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CASE REPORT

Diagnostic challenges of an unusually large schwannoma of the mandible: Report of a case

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Abstract Schwannomas are slow-growing, benign neoplasms arising from the Schwann cells and are commonly reported as peripheral tumors in the head and neck region. Central intramandibular schwannomas are extremely rare lesions. We report a case of intramandibular schwannoma in a 70 year old male patient. Panoramic radiography revealed a large, multilocular radiolucent lesion with distinct borders involving the right mandibular body and ramus. A complete excision was achieved by removing the tumor followed by reconstruction of the mandible. The clinical, radiological, and histopathological features are discussed within the context of this case.

1. Introduction

Schwannomas or neurilemmomas are slow-growing, benign neoplasms commonly seen as peripheral tumors in the head and neck region and are derived from Schwann cells, the sheath cells that cover myelinated nerve-fibers (Belli et al., 1997; Zhang et al., 2012). These rare, neoplasms arise from any cranial, peripheral, or autonomic nerve that contains Schwann cells. Schwannomas of the head and neck account for approximately 25–45% of extracranial Schwannomas reported (Colreavy et al., 2000).

Although Schwannomas of the bone are rare, they are capable of affecting any bone. While those neoplasms arising from the medullary cavities of the bone and mimicking primary bone tumors are defined as intraosseous schwannomas, they may be termed according to the bone of origin, when originating from the nerves housed within canals and foramina in the bone (Gordon, 1976; Ida et al., 2011). Schwannomas of the jaw are extremely rare and only forty-nine cases of intramandibular schwannoma have been reported so far in the indexed literature (DeLeonibus et al., 2017). They are usually described as asymptomatic, encapsulated tumors which typically arise in association with a nerve trunk and push aside the nerve along with adjacent structures as they grow (Colreavy et al., 2000). Consequently, this causes enlargement of the canal housing

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the neurovascular bundle. Swelling due to bony expansion is the most commonly reported symptom among patients with the intramandibular variant of schwannoma. Nevertheless, nearly fifty percent of the cases are reportedly associated with pain and paresthesia in addition to bony expansion (Park et al., 1999).

Radiologically, intramandibular schwannoma presents as a unilocular or multilocular well-defined radiolucency. In the head and neck region, they have primarily been reported to occur in the posterior part of the mandible, mimicking other benign conditions such as ameloblastoma or odontogenic keratocyst (Park et al., 1999). The objective of this paper is to present a case of an extensive intramandibular schwannoma involving the right posterior mandible, which was surgically treated by resection of the tumor along with a recurrence-free follow up period of three years. This case is not only being reported for its differential diagnosis with common odontogenic lesions, but also because it is one among the first of its kind to be reported from Saudi Arabia.

2. Case report

A 70 year old male patient reported to the out-patient clinic of a tertiary level dental hospital in Riyadh, Saudi Arabia, with the history of a slow-growing painless swelling in the right mandible over the past one year. There was no associated history of trauma or infection at the site of the lesion. The patient presented a history of medical management for type 2 diabetes mellitus and hypertension, with oral hypoglycemic and antihypertensive medications respectively. Clinical examination revealed facial asymmetry with mild swelling over the right mandibular ramus. The skin overlying the swelling was unremarkable in color, texture and consistency. Upon intra-oral examination, the patient had an edentulous right mandibular molar area with buccolingual expansion of the mandibular cortex, extending posteriorly up to the mandibular ramus. On palpation, the swelling was non-tender, non-compressible and the overlying mucosa was apparently normal. The second premolar was the distal most tooth in the right mandibular quadrant and it demonstrated clinical signs of gingival recession without any associated tenderness or mobility (Fig. 1).

A panoramic radiograph revealed well defined, corticated, multilocular radiolucent lesion involving the right mandible extending anteroposteriorly from the mesial aspect of the lower right second premolar up to the posterior border of the right mandibular ramus, and from the condylar neck and coronoid process to the inferior border of the mandible superoinferiorly. Radiographic evidence of expansion and thinning of the cortical boundaries of the mandible were appreciated. While a thin layer of bone was seen in the superior and inferior borders of the mandible, the mandibular canal was obliterated or could not be appreciated radiographically (Fig. 2a).

Intravenous contrast enhanced computed tomography (CT) scanning of the involved regions revealed a radiolucent lesion causing expansion and thinning of the buccal and lingual cortices of the right mandibular body and ramus, along with multiple sites of cortical perforation. Although the mandibular canal could not be seen, the mandibular foramen was expanded and the mental foramen appeared normal. Contrast enhancement of the lesion was not observed in CT. More proximally, the lesion was not only seen widening the mandibular foramen, but also was seen expanding into the

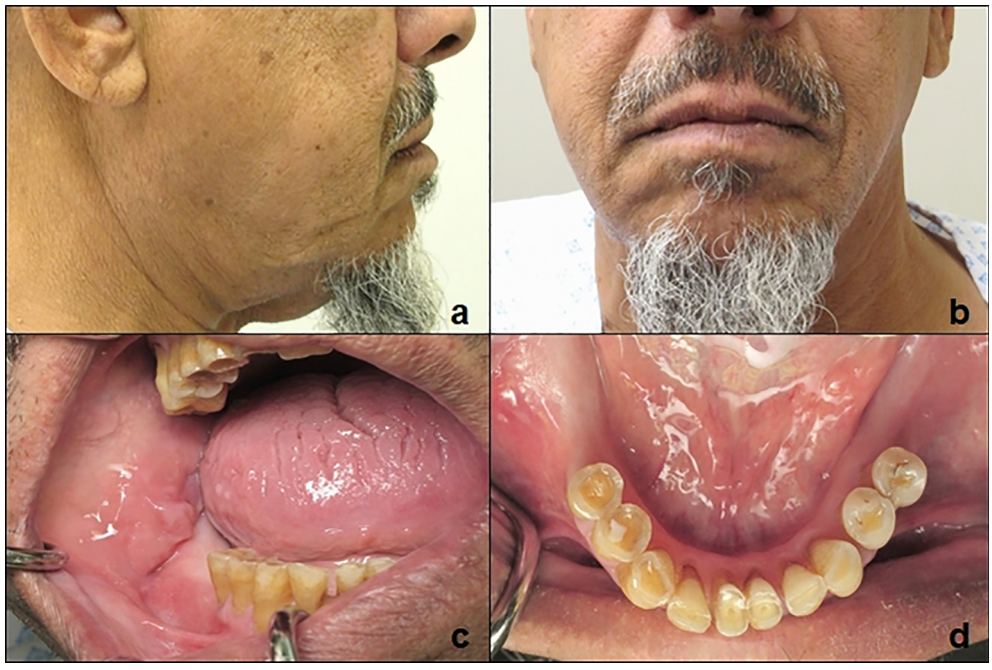

Fig. 1 Pre-operative clinical presentation of the patient. **(a)** Right lateral extra-oral view of the mandibular body and ramus region. **(b)** Frontal extra-oral view of the maxillo-mandibular region revealing mild facial asymmetry on the right side. **(c)** Intra-oral view of the right mandibular quadrant showing a partially edentulous arch and minimal clinical swelling due to bony expansion and normal overlying mucosa. **(d)** Intra-oral occlusal view of the mandibular arch showing asymmetrical bony expansion on the right side in comparison to the left side.

right pterygomandibular (PM) space and displacing medially, the anatomic structures present within the prevertebral space. CT Sections at the level of the cranial base showed symmetrical anatomical structures such as the foramen ovale and Meckel's cave, thereby ruling out any intracranial extension of the lesion (Fig. 2b and c).

Incision biopsy under local anesthesia yielded a small and fragmented specimen, which upon histological examination, was mostly solid and not cystic. However, diagnosis of a lesion of cystic nature was suggested based on the panoramic radiographic image, which was available at the time of initial biopsy. A histological diagnosis of degenerated or ancient schwannoma was not considered due to the lack of features such as degenerative atypia, pseudocystic spaces and calcifications. Although no immune stains were performed on the initial biopsy specimen, more representative tissue was requested for conclusive histopathological diagnosis.

Considering the anatomical extent of the tumor to the PM space, bone thinning and the clinically aggressive nature of the lesion, total resection of the tumor by right hemimandibulectomy with disarticulation of the condyle was planned and explained to the patient along with different reconstruction modalities. The patient consented to the surgical procedure for tumor excision and reconstruction using a reconstruction plate along with prosthetic condyle.

Pre-operatively, the patient's medical condition was optimized for general anesthesia (GA) and surgery. Under GA, a submandibular approach with retromandibular extension was utilized to expose the affected mandible. The reconstruction plate was adapted and screw sites were drilled to indicate their location post resection. Subsequently, tumor resection was done and immediate reconstruction of the mandible was achieved using the pre-adapted reconstruction plate (Fig. 3). The postoperative period was uneventful and the patient was discharged 7 days postoperatively. Long-term follow-up of the patient for three years showed no clinical or radiological signs of recurrence of the lesion (Fig. 3d).

Gross pathological findings revealed a surgically resected lesion involving the right mandible extending from the mesial aspect of the right mandibular second premolar, in the body of the mandible, up to the condyle. The specimen measured 11.5 cm × 7.0 cm × 1.5 cm, along with bony expansion in the buccal and lingual sides of the mandible. The inferior alveolar neurovascular bundle passing through the eroded mandibular foramen was elastic in consistency and brownish-white in color (Fig. 3). Microscopically, hematoxylin and eosin stained

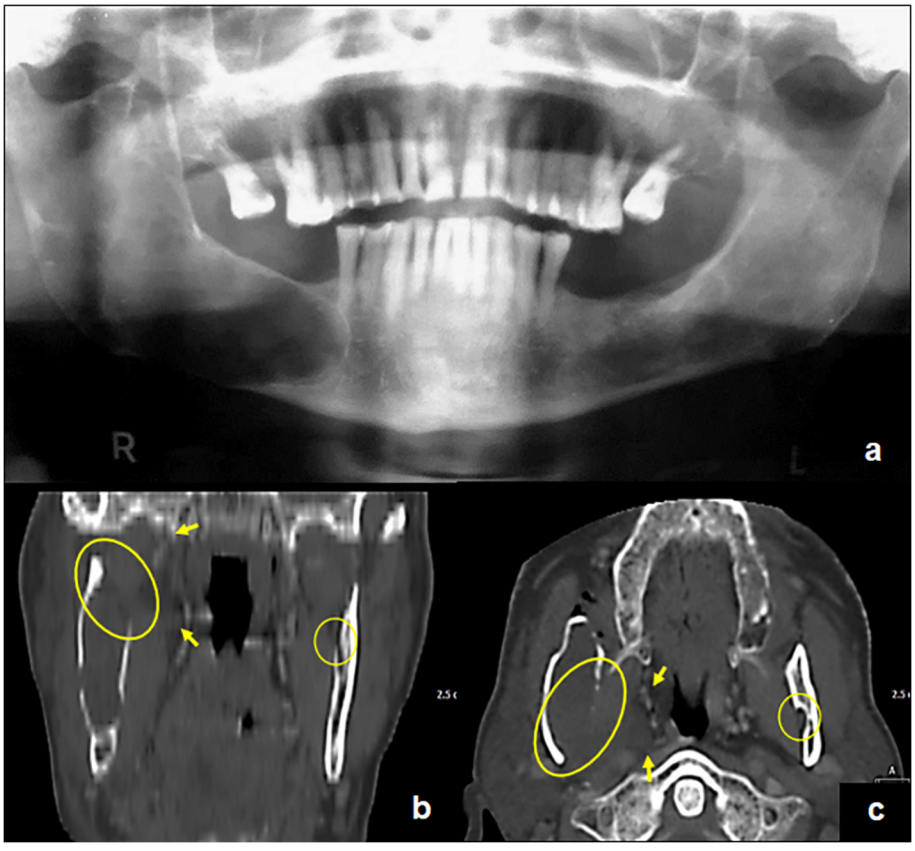

Fig. 2 Pre-operative radiographic evaluation of the patient. **(a)** Panoramic radiograph revealing multilocular radiolucent lesion involving the right mandibular body and ramus, extending anteroposteriorly from the second premolar to the entire mandibular ramus. Obliteration of the mandibular canal could be appreciated. **(b)** Contrast enhanced computed tomographic (CT) image showing a coronal section through the lesion as it extends across the mandibular foramen into the pterygomandibular space. Enlargement of the mandibular foramen (marked with yellow circles) on the right side in comparison to the unaffected left side can be appreciated clearly. **(c)** Contrast enhanced CT image showing an axial section through the lesion as it extends across the mandibular foramen into the pterygomandibular space. Enlargement of the mandibular foramen (marked with yellow circles) on the right side can be appreciated in this CT section too.

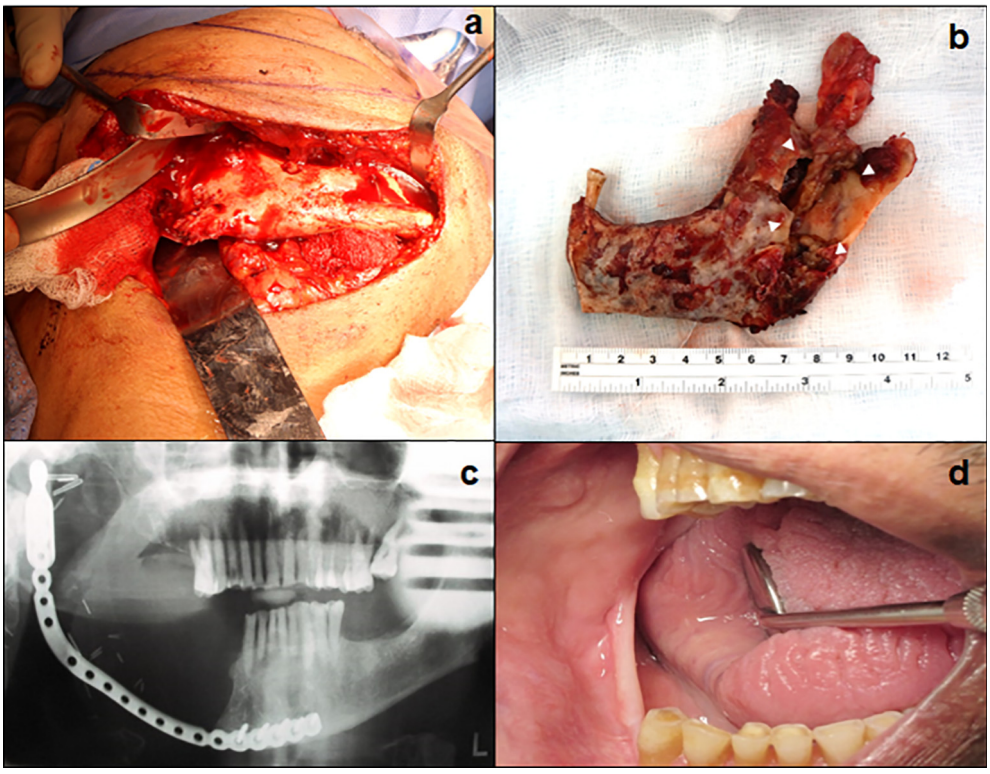

Fig. 3 Intra-operative and post-operative clinical and radiographic presentation. **(a)** Surgical approach to the tumor through an extended sub-mandibular incision and exposure of the tumor. **(b)** Resected specimen of the tumor involving the right mandibular body and ramus regions showing areas of cortical perforation on the lingual side and the inferior alveolar neurovascular bundle along with enlargement of the mandibular foramen (white arrows). **(c)** Panoramic radiograph showing the mandible, post resection of the tumor and reconstruction using a reconstruction plate. **(d)** Intra-oral view of the right mandibular quadrant 3 years postoperatively showing satisfactory mucosal coverage at the resection site, acceptable mouth opening and no clinical signs for lesion recurrence.

sections revealed multiple fragments of soft tissue consisting of interlacing fascicles of spindle-shaped cells with indistinct cytoplasmic borders in moderately fibrous stroma containing small to medium sized ectatic blood vessels. In focal areas, the tumor cells exhibited palisading of the nuclei characteristic of "Antoni type A" pattern. However, the majority of the tumor cells were haphazardly arranged within the fibrillar stroma (Antoni type B), along with interspersed Verocay bodies. The neoplastic cells were strongly positive for S100, vimentin, CD56 and negative for cytokeratin (Fig. 4). The histopathological and immunohistochemical features were in support of a diagnosis of schwannoma.

3. Discussion

In this report, we present the clinical, radiographic and surgical management of a case of an extensive intramandibular schwannoma in a 70 year old male patient.

Schwannoma is an infrequent, benign, encapsulated tumor arising from the Schwann cells. They are often reported during the fourth and fifth decades of life and exhibit a 1.6:1 female predilection (Subhashraj et al., 2009; Gainza-Cirauqui et al., 2013). Around 25–45% of all schwannomas reported, are from the head and neck region, out of which, the incidence of oral cavity schwannomas is between 1 and 12%, with a preponderance towards the dorsal and ventral surfaces of the tongue and the buccolabial vestibule (Jahanshahi et al., 2011; Buric et al., 2009). Intramandibular schwannomas are extremely rare, mainly occurring in the posterior areas of the body and the ramus of the mandible (Nakasato et al., 2000).

Schwannomas within the mandible may mimic benign odontogenic tumors in their clinical and radiographic presentation (Gallego et al., 2009). Towards establishing a diagnosis, routine radiographs are of little significance because they do not possess any features that distinguish schwannoma from other lesions. However, CT imaging has enabled better diagnosis of schwannomas of the head and neck region, wherein intramandibular and maxillary schwannomas demonstrate a well-demarcated, unilocular or multilocular radiolucency with a thin, sclerotic border (Zhang et al., 2012). MRI scans provide much more valuable information for pre-operative diagnosis and treatment planning (Sun et al., 2011). In the present case, the tumor was initially diagnosed as an ameloblastoma based on its clinical presentation and plain radiographs. However, CT imaging revealed a unilocular radiolucency with remarkable enlargement of the lesion above the mandibular canal extending posterosuperiorly into the PM space. This gave rise to the suspicion towards an ameloblastoma with intraneural spread or a tumor of neurological origin. The need for an MRI imaging in the present case was ruled out due to the greater radiographic detail obtained using contrast enhanced CT imaging and the absence of any evidence of intracranial extension.

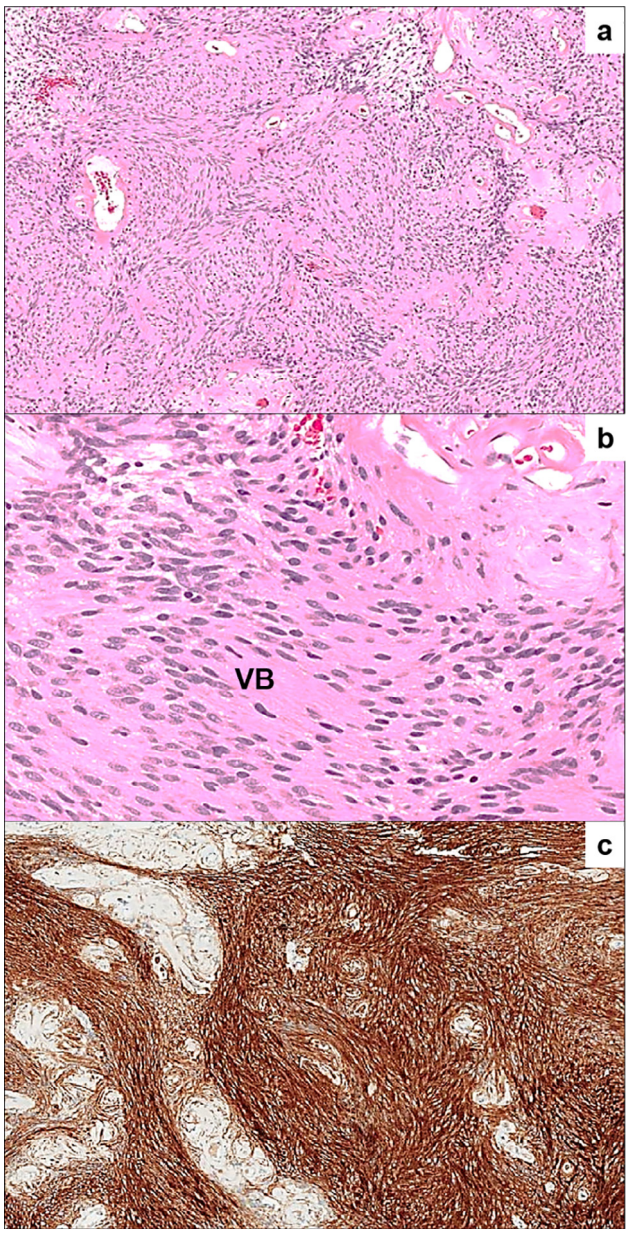

Fig. 4 Histopathological findings. Hematoxylin and eosin stained sections reveal spindle cell tumor with prominent focal palisading, "Antoni A" and interspersed eosinophilic incomplete Verocay bodies (VB) (**a**, ×20, **b**, ×40). Almost all of the tumor cells demonstrated strong positivity for anti-S100 and anti-CD56 (**c** ×40).

Intramandibular schwannomas have been reported to be associated with mobility or displacement of adjoining teeth and clinical symptoms ranging from pain to paresthesia (Zhang et al., 2012). In the present case, there were no clinical signs of pain or paresthesia reported by the patient, who apparently had noticed the swelling only in the preceding year. Nevertheless, no clear history regarding the status of previously extracted right mandibular posterior teeth was available. The right mandibular second premolar, the only tooth clinically and radiographically associated to the tumor, exhibited gingival recession without any tenderness to percussion or mobility.

In addition to the classic schwannoma, several histopathologic variants such as the cellular, plexiform, epithelioid, ancient and melanotic types have been reported (Zhang et al., 2012). Histological features characteristic of schwannomas include encapsulation, small eosinophilic masses called "Verocay bodies" seen in a fibrillary stroma, palisading nuclei, and Antoni type-A and B tissues. The Verocay bodies are typically characterized by two compact rows of well-aligned nuclei separated by eosinophilic fibrillar cell processes (Goldblum et al., 2013). Isolated Antoni type-A and B tissues, and alternating Antoni-A and B regions are the two types of tissue arrangement evidenced histologically in schwannomas. Typically palisaded, the Antoni type-A tissues are made up of aligned fusiform cells. On the other hand, Antoni type-B tissues are composed of a smaller number of spindle cells, randomly arranged within a loose myxomatous stroma (Asaumi et al., 2000; de Lacerda et al., 2006).

It is imperative to use immunohistochemistry techniques while diagnosing suspected neural tumors like schwannomas, as these tumor cells typically exhibit positivity for S-100 protein (Chrysomali et al., 1997). Schwannomas present a strong positive immunoreactivity to S-100 protein in almost all tumor cells, in contrast to other neural tumors like neurofibromas or peripheral nerve sheath tumors (Asaumi et al., 2000). In the present case, the majority of the tumor cells in the resected specimen displayed histologic characteristics of schwannoma such as Antoni type-B tissues with focal areas of Antoni type-A arrangement. Moreover, the neoplastic cells were strongly positive for S-100, vimentin, CD56 and negative for cytokeratin, which were all in support of a diagnosis towards schwannoma.

The surgical treatment plan for resection of the mandible was arrived at after considering the anatomic location of the lesion, wherein odontogenic cysts or tumors are associated with a high rate of recurrence and are typically aggressive in nature. Some of the common differentially diagnosed examples include ameloblastoma, odontogenic keratocyst, calcifying odontogenic cyst and glandular odontogenic cyst (Regezi, 2002). Moreover, the suspicion of a tumor with extraneural spread or a tumor of neural origin extending to the PM space (Sun et al., 2011) further warranted the treatment decision of complete resection in order to achieve a better prognosis.

There is no clear surgical protocol available in the literature for management of intramandibular schwannomas. Nevertheless, complete removal has been considered as the best option since schwannomas of the posterior mandible, although overwhelmingly benign with minimal discomfort, are associated with a risk of intracranial spread (Zhang et al., 2009; Mahmood et al., 2013). For smaller lesions, the treatment of choice is conservative surgical enucleation and periodic follow-up examinations. Enucleation should be considered for tumors without extensive mandibular destruction, in order to conserve the mandible. In cases with extensive destruction, segmental resection of the involved mandible has been mandated along with surgical reconstruction to reach satisfactory facial symmetry and oral function. In the reported case, considering the size of the lesion, its extension, a total resection of the tumor mass was performed. Furthermore, owing to the patient's non-preference for advanced reconstructive modalities, a simple reconstruction plate with a prosthetic condylar head was placed to maintain mandibular continuity and function. Long-term post-operative follow up of the

patient revealed reasonable facial symmetry, stable occlusion and no clinical or radiographic signs of recurrence.

4. Conclusion

Intramandibular schwannomas although extremely rare, are reported and deserve to be included as a differential diagnosis among other benign odontogenic and non-odontogenic tumors of the maxillo-mandibular region. It is imperative that the dental and oral surgeons are aware of this lesion and its clinical and radiographic presentations. With confounding clinical presentations, as reported in this case, clinicians must make every extra effort to obtain a proper diagnosis. Furthermore, owing to its rarity, multicenter reporting of intramandibular schwannomas should be advocated to identify their patterns of incidence and necessary surgical management protocols.

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Conflict of interest

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Ethical approval

This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent

Informed consent was obtained from all individual participants included in the study.

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