Intraosseous neurilemoma of the mandible with unusual multilocular presentation: a case report

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This case report describes an intraosseous neurilemoma, observed radiographically as a multilocular lesion, in a 12-year-old patient. Physical examination revealed facial asymmetry, swelling on the right side of the mandibular body, and bone expansion in the region of the base of the buccal sulcus. Panoramic radiography revealed a multilocular radiolucency on the right side of the mandibular body in the periapical region extending from the distal region of the first premolar root to the second molar and adjacent to the third molar. Evaluation of a radiograph obtained 3 years earlier for an orthodontic assessment revealed that the lesion was present, appearing as a unilocular radiolucency near the root of the mandibular right first molar. An incisional biopsy of the multilocular lesion was performed, and Antoni A and Antoni B histologic patterns were identified by microscopic analysis. Immunohistochemical analysis was conducted, and neoplastic cells stained positive for the S-100 protein. The patient underwent conservative surgical excision of the lesion, and no recurrence was observed during 7 years of clinical follow-up. Based on analysis of the present case and previous cases reported in the literature, intraosseous neurilemoma, especially its multilocular variant, is an uncommon neoplasm. In the present case, evaluation of the lesion when it was first radiographically detectable, prior to orthodontic treatment, would have permitted a more limited surgical approach for the excision of a small intraosseous lesion.

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Neurilemoma is a benign neoplasm derived from Schwann cells, which form the myelin sheaths that surround the nerves.[1-8] This tumor was first described by Verocay in 1910.[9] Studies indicate that 24% to 48% of neurilemomas are found in the head and neck region.[7,10-13] Most tumors affecting the soft tissues, especially the tongue, are asymptomatic and grow slowly, and intraosseous lesions are uncommon.[1,4,6-8,14-18] Neurilemomas typically develop in the second or third decade of life, and a slight female predilection has been reported.[1,6-8,19,20] The indicated treatment is conservative surgical removal. After complete excision, recurrence is uncommon because of encapsulation of the tumor.[1,6,7,19-23]

The present case report describes an uncommon case of intraosseous neurilemoma involving the mandibular body and discusses the findings in light of the literature on the subject. The objective of this report is to describe the clinical, imaging, and histologic characteristics and treatment of an uncommon case of intraosseous neurilemoma that developed in the mandible of a 12-year-old girl under orthodontic follow-up whose lesion had already been detected during initial orthodontic assessment.

Case report

This report was approved by the Ethics Committee of the Institute of Science and Technology, São Paulo State University, São Paulo, Brazil. All procedures performed were in accordance with the ethical standards of the Helsinki declaration on medical research involving human subjects. The patient's parent signed an informed consent agreement.

A 12-year-old girl presented at the outpatient clinic of the Department of Biosciences and Oral Diagnosis, Institute of Science and Technology, São Paulo State University, São José dos Campos, Brazil, reporting pain in the right posterior region of the mandible. During history-taking, the patient reported no systemic alterations, surgeries, or hospitalizations. However, she had been undergoing orthodontic therapy for 3 years, including monthly follow-up examinations by the dentist providing treatment.

Facial asymmetry and swelling on the right side of the body of the mandible were observed during the extraoral physical examination (Fig 1). The submandibular lymph nodes were mobile, soft, regular, and painless. Intraoral examination revealed bone expansion in the region of the base of the vestibular sulcus (Fig 2). The mucosa lining the sulcus appeared normal, but crepitant areas were detected on palpation. The patient used a fixed orthodontic appliance.

Imaging examinations were requested. Panoramic radiography revealed a multilocular radiolucency on the right side of the mandibular body in the periapical region extending from

Fig 1. Facial asymmetry and swelling on the right side of the body of the mandible.

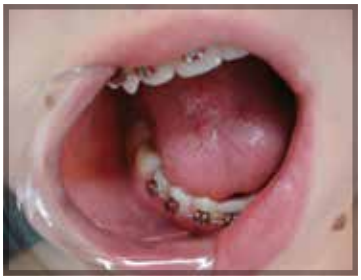

Fig 2. Bone expansion in the base of the right buccal sulcus.

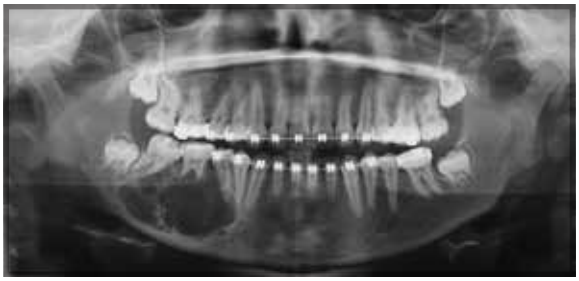

Fig 3. Multilocular radiolucency extending from the first premolar to the second molar on the right side of the mandibular body.

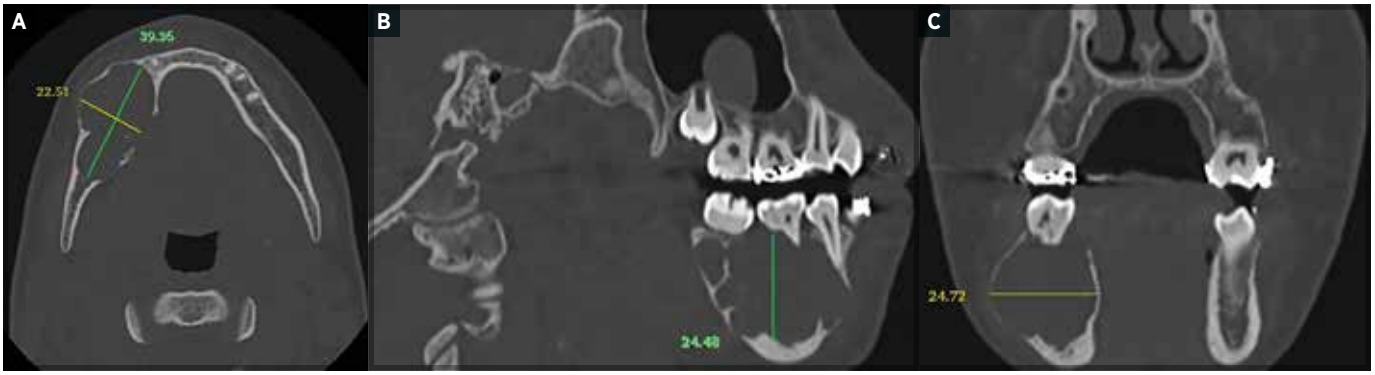

Fig 4. Cone beam computed tomograms showing a multilocular, hypodense lesion (dimensions indicated in millimeters) causing expansion, thinning, and areas of rupture in the buccal and lingual bone plates. A. Axial section. B. Sagittal section. C. Coronal section.

the distal region of the first premolar root to the second molar and adjacent to the third molar (Fig 3). Computed tomography showed a multilocular, hypodense lesion causing expansion, thinning, and areas of rupture in the buccal and lingual bone plates (Fig 4). The mandibular right third molar was situated adjacent to the lesion but was not affected. In addition, a panoramic radiograph obtained 3 years earlier, before the start of orthodontic treatment, was evaluated (Fig 5). The lesion was present at that time, appearing as a unilocular radiolucency near the root of the mandibular right first molar.

Based on the clinical and imaging findings, the differential diagnosis consisted of ameloblastoma and keratocystic odontogenic tumor. For establishment of the final diagnosis, exploratory puncture was performed, followed by an incisional biopsy. The exploratory puncture indicated a solid lesion. Because ameloblastoma was included in the differential diagnosis, the incisional biopsy was performed by means of a full-thickness intrasulcular mucoperiosteal flap followed by detachment for bone exposure. A bone window was opened, and tissue was collected from the lesion via curettage to obtain a representative sample. Histopathologic examination of the specimen revealed the fragment of a partially encapsulated neoplasm of neural origin intermingled with dense connective tissue. The neoplasm was formed by lobules consisting of Schwann cells, which were organized either in palisades with central acellular eosinophilic areas (Antoni A pattern) or in a random arrangement (Antoni B pattern).

Immunohistochemical analysis of the specimen was performed, and the neoplastic cells stained positive for the S-100 protein. Staining with the Ki-67 protein revealed the absence of intense proliferative activity. Based on the histologic data, the final diagnosis was intraosseous neurilemoma.

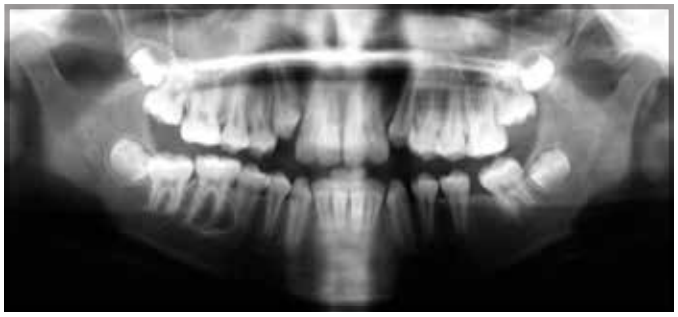

Fig 5. Panoramic radiograph obtained 3 years earlier, prior to orthodontic treatment, showing a unilocular, radiolucent lesion near the root of the mandibular right first molar.

The patient underwent conservative surgical excision of the lesion and extraction of the involved teeth from the second premolar through the third molar. She was reexamined at regular intervals after surgery. No recurrence was observed in 7 years of clinical follow-up (Fig 6).

Discussion

Neurilemoma, also called *schwannoma*, is a benign neoplasm originating from Schwann cells, which form the myelin sheath surrounding the axon.[2,5,7,20,24] The etiology of this tumor is

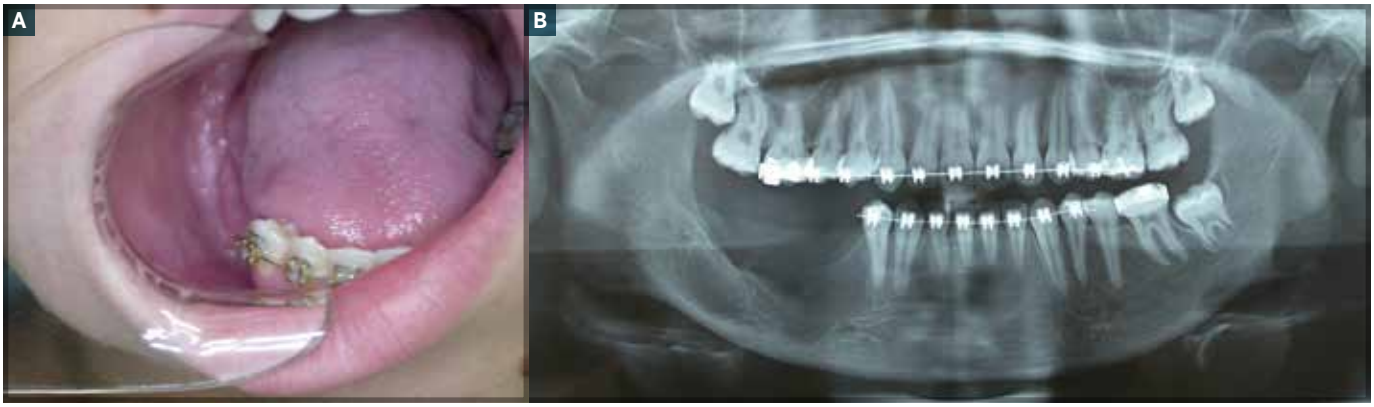

Fig 6. Imaging obtained at the 7-year follow-up, confirming that the lesion had not recurred. A. Intraoral view. B. Panoramic radiograph.

Table. Cases of intraosseous neurilemoma involving the mandible and maxilla.

	Patient		Lesion[a]		
Study	**Age, y**	**Sex**	**Radiographic features**	**Signs and symptoms**	**Location**
Villanueva et al (1995)[27]	14	F	Radiolucent, unilocular, circumscribed lesion	None	Anterior maxilla
Buric et al (2009)[1]	23	F	Radiolucent, unilocular, circumscribed lesion	Bone expansion and pain	Right posterior mandible
Chi et al (2003)[19]	16	F	Radiolucent, unilocular, circumscribed lesion	External root resorption of tooth adjacent to lesion	Anterior mandible
de Lacerda et al (2006)[21]	11	M	Radiolucent, unilocular, circumscribed lesion	Bone expansion, pain, and paresthesia	Anterior mandible
Hsieh et al (2009)[18]	54	M	Radiolucent, unilocular, circumscribed lesion	Tooth mobility of first molar and external root resorption of molars	Right posterior mandible
Jahanshahi et al (2011)[6]	11	F	Radiolucent, unilocular, circumscribed lesion	Bone expansion	Right posterior mandible
Zhang et al (2012)[7]	35	M	Radiolucent, bilocular, circumscribed lesion	Bone expansion and mild paresthesia on right side of lower lip	Right posterior mandible
	29	F	Radiolucent, unilocular, circumscribed lesion	Bone expansion	Left posterior mandible
Lambade et al (2013)[22]	33	M	Radiolucent, unilocular, circumscribed lesion	Bone expansion	Anterior mandible
Suga et al (2013)[8]	33	M	Radiolucent, unilocular, circumscribed lesion	Bone expansion	Anterior mandible
Meundi et al (2015)[20]	20	F	Radiolucent, unilocular, circumscribed lesion	Bone expansion	Anterior maxilla

Abbreviations: F, female; M, male.

[a]In every case, the lesion was treated with surgical excision.

unknown.[2,20] Neurilemomas grow slowly and may therefore be present in the oral cavity for years without manifesting major symptoms other than swelling.[16] Generally, the nerve that gives origin to the tumor is responsible for the symptoms.[25]

No sex predilection for neurilemoma has been reported, although the lesions are slightly more prevalent in women, with a female to male ratio of 1.6:1.[2,5,7,19,20] This predilection increases in the case of intraosseous neurilemoma, which has a female to male ratio of 4:1.[7,20] Neurilemomas are more common in the second decade of life but can affect all age groups.[2,5,7,12,20,23] Studies indicate that 24% to 48% of neurilemomas are found in the head and neck region.[7,10-13] The tongue, hard palate, floor of the mouth, buccal mucosa, mandible, and submandibular region are the most affected sites.[2,5,7,20,25]

Clinically, most neurilemomas appear in soft tissues as single, submucosal, well-circumscribed, fixed, painless nodules that are yellow, blue, white, or mucosa colored, measure 1 to 5 cm, and evolve over a period of 3 months to 5 years.[2,5,7,20] The clinical differential diagnosis of neurilemoma includes fibroma, lipoma, and benign salivary gland tumors.[24]

Central neurilemomas primarily affect the mandible and are rare in the maxilla, probably because of the extensive intraosseous path of the inferior alveolar nerve.[7,26,27] In the mandible, neurilemomas generally affect the posterior region but can also be found in the anterior region.[19] Bone expansion at the site is the main complaint of patients. With the development of the tumor and consequent volume increase, the nerve fibers on the tumor capsule may be compressed, resulting in paresthesia.[21] However, the affected teeth remain vital.[20]

Definitive diagnosis of neurilemoma is made through microscopic analysis of a specimen obtained by incisional or excisional biopsy and submitted to histochemical and immunohistochemical staining.[5,19,28] In addition to the combination of panoramic radiography and computed tomography with 3-dimensional reconstruction, periapical and occlusal radiography, as well as aspiration biopsy, can be used for the diagnosis of this tumor.[7,20] Magnetic resonance imaging is another method used to diagnose neurilemomas in soft tissue.[2]

Neurilemomas exhibit diverse histologic features. Most tumors possess a capsule, beneath which 2 associated histologic patterns can be observed. The Antoni A pattern consists of closely packed Schwann cells with elongated nuclei that form bundles and are arranged in palisades. The acellular areas between the rows of nuclei are called *Verocay bodies*.[9] In the Antoni B pattern, the Schwann cells are loosely arranged, can be elongated, spindle-shaped, or stellate, and exhibit fewer cellular areas and a higher proportion of connective tissue.[7,19,29-32] Immunohistochemical markers such as S-100 and Leu-7 can complement the histologic diagnosis.[32] The S-100 protein is recognized as a good marker for confirmation of this tumor because it is expressed by Schwann cells.[33]

The recommended treatment for neurilemoma is conservative surgical removal.[1,22,25] If the nerve of origin is visible during the procedure, it should be carefully separated to preserve its function, although this may not always be possible.[25,34] The prognosis for neurilemoma is good, and recurrence is unlikely, as is malignant transformation.[11,24] However, it is important to continue follow-up examinations of the patient for more than 1 year.[33]

The present report describes an uncommon clinical case of intraosseous neurilemoma in a young girl. The Table presents the age and sex of affected patients as well as the radiographic features, main signs and symptoms, and locations of intraosseous neurilemoma reported in the literature over the last 15 years.

The patient in the present report complained of pain and exhibited bone expansion on the right posterior area of the mandible. Buric et al and de Lacerda et al also reported cases in which the patients reported painful symptoms, while the patients described by Lambade et al and Jahanshahi et al did not complain of pain.[1,6,21,22] The presence of paresthesia has also been reported, and swelling is a common sign observed in the cases described in the literature.[1,6-8,19-22]

In the present case, the neurilemoma was located on the right side of the mandibular body in the periapical region and extended from the first premolar to the second molar. Buric et al, Jahanshahi et al, and Zhang et al also described neurilemomas located in the posterior mandible.[1,6,7] However, different locations, including the anterior mandible, have been reported in the literature.[8,19,21,22] Neurilemomas in the anterior maxilla were described by Meundi et al and Villanueva et al.[20,27]

Radiographically, the present case exhibited characteristics of a multilocular lesion, while radiolucent and circumscribed images of this tumor have been reported in the literature.[1,6,8,19-22,27] Only Zhang et al described a bilocular lesion in a case they reported in 2012.[7] It should be noted that the lesion observed on the initial radiograph of the girl in the present case, prior to orthodontic treatment, did not show a direct relationship with the bundle of the inferior alveolar nerve.

The histologic features observed in the present case are characteristic of neurilemoma and agree with those reported in the literature.[7,19,29-32] The immunohistochemical analysis demonstrating positive staining for S-100 confirmed the neural origin and definitive diagnosis of neurilemoma, as reported in the literature.[1,3-5,7,19,22,35] In addition, the observed low reactivity to Ki-67, which demonstrates low proliferative activity, was compatible with the benign nature and slow growth of the tumor.[36]

The patient in the present report underwent conservative surgical excision of the tumor and extraction of the involved teeth and remains on a regular recall schedule for follow-up. This approach is consistent with the treatment recommended in the literature.[1,6-8,19-22,27]

Conclusion

Based on the analysis of the present case as well as previous cases described in the literature, it can be concluded that intraosseous neurilemoma, especially its multilocular variant, is an uncommon neoplasm. In the present case, evaluation of the lesion when it was first radiographically detectable, prior to orthodontic treatment, would have permitted a more limited surgical approach for the excision of a small intraosseous lesion.

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Disclaimer

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