CASE REPORT

Intraosseous schwannoma of the mandible

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ABSTRACT
Schwannomas (neurilemmomas) are slow-growing, benign neoplasms derived from schwann cells, the sheath cells that cover myelinated nerve fibers. These tumors most commonly arise in the soft tissues of the head and neck, as well as on the flexor surfaces of the upper and lower extremities. Intraoral lesions are uncommon, however, and intraosseous schwannomas are even rarer. In the Mayo Clinic series of 11,087 primary bone tumors, 14 cases of intraosseous schwannoma were identified, accounting for less than 1% of these benign primary bone tumors. The most common site of occurrence is the mandible, a characteristic traditionally attributed to the long intraosseous path of the inferior alveolar nerve. In this article, we describe an additional case occurring in the mandible of a 15-year-old boy.
Key words: Intraosseous schwannoma, mandible, neurilemmoma

INTRODUCTION

Schwannomas were first established as a pathologic entity in 1910 by Verocay who called them neurinomas. Schwannoma (also known as neurilemmoma, neurolemmoma, neurinoma, perineural fibroblastoma, and peripheral nerve sheath tumor) is a slow-growing, benign neoplasm derived from Schwann cells, which are sheath cells that cover myelinated nerve fibres.[1] The soft tissue of the head and neck region is one of the most common sites for benign nerve sheath tumors as well as the flexor surfaces of the extremities.[2] Intraoral development is uncommon (only 1%). Most common site of occurrence is the mandible, attributed to the long intraosseous path of the inferior alveolar nerve. In the current medical literature, there are 44 acceptable cases of intraosseous schwannoma of the jaws, 39 of the mandible and five in the maxilla, representing less than 1% of the primary tumors of the bones. Other sites reported include the sacrum, vertebra, clavicle, ribs, humerus, radius, ulna etc.[3]

CASE REPORT

A 15-year-old male patient reported with the chief complaint of swelling involving the lower jaw since 6 months. Initially, the swelling was of peanut size and had been gradually increasing, and reached the present size. There was no associated pain or discomfort. The patient's medical history, drug history, and general physical examination were all non-significant.

Clinical examination revealed a diffuse 4 cm × 4 cm swelling, extending from the corner of the mouth to the angle of the mandible, roughly being quadrilateral in outline. Figure 1 shows extraoral photograph with swelling on the left side of the face. Figure 2 shows intraoral photograph with the swelling in the left lower buccal vestibule, extending anteriorly from the distal surface of lower left first premolar to the ascending ramus posteriorly. Buccal and lingual cortexes were expanded. The premolar and molar on the involved side were grade 3 mobile.

Figures 3 and 4 radiographically revealed the lesion as a large multilocular radiolucency extending from distal of first premolar to the ramus of the mandible. Root resorption was seen in lower left 2nd premolar and 1st and 2nd molar. Lower border of the mandible appeared thinned out.

Incisional biopsy consisted of multiple pieces of soft tissues, measuring about 1 mm × 5 mm in diameter, brownish white in color, soft in consistency, and round to oval in shape. Microscopically, Figures 5 and 6 revealed highly cellular stroma with cells arranged in sworls and whorls, with interspersed vascular channels. The cell boundaries were indistinct. Antoni type A and B type arrangement of cells was seen. A few areas showed verocay bodies. A few nerve bundles, collagen fiber bundles, myxoid areas, endothelial lined vascular channels

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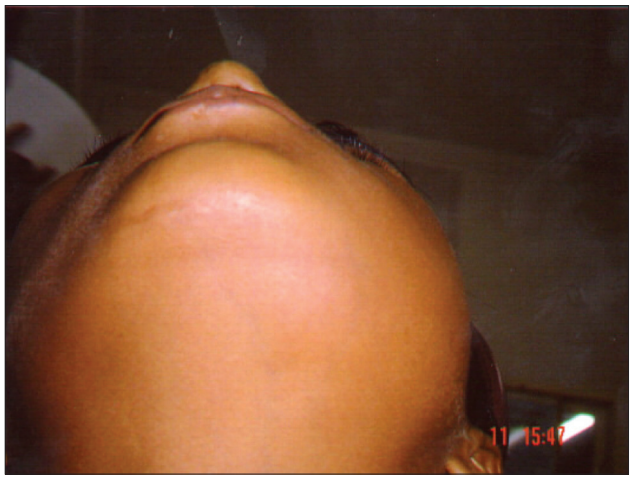

Figure 1: Extraoral photograph with swelling on the left side of the face

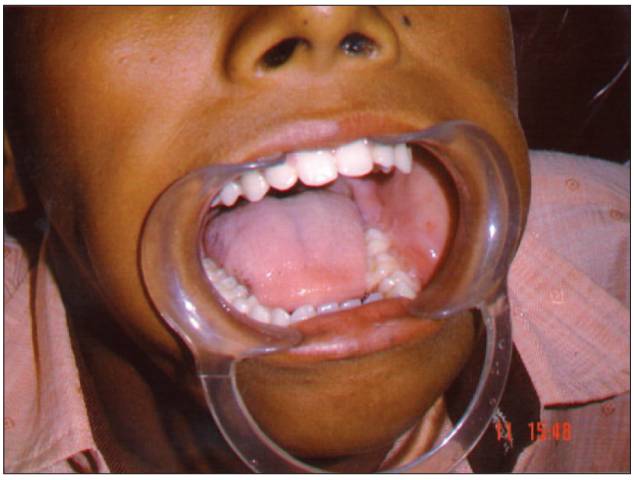

Figure 2: Intraoral photograph with lesion on the left side of the jaw

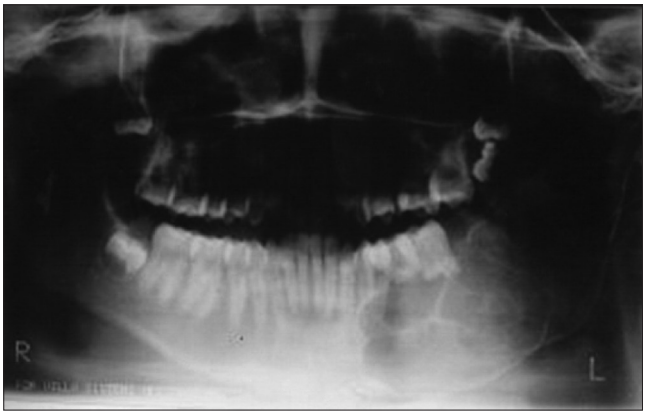

Figure 3: Photograph of orthopantomogram with multilocular radiolucency on the left side of the mandible

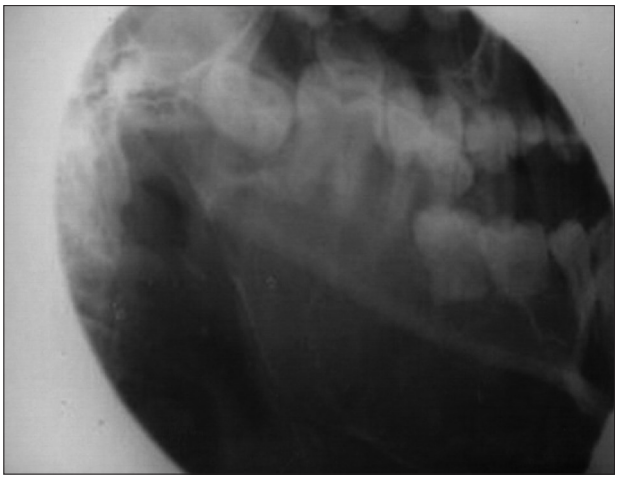

Figure 4: Lateral oblique view of the lesion

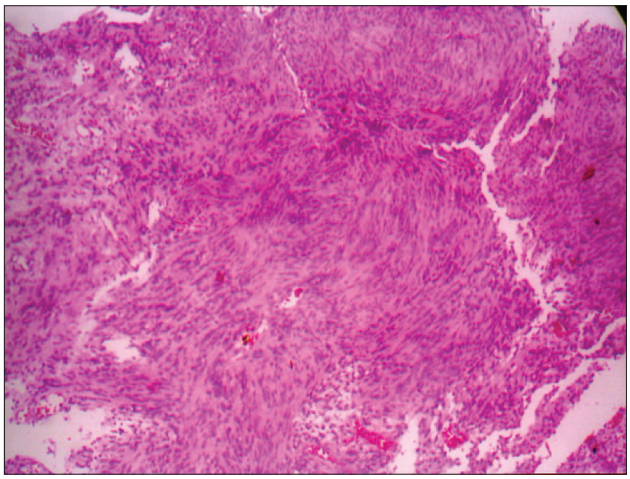

Figure 5: Photomicrograph, H and E stained section, 10× view of the lesion

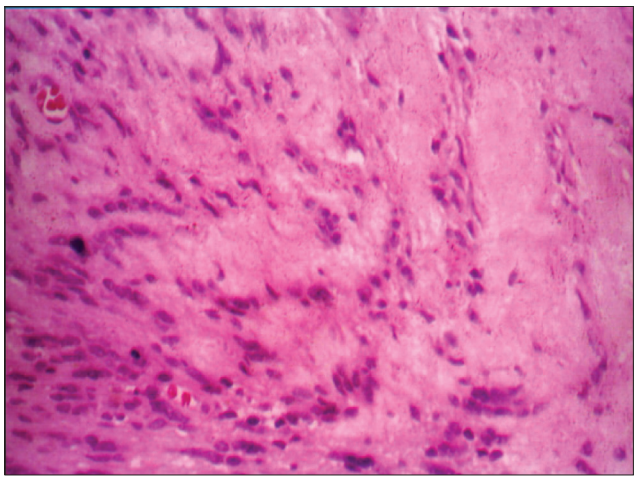

Figure 6: Photomicrograph, 40× view of the lesion

with RBC's, and extravasated RBC's were seen. Based on the findings, the diagnosis of schwannoma was given.

DISCUSSION

Schwannoma is a benign tumor, apparently derived from the Schwann cells, which may arise from any myelinated nerve fibres.[4] Schwannoma rarely occurs in the oral cavity. Intraosseous schwannomas are rare, (less than 1%) but when they occur, the mandible is the most commonly affected site. Most cases reported in the mandible had a more posterior

location, corresponding to the intraosseous course of the inferior alveolar nerve.[5]

There are three mechanisms by which schwannomas may involve a bone:

1. A tumor may arise centrally within a bone,
2. A tumor may arise within a nutrient canal, or
3. A soft tissue or periosteal tumor may cause secondary erosion and penetration into bone.[1]

This case demonstrates an example of schwannoma centrally within a bone.

Schwannomas most often occur in the fourth and fifth decade of life with a 1.6:1 female predilection. The duration varies from few months to a few years.[1] Clinically, neurilemmoma is a slow-growing tumor that may be present for years before becoming symptomatic. Swelling is the most common symptom, but pain or paresthesia may be present in about 50% of cases.[6] Radiographic findings of intraosseous mandibular schwannoma show a great variation, from unilocular to multilocular, with or without well-defined borders of the lesion, and cortical expansion.[2]

The present case revealed a large multilocular radiolucency extending from distal of first premolar to the ramus of the mandible. Root resorption was seen in the lower left 2nd premolar and 1st and 2nd molar. Lower border of the mandible appeared thinned out. When seen as a gross specimen, the tissue of schwannoma is solid, roundly lobulated, and grayish white in color. It is soft or moderately firm.[7] Most tumors are encapsulated. They ranged in size from 1.0 to 19.5 cm and had a mean greatest diameter of 6.2 cm.[8]

Histologically, schwannomas are described as Antoni A type or Antoni B type with verocay bodies with palisading nuclei. [9] Chrysomali *et al*. reported that schwannomas consistently showed positive staining for S-100 protein in most of the tumor cells. CD57 positive cells varied from 0.1% to 10% in: Schwannomas. Malignant transformation of the schwannoma is almost unknown although one acceptable example has been reported. (carstens and schrodt, 1969). However, recurrence of the schwannoma is possible if it is inadequately excised. [10] Schwannomas are resistant to radiotherapy.[11] Because it is a well-encapsulated lesion, the treatment of choice is conservative surgical enucleation with periodic follow-up.

As diagnostic tools, ultrasonography, computed tomography, and magnetic resonance imaging may be helpful for estimation of tumor margins as well as infiltration of surrounding structures. Nevertheless, they should not be considered as routine or indispensable procedures.

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