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Intraosseous Schwannoma of the Mandible - A Case Report

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ABSTRACT

Schwannomas, originally called neurilemmoma, are benign and slow growing peripheral nerve sheath neoplasms. They originate from Schwann cells, which are the myelin depositing cells of the peripheral nervous system, thus deriving its name. They can be found in both soft tissue and bone. When found within bone, they are referred to as intraosseous, or central, schwannoma. Intraosseous lesions are exceptionally rare, representing less than 1% of all primary tumors of the jaws. Conservative excision is the treatment of choice and is associated with a near 100% cure rate. Although extremely rare, schwannomas can undergo malignant transformation. The aim of this report is to describe an additional intraosseous schwannoma of the mandible and provide further literal review literature on this rare entity.

CASE REPORT

A 52-year-old Caucasian female was referred to the University of Tennessee Health Science Center Department of Oral and Maxillofacial Surgery by her orthodontist. The referral noted the incidental finding of a radiolucent lesion in the left mandible found on routine radiographic imaging. Upon initial consultation, intraoral examination revealed slight buccal expansion of the left posterior mandible. Teeth #'s 17-22 all tested vital, were non-mobile, and had no associated tenderness or other signs or symptoms of infection. Of note, the patient also denied any history of pain or neurosensory disturbances. A panoramic radiograph demonstrated a well-circumscribed, radiolucent lesion with scant radiopacities and root resorption of adjacent teeth (Figure 1).

A CBCT was taken, which revealed a radiolucent lesion of the left mandible with scant radiopacities noted within. The lesion also demonstrated loss of the buccal cortex and root resorption of adjacent teeth #'s 19 and 20 (Figures 2-5). The differential diagnosis included desmoplastic ameloblastoma, calcifying odontogenic cyst, calcifying epithelial odontogenic tumor, desmoplastic fibroma, ossifying fibroma, ameloblastic fibro-odontoma, schwannoma, vascular lesion, osteosarcoma, and metastatic cancer from the breast or prostate.

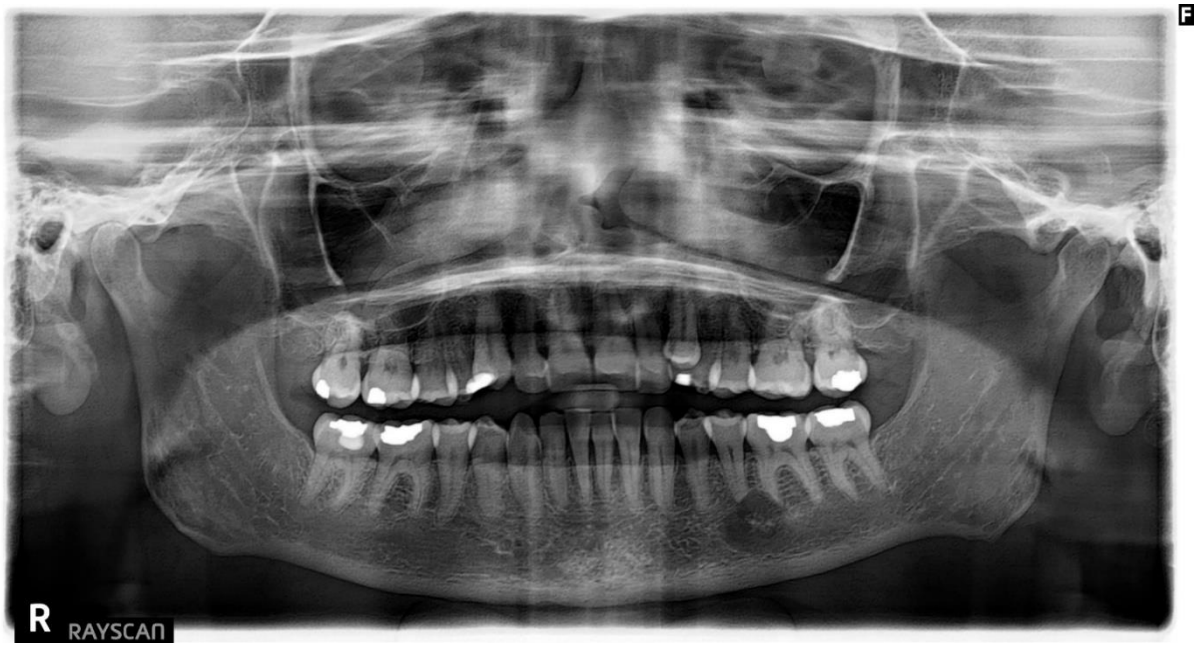

Figure 1: Panoramic radiograph at initial exam with schwannoma apical to #s 19 and 20.

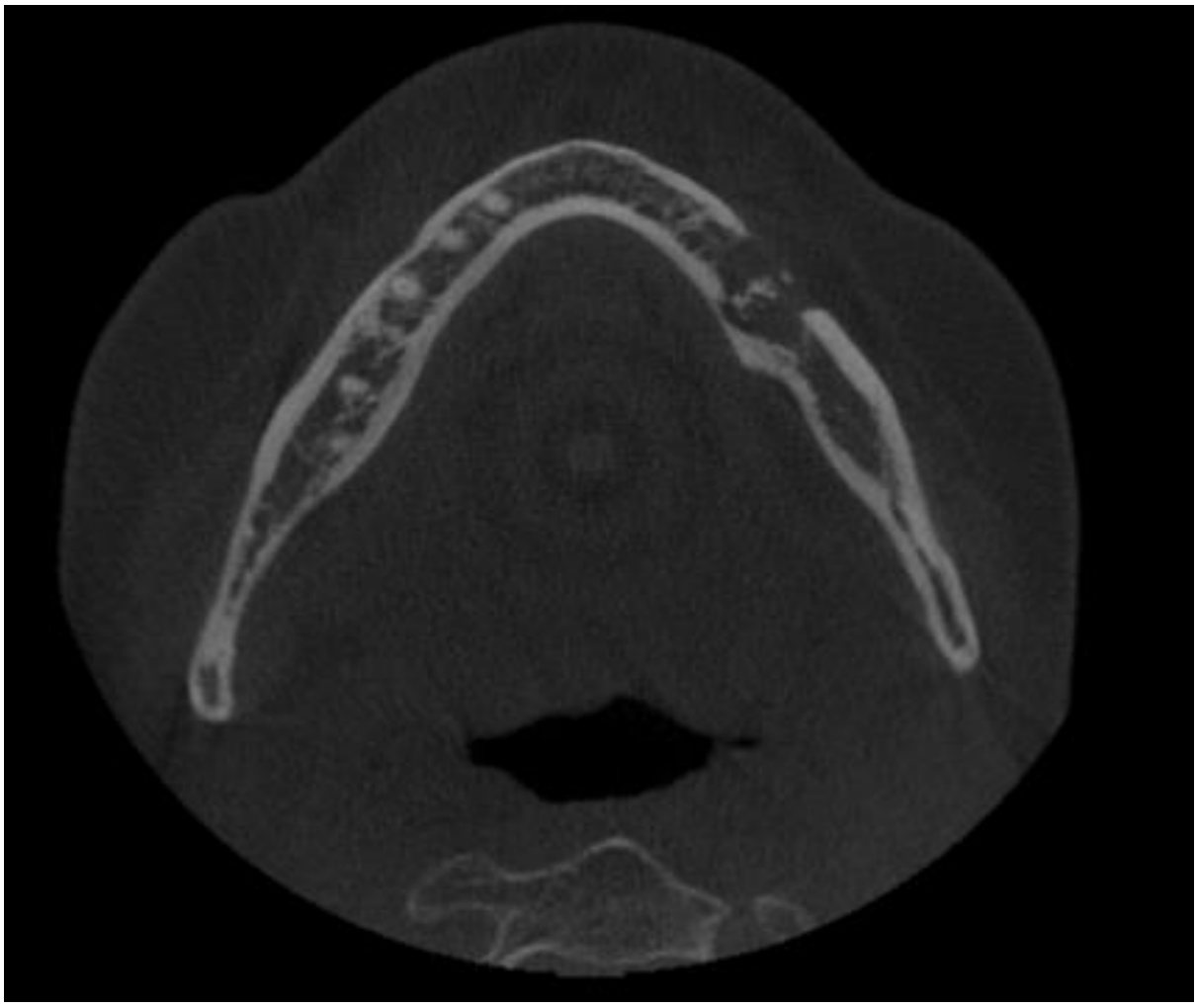

Figure 2: CBCT axial slice of left mandible schwannoma.

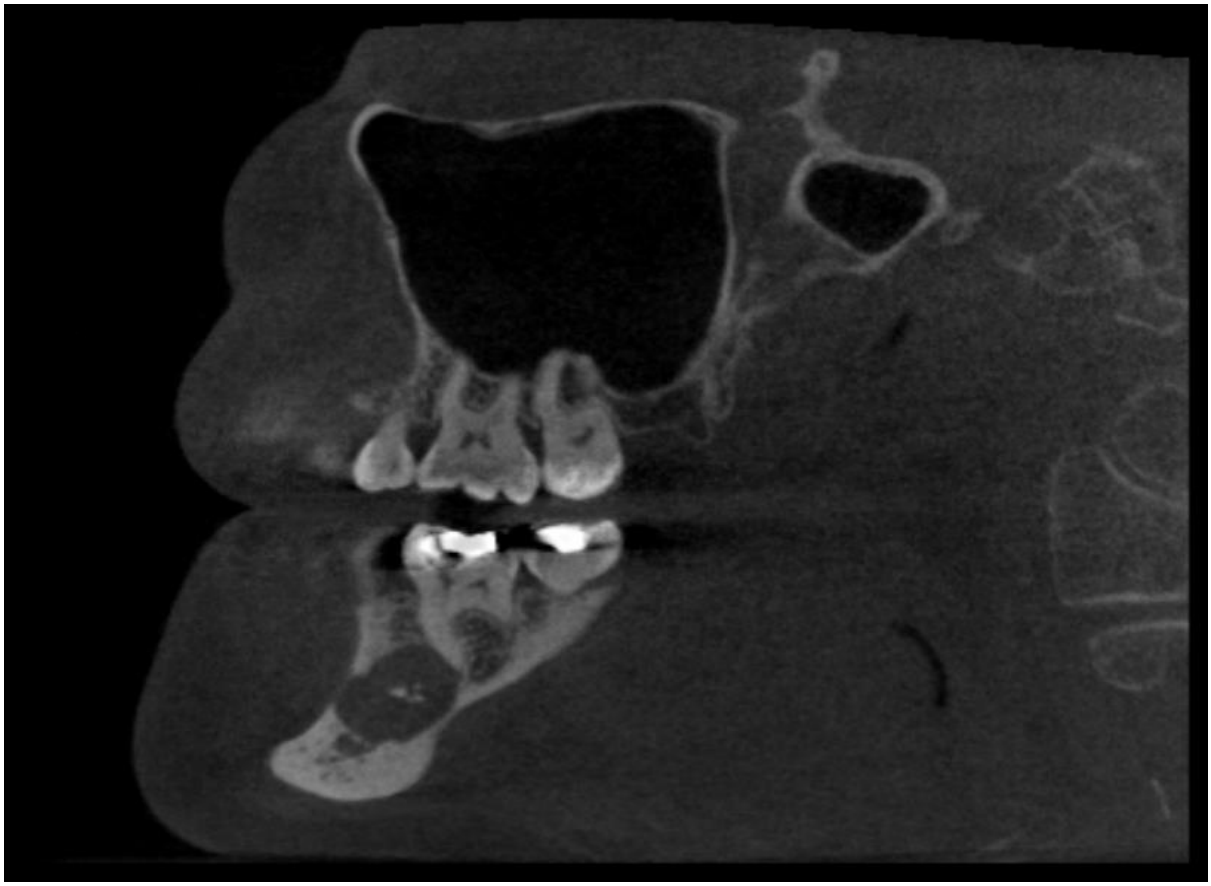

Figure 3: CBCT sagittal slice of left mandible schwannoma.

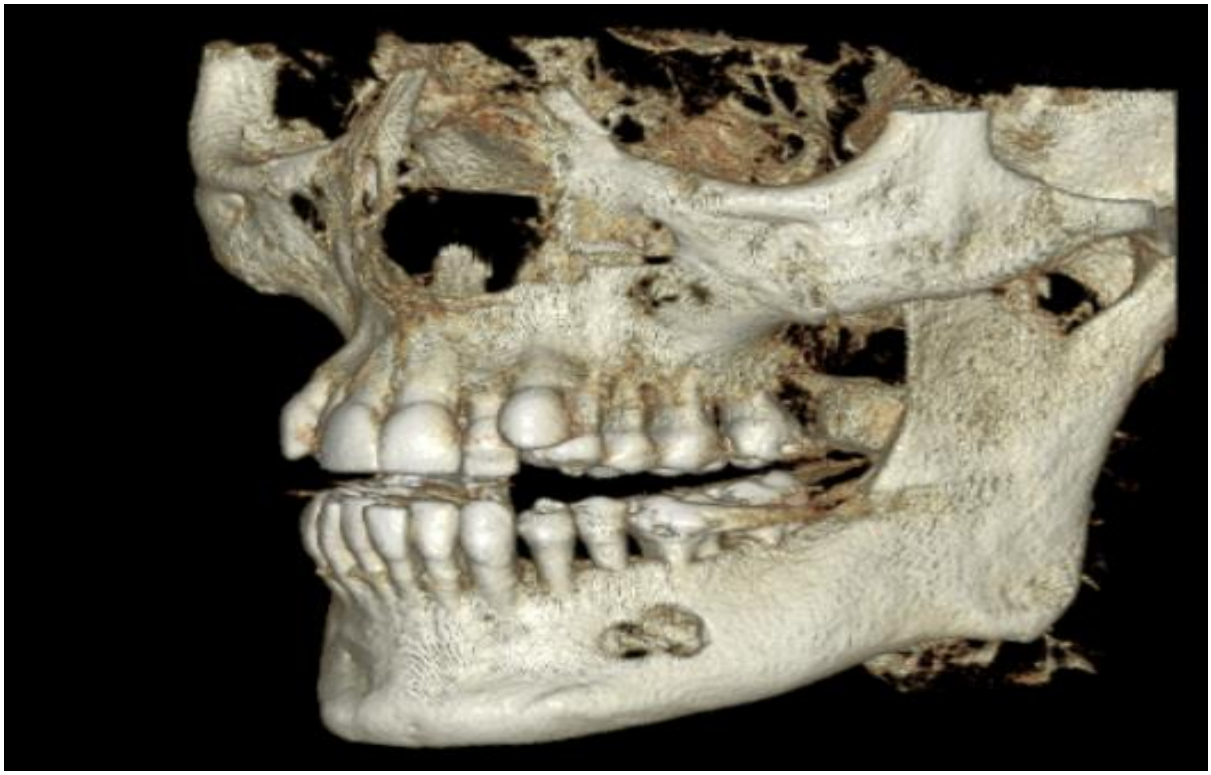

Figure 4: CBCT 3D Reconstruction demonstrating the left mandible schwannoma.

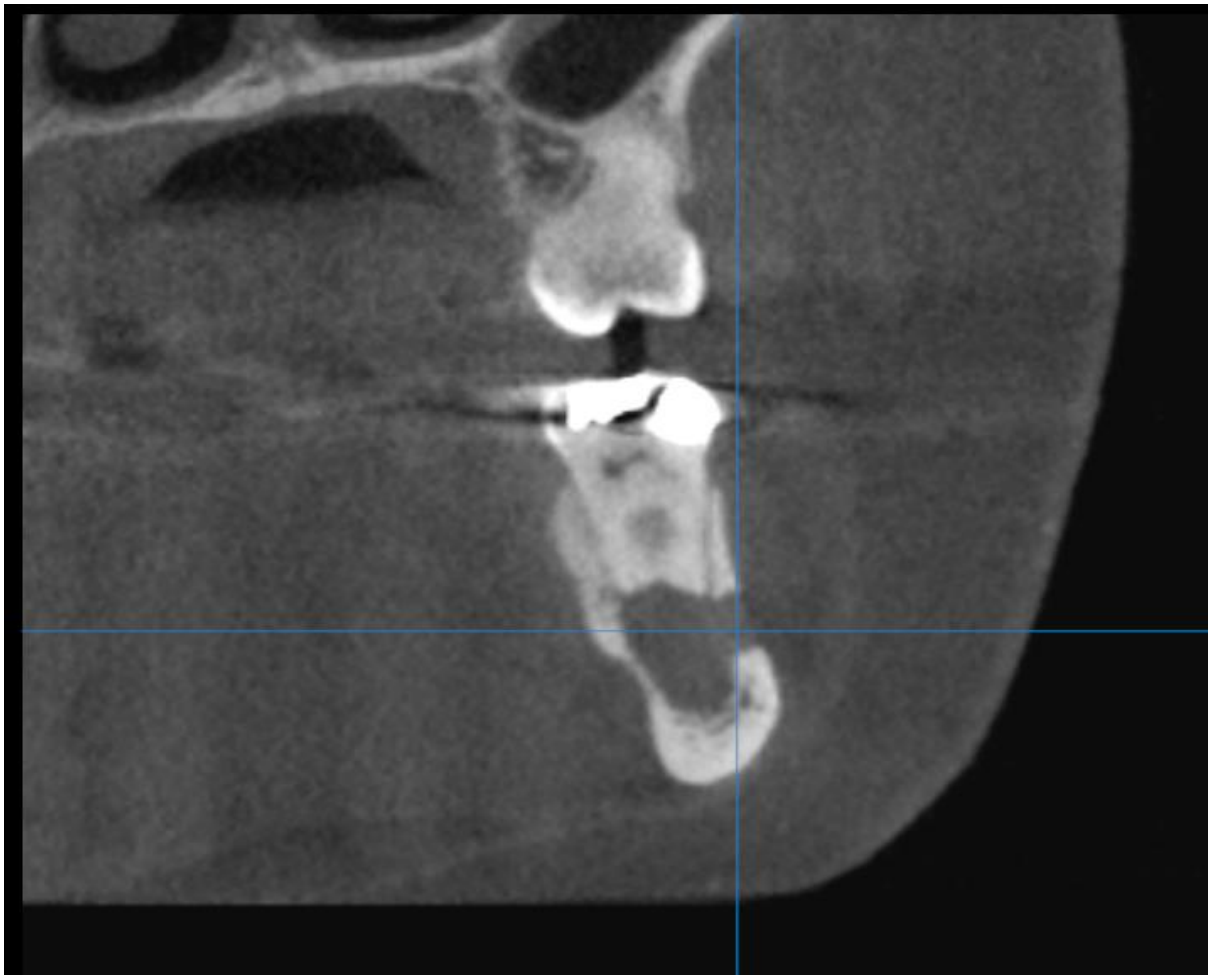

Figure 5: CBCT coronal of schwannoma in left mandible near the mental foramen.

The patient underwent an incisional biopsy under IV sedation to obtain a definitive diagnosis prior to rendering definitive treatment (Figure 6). A #15 blade was used to make a sulcular incision from teeth #19 to #21. A full thickness mucoperiosteal flap was reflected to reveal the underlying mandible. A small area of buccal cortex perforation was noted. Through the pre-existing perforation, the lesion was aspirated using an 18-gauge needle and 10 cc syringe to rule out a vascular lesion. The aspirate was negative for any heme but was noted to contain a minimal amount of straw-colored fluid. A high-speed handpiece and fissure bur were used to extend the pre-existing buccal cortical perforation into an approximately 8 x 8 mm bony window. The lesion was readily encountered, and a double-ended curette was used to enucleate a portion of the lesion. Two separate samples were taken from the lesion and placed in a formalin solution and submitted for histopathological examination.

Histopathological examination performed at UTHSC revealed fragments of fibrous connective tissue populated by spindled, wavy nuclei with some suggestion of palisading Antoni A tissue. The lesion exhibited positive staining with the immunohistochemical marker S100. All appropriate controls were viewed simultaneously and deemed adequate. Therefore, a diagnosis of benign peripheral nerve sheath tumor was favored, suggestive of schwannoma. The patient returned to clinic approximately two weeks after the incisional biopsy and was informed of her diagnosis. She was subsequently worked up for conservative surgical excision of the lesion in the operating room under general anesthesia.

The patient was taken to an outpatient surgery center for definitive treatment under general anesthesia. The lesion was accessed in the same manner as previously described during the incisional biopsy procedure. A surgical handpiece and fissure bur were used to further extend the bony window to include the entire lateral aspect of the buccal cortical plate overlying the lesion. The schwannoma was removed with careful and conservative surgical excision. The patient was discharged to home the same day as the procedure.

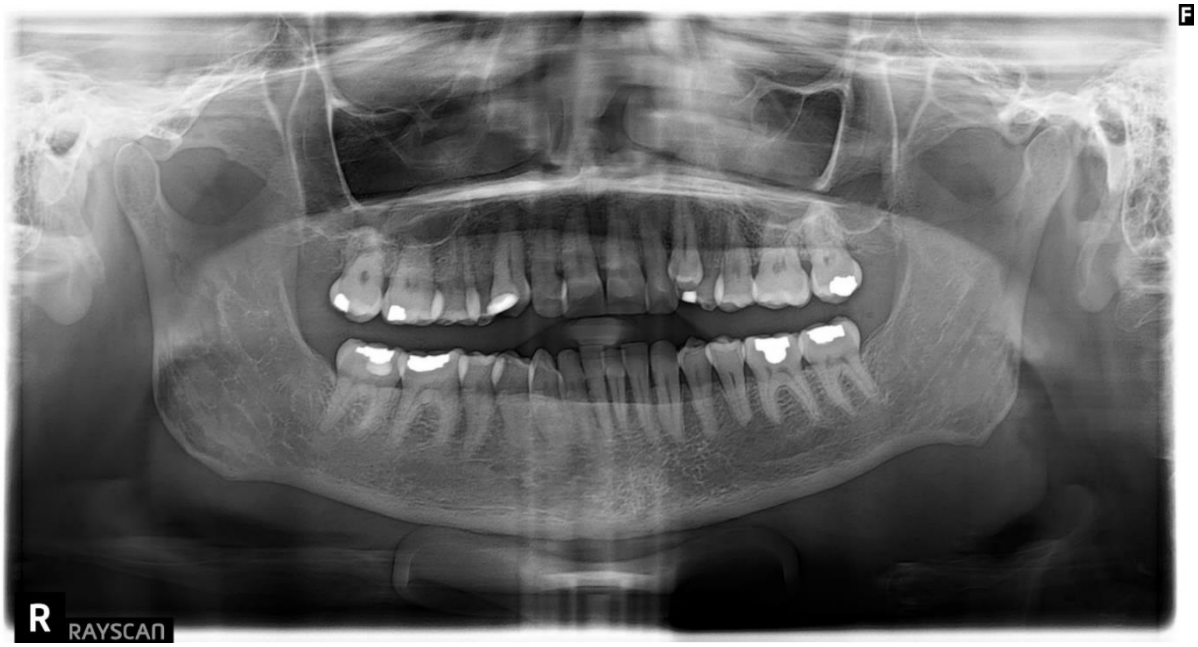

Figure 6: Pre-op panoramic radiograph taken at time of biopsy.

The patient was followed for routine post-operative care at routine intervals. Of note, the patient had paresthesia of the left mental nerve distribution following surgery but otherwise had an uneventful post-operative course. A panoramic radiograph was taken at subsequent follow-up appointments to assess healing and for surveillance. The patient was brought back for routine follow-up visits at 2 weeks, 6 weeks, 10 weeks, 4 months, 6 months, 10 months, and 20 months post-operatively with panoramic radiographs being taken at 1 week (Figure 7), 4 months (Figure 8), 6 months (Figure 9), 10 months, and 20 months (Figure 10). Consecutive panoramic radiographs demonstrated bony infill of the defect with no signs of lesion persistence or recurrence. The left mental nerve paresthesia progressively improved at subsequent follow up appointments.

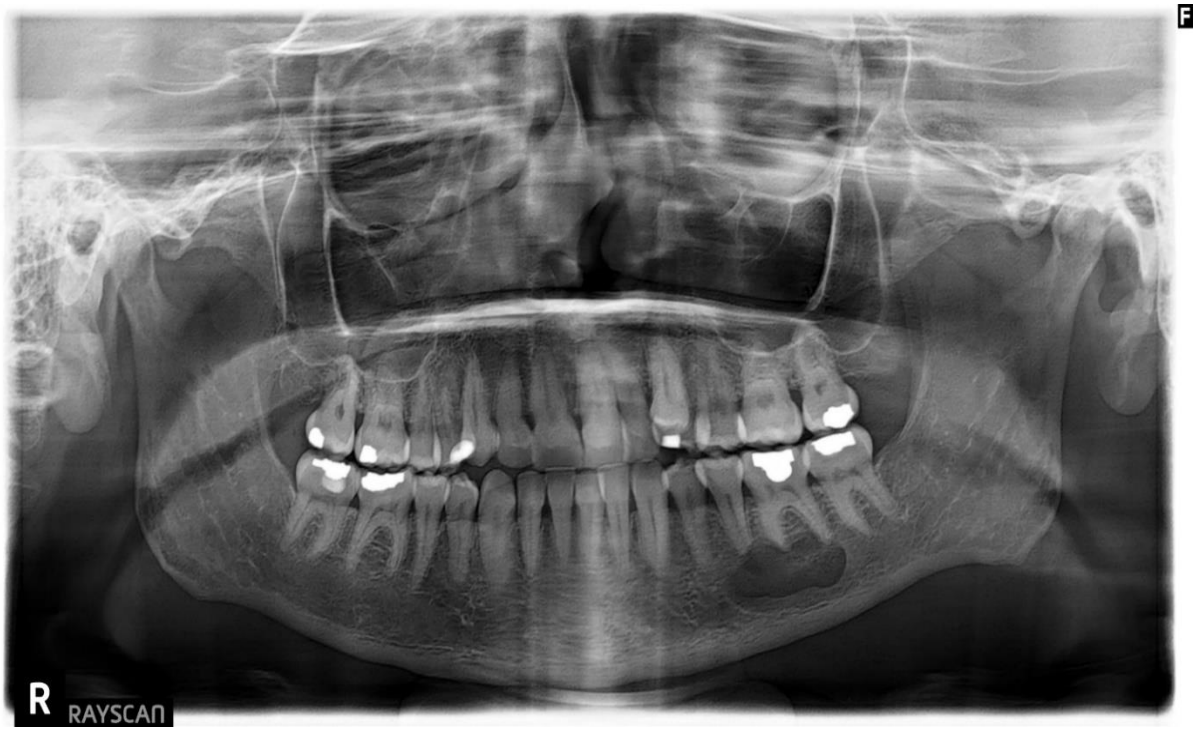

Figure 7: Panoramic radiograph 1-week post-op.

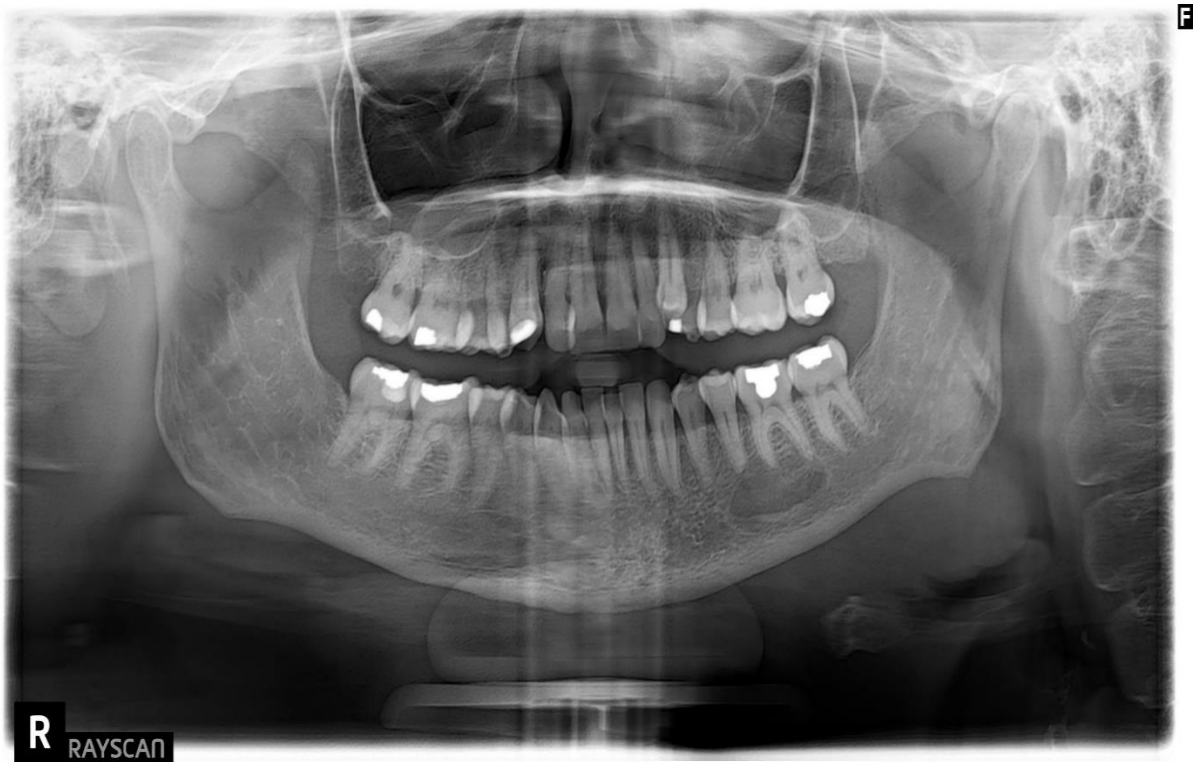

Figure 8: Panoramic radiograph 4 months post-op.

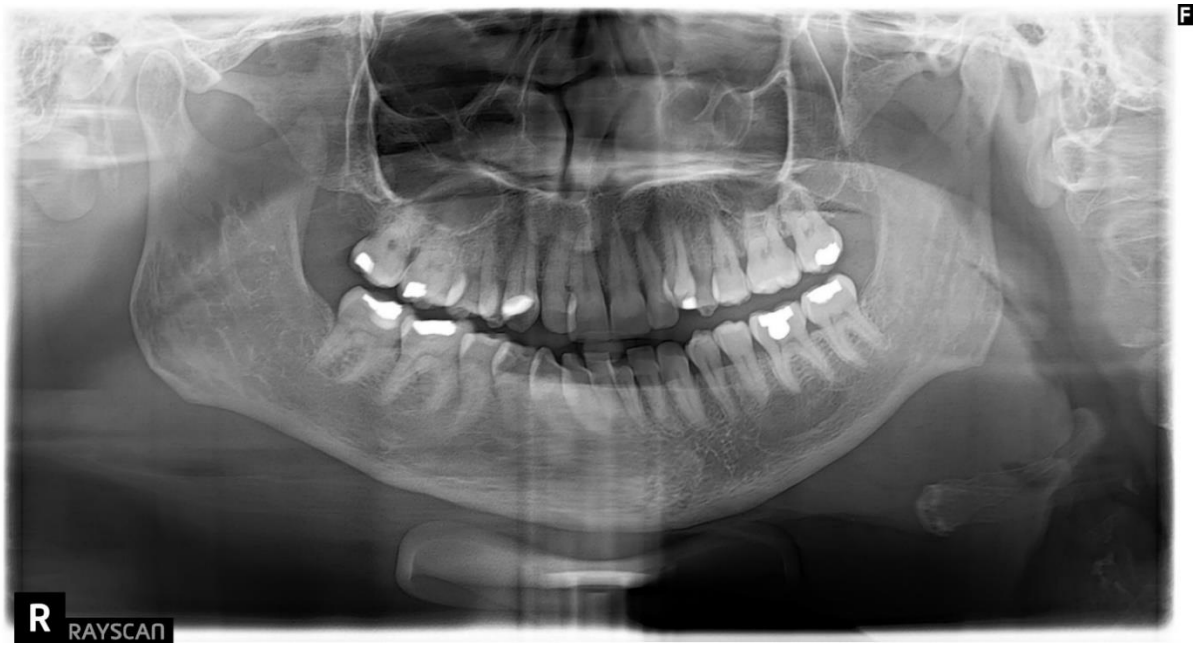

Figure 9: Panoramic radiograph 6 months post-op.

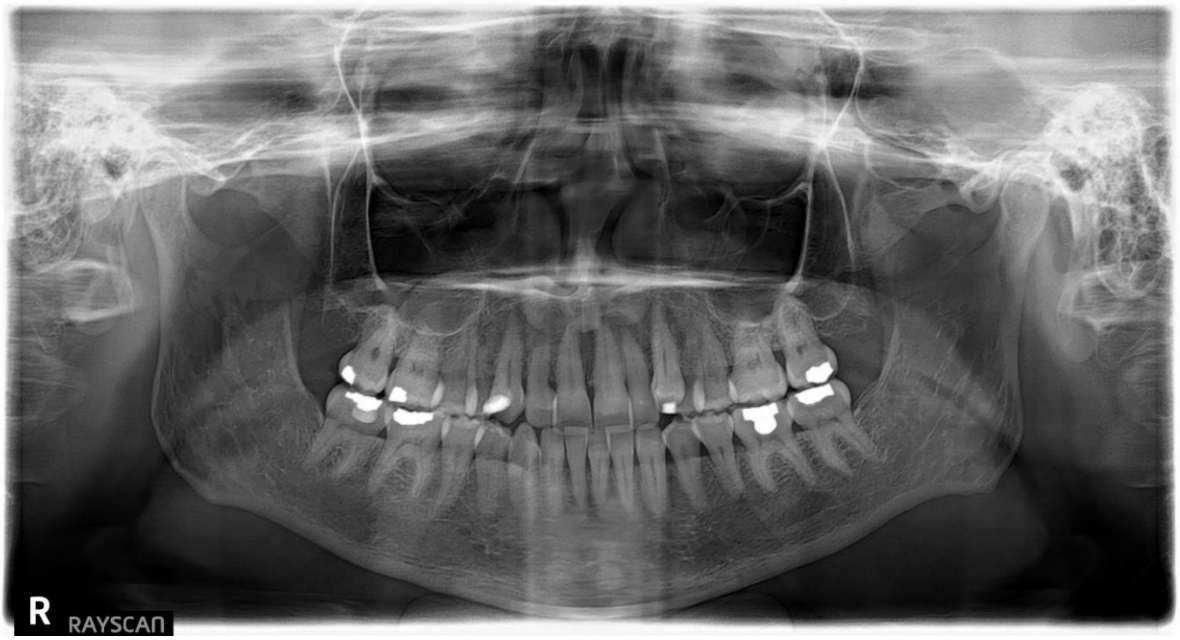

Figure10: Panoramic radiograph 20 months post-op.

DISCUSSION

Schwannomas are classified as one of the most common benign peripheral nerve sheath neoplasms [1]. They are known to most commonly affect the head and neck but can also be present in other parts of the body, such as the extremities. The average age range for the development of schwannoma is the 6th to 7th decade of life, which is consistent with the patient presented in this report [2]. Clinically, schwannomas present with swelling in the bone with or without the presence of pain and paresthesia. The patient presented in this case did have noted expansion but did not have any history of pain or paresthesia. Classical radiographic features of a schwannoma include

external root resorption, cortical thinning, spotty calcifications, cortical expansion, and peripheral scalloping [3]. The radiographic findings in this report included each of these features.

As its name suggests, schwannomas are derived from Schwann cells, which are a type of glial cell. Schwann cells are ultimately derived from neural crest cells and function to deposit myelin onto neurons of the peripheral nervous system [4]. This allows for rapid conduction of electrical impulses along the nerve via Nodes of Ranvier.

In the oral cavity, schwannomas tend to appear in the soft tissue of the floor of the mouth or tongue. Although extremely rare, schwannomas can also present in the bones of the face, such as the mandible or maxilla. One author presenting a case report on intraosseous schwannoma of the mandible in 2011 noted there only being 45 reported cases at that time [5]. When present in the jaws, these tumors present as a unilocular radiolucency, which often demonstrates bone and tooth root resorption in the affected areas [6].

In addition to schwannoma, there exist other peripheral nerve sheath tumors, such as neurofibroma, perineuroma, traumatic neuroma, and malignant peripheral nerve sheath tumor [7]. Proper diagnosis is therefore of utmost importance. This is typically achieved histologically and clinically. Histologically, schwannomas, are made up completely of Schwann cells that are contained within a capsule. Furthermore, schwannoma tends to be attached to the parent nerve peripherally [6,7]. To further differentiate schwannoma from other lesions, schwannoma typically presents with distinct histologic S100+ staining (Figure 11) and unique histologic architecture consisting of Antoni A tissue with associated Verocay bodies as well as Antoni B tissue [3,7]. Antoni A tissue, named after the neurologist, Nils Ragnar Eugène Antoni, contains highly cellular regions with tightly packed spindle-shaped cells. They are typically demarcated by elongated nuclei in two rows of palisades with a central acellular zone, which are referred to as Verocay bodies (Figure 12) [8]. These palisades were initially identified and subsequently named by Jose Juan Verocay, who discovered them in 1910.7 Antoni B zones, in contrast to Antoni A zones, are loosely organized with myxomatous and cystic changes. These areas may represent degenerated Antoni A tissue. Most schwannomas show varying proportions of both Antoni A and Antoni B tissue which are discrete and separate from one another; however, some regions may show a transitional zone with a merger between the two tissue types [8].

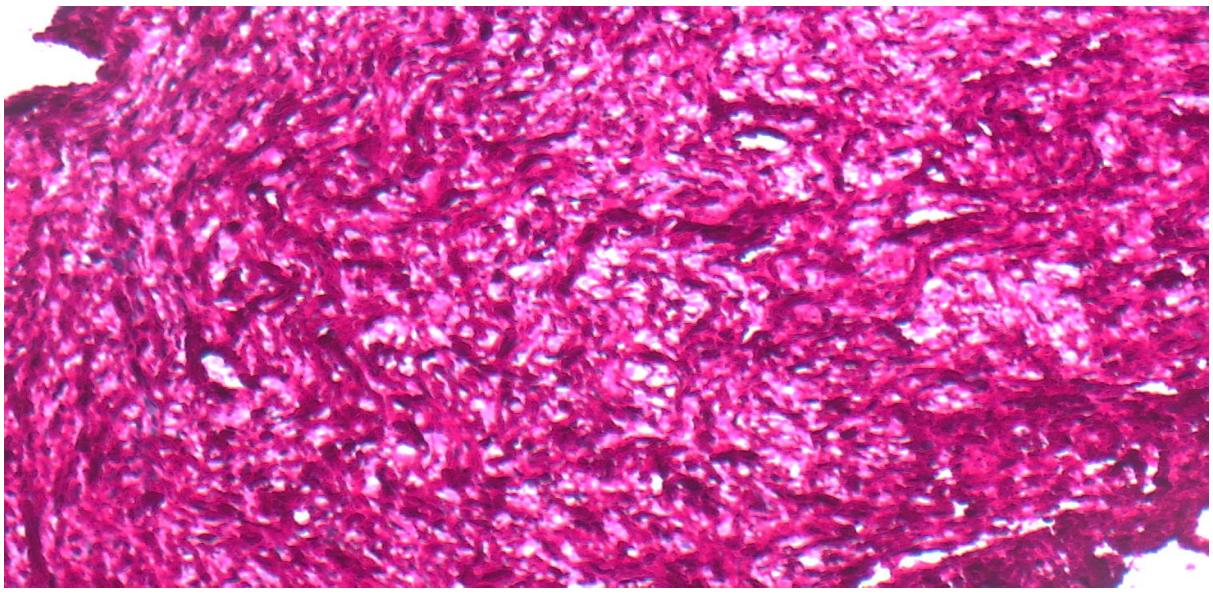

Figure 11: Histologic photomicrograph – power x200 demonstrating S100+ staining.

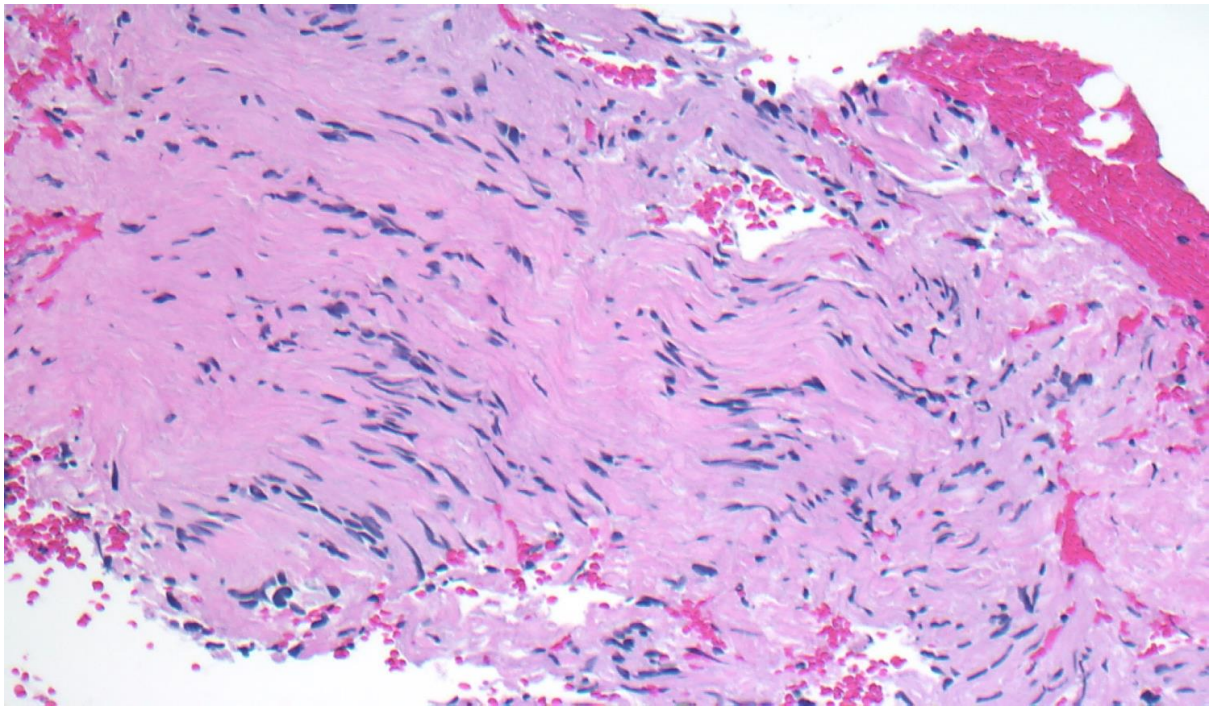

Figure 12: Histologic photomicrograph – power x200 H & E stain demonstrating the palisading pattern of Antoni A zones.

Schwannomas, originally called neurilemmoma, are benign and slow growing peripheral nerve sheath neoplasms originating from Schwann cells. They can be found in both soft tissue and bone. When found within bone, they are referred to as intraosseous, or central, schwannoma. They have characteristic histopathologic features that suggest their diagnosis, which include Antoni A and B tissue with S100+ immunohistochemical staining. In addition to schwannoma, there are multiple other peripheral nerve sheath tumors, one of which is malignant. Understanding the clinical, radiographic, and histopathologic features of schwannoma are paramount in rendering a proper diagnosis. For schwannoma, conservative excision is the treatment of choice and is associated with a near 100% cure rate.6 Due to the rarity of intraosseous schwannomas of the mandible and maxilla, further research needs to be conducted to fully understand this subset of lesion and the pathological process inciting its formation.

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