they are more differentiated, they may exhibit nuclear palisading, whorling and Verocay bodies. The Antoni B areas are less cellular and less organized, and they often contain prominent thickened blood vessels.\textsuperscript{17} In addition to this classic form, there are several histopathologic variants that include the cellular, plexiform, epithelioid, ancient, and melanotic types. The present case was of plexiform variant microscopically, immunoreactivity for S-100 protein is routinely observed in neurilemmomas, where as absent in other lesions, it is also positive for CD34, and epithelial membrane antigen.\textsuperscript{18} Malignant form of Schwannomas has also been documented in literature.

The treatment of choice for neurilemmomas is conservative surgical enucleation with periodic follow-up, recurrence is uncommon.\textsuperscript{19} In the present case, the patient was followed up for six months with no clinical or radiographic signs of recurrence.

**CONCLUSION**

In conclusion, we present a neurilemmoma that developed in an intraosseous location within the ramus of mandible. Although very rare, intraosseous neurilemmoma should be included in the differential diagnosis of painless soft tissue mass, radiographically benign appearing, multilocular radiolucent lesions arising in jaws and any odontogenic cysts and tumors.

**REFERENCES**