








Múltiplos odontomas complexos em mandíbula: relato de um caso raro e revisão da literatura

Múltiples odontomas complejos de la mandíbula: reporte de un caso raro y revisión de la literatura

Multiple complex odontomas of the mandible: a rare case report and literature review

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Fecha correspondencia:

Recibido: diciembre de 2021.

Aprobado: febrero de 2022.

Forma de citar:

Rodrigues da Silva W, Gil de Farias-Morais H, de Macêdo-Andrade AC, Sousa e Silva N, Rifausto JL, Maciel-Dantas WR, de Lisboa-Lopes-Costa A. Múltiplos odontomas complexos em mandíbula: relato de um caso raro e revisão da literatura. Rev. CES Odont 2022; 35(1): 60-69. <https://doi.org/10.21615/cesodon.6542>

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Ética de publicaciones

Revisión por pares

Resumo

Odontomas são as lesões odontogênicas mais comuns nos ossos gnáticos mais comuns. Existem dois tipos principais: odontoma complexo que é composto principalmente de calcificação amorfa, e odontoma composto que é caracterizado por estruturas irregulares semelhantes à dentes. Odontomas são geralmente assintomáticos e, na maioria dos casos, envolvem apenas um quadrante da maxila ou mandíbula. O tratamento escolhido é a remoção cirúrgica.

Palavras-chave: odontoma; múltiplo; tumores odontogênicos.

Resumen

Los odontomas son las lesiones odontogénicas más comunes en los huesos gnáticos más comunes. Hay dos tipos principales: odontoma complejo, que se compone principalmente de calcificación amorfa, y odontoma compuesto, que se caracteriza

Gestión por Open Journal
System

DOI: 10.21615/cesodon.6542

ISSNe 2215-9185

ISSN 0120-971X

Publica con nosotros

por estructuras irregulares similares a dientes. Los odontomas suelen ser asintomáticos y, en la mayoría de los casos, afectan solo un cuadrante del maxilar o la mandíbula. El tratamiento elegido es la extirpación quirúrgica.

Palabras clave: odontoma; múltiple; tumores odontogénicos.

Abstract

Odontomas are the most common tumor-like (hamartoma) odontogenic lesions in the gnathic bones. There are two main types: complex odontoma which is composed mainly of amorphous calcification, and compound odontoma which is characterized by irregular tooth-like structures. Odontomas are generally asymptomatic and in most cases involve only one quadrant of the maxilla or mandible. The treatment of choice is surgical removal. The aim of the present study is to report a rare case of multiple complex odontomas affecting the mandible and to review the literature on the clinical and pathological features of these tumors.

Keywords: odontoma; multiple; odontogenic tumors.

Introduction

Odontoma is a common tumor-like (hamartoma) and generally intraosseous odontogenic lesion that can interfere with the eruption of primary and permanent teeth ^(1, 2). There are two types of odontoma: compound and complex. Compound odontoma is formed by multiple irregular tooth-like structures, while complex odontoma is composed of amorphous calcification and dysplastic dentin mixed with enamel ^(1, 3, 4). Both types usually occur as solitary lesions in the gnathic bones ⁽⁵⁾.

The etiology of odontoma is still uncertain. Some authors reported the possibility of an association with local trauma, inflammatory and/or infectious processes, odontoblastic hyperactivity, and genetic alterations ^(2, 6). Clinically, odontomas are characterized by slow growth, are generally asymptomatic, and do not cause cortical bone expansion; thus, these tumors are usually discovered by routine radiography ⁽⁶⁾. The imaging findings vary according to the type of odontoma and degree of mineralization ⁽⁵⁾. Although they can affect any site in the gnathic bones, compound odontomas are more common in the anterior maxilla, while complex odontomas are more prevalent in the posterior region of the mandible ^(1, 4).

Multiple odontomas (MOs) are characterized by numerous odontomas involving one to four quadrants of the gnathic bones ^(4, 5). These odontomas can be associated with some syndrome such as Gardner's syndrome, otodontal syndrome, and cleidocranial dysostosis ^(5, 7-9). In addition, patients with MOs may have some malformation such as esophageal stenosis and

pulmonary and hepatic alterations ⁽⁵⁾. Within this context, the aim of the present study was to describe a case of complex MOs in the mandible, emphasizing the clinical and pathological features of these tumors.

Case report

A 16-year-old girl was sent to an oral-maxillofacial surgery referral service because of a lesion detected by routine radiography. The imaging exam had been performed 30 days earlier and the duration of the lesion was unknown. During anamnesis, the person responsible for the patient reported that she had been exhibiting mental/behavioral agitation since age 7 after she had developed a headache followed by the loss of balance. However, despite seeking specialist services, no diagnosis could be established that would explain her alterations. Extra- and intraoral physical examination revealed no noteworthy alterations (Figure 1A and 1B).



Figure 1. Initial clinical appearance. **A)** Extraoral view. **B)** Intraoral view.

Panoramic radiography revealed two radiolucent lesions, with well-defined margins, with radiopaque foci inside. Each of the lesions located on the ascending ramus of the mandible. In addition, a periapical lesion is also observed on tooth 46 (Figure 2). In view of the clinical and radiographic features, the diagnostic hypotheses were calcifying epithelial odontogenic tumor (lesion in the right mandibular region) and odontoma (lesion in the left mandibular region). An incisional biopsy was thus performed, which revealed a developing complex odontoma at both sites.



Figure 2. Panoramic radiography. Right image: unilocular, radiolucent lesion with well-defined borders. Radiopaque foci inside the lesion. The lesion caused narrowing of the mandibular canal on the right. Left image: unilocular, radiolucent lesion with well-defined borders. Radiopaque foci inside the lesion.

The treatment of choice was enucleation ([Figure 3](#)), followed by curettage of the bone defect. The removed material was sent for anatomopathological analysis ([Figure 4](#)). Histopathological examination of the hematoxylin/eosin-stained specimen revealed the presence of tooth-like structures formed by a matrix of enamel and dentin, as well as regions containing dental papillae. These findings confirmed the diagnosis of complex odontoma ([Figure 5](#)). The patient was also referred for endodontic treatment of tooth 46. She has been under follow-up for 6 months and there were no signs or symptoms of recurrence of lesions in the mandibular ramus, however, she refused to perform the treatment of the periapical lesion in 46 and new lesions appeared carious in elements 15 and 12 ([Figure 6](#)).



Figure 3. Intraoperative image.

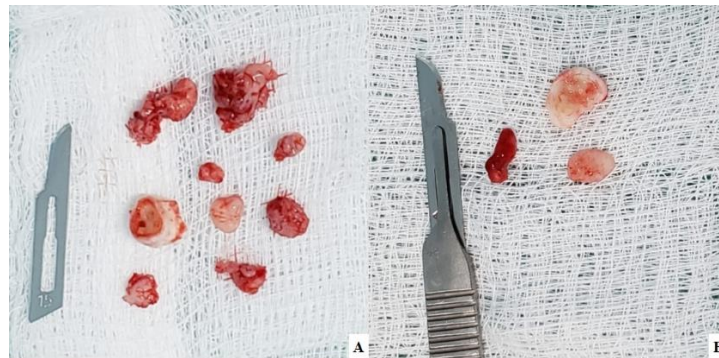


Figure 4. Surgical specimens. **A)** Surgical specimen of the right lesion. **B)** Surgical specimen of the left lesion.

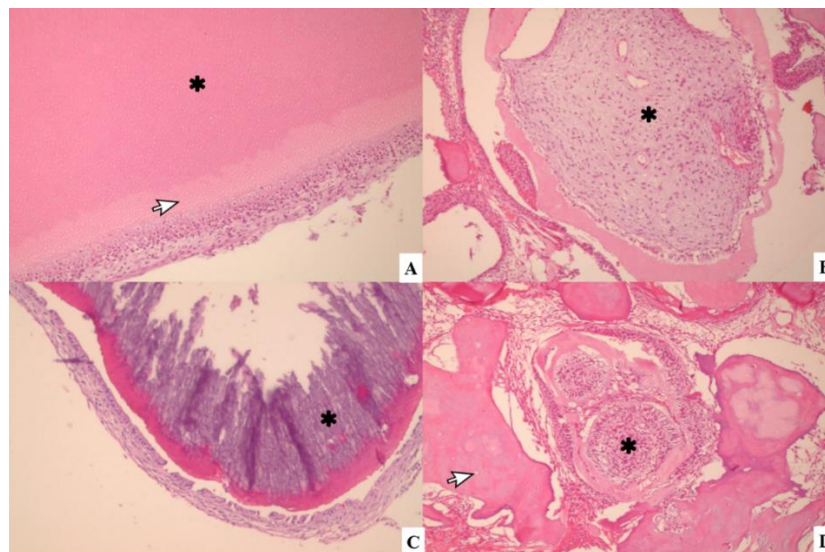


Figure 5. Histopathological features (hematoxylin-eosin). **A)** Different degrees of mineralization of dentin. Pre-dentin (arrow) and mature dentin (asterisk) (200x). **B)** Dental papilla (asterisk) (200x). **C)** Enamel matrix (asterisk) (200x). **D)** Cementum-like area containing cementocyte lacunae (arrow) and islands of ameloblastomatous epithelium. Note the inverted polarization at the periphery (asterisk) (200x).



Figure 6. Control panoramic radiograph showing the absence of recurrence.

Discussion and conclusions

In recent years, several epidemiological studies have investigated the prevalence of odontogenic tumors; among these tumors, odontomas occupy the top of the prevalence pyramid^(10,11). Despite the large number of odontoma cases, MOs are extremely rare in humans and their prevalence is still unknown⁽⁷⁾. In a review of the English literature, we found only 15 cases of MOs located at different sites and not associated with any syndrome (Table 1). Multiple odontomas are usually diagnosed in the first three decades of life. There are only two cases reported in a 53-year-old woman and in a 58-year-old man^(4, 12). Thus, this rare case is the 16th case of MOs described in the literature.

Table 1. Multiple odontomas reported in the literature.

Author	Gender/Age (y)	Location	Type	Treatment
Straith (1950)	M/21	Anterior maxila	Not defined	Excision
Schreiber (1963)	M/9	Anterior maxila	Not defined	Excision
Bader (1967)	F/17 days	AP maxila and AP mandible	Compound and complex	Excision
Malik (1974)	F/7	AP maxila and AP mandible	Compound	ND
Iwamoto (1999)	F/15	Posterior mandible	Not defined	Excision
Ajike (2000)	F/15	AP maxila and AP mandible	Compound	Excision
Bordini (2008)	M/17	AP maxila and AP mandible	Compound	Excision
Jeiaraj (2008)	M/58	Anterior mandible	Complex	Excision
Wanjari (2011)	F/4	AP maxila and AP mandible	Compound and complex	Excision
Erdogan (2014)	M/27	AP maxila and AP mandible	Compound	Partial excision
Guledgud (2014)	M/13	AP maxila and anterior mandible	Complex	Excision
Miloglu (2014)	F/30	Posterior mandible	Complex	Excision
Sun (2015)	M/14	AP maxila and AP mandible	Complex	Partial excision
Gujjar (2015)	M/25	AP maxila and AP mandible	Compound	Excision
Nammalwar (2018)	M/12	Posterior mandible	Complex	Excision
Botelho (2019)	F/53	Posterior mandible	Complex	Excision
Present case	F/16	Posterior mandible	Complex	Excision

M, male; F, female; AP, anterior and posterior; ND, not described.

The exact etiology of odontomas is uncertain, although some authors suggested environmental (local traumas, infection, or inflammation) and genetic factors (cleidocranial dysostosis, Gardner's syndrome, Hermann's syndrome, and Pierre-Robin syndrome) as probable causes⁽¹³⁻¹⁵⁾. For MOs,⁽¹⁶⁾ suggested a possible genetic etiology since partial duplication of chromosome 11 q13.3 may confer a gain of function of the FGF3 and FGF4 genes. However, in the present case, there was no family history of syndromes and the patient was the first known case of MOs in the family. In addition, although some systemic symptoms were identified during anamnesis, the patient has always been followed up by a specialist team and no associated syndrome has been found so far.

Odontomas are commonly associated with the permanent dentition and are asymptomatic lesions discovered incidentally during routine radiography ^(4, 7), as demonstrated in the present case. However, when symptomatic, pain, tooth devitalization and root and/or bone resorption resulting from the pressure exerted by their growth may be observed. Odontomas are generally small but may exhibit sufficient growth to cause bone expansion ⁽¹⁷⁾. Radiographically, the lesions of the present case were large at diagnosis but no bone expansion was observed clinically.

Most cases of odontomas occur between the second and fourth decades of life ⁽⁴⁾ at intraosseous locations ⁽⁷⁾. There is no apparent site predilection but the maxilla is more often involved than the mandible ⁽¹⁸⁾. Compound odontomas are generally found in the anterior maxilla, while most odontomas located in posterior areas, especially of the mandible, are complex odontomas ⁽¹⁹⁾. The case reported here is consistent with these findings.

The radiographic features of odontoma are associated with the stages of development and mineralization. In the early stage, there is no calcification of dental tissues and radiolucent findings predominate; however, completely radiolucent odontomas without calcifications are rare. Partial calcification within a cystic radiolucency is frequently observed, indicating a developing lesion as in the present case in which a more extensive radiolucent ring was observed. In the final stage, calcifications occupy most part of the tumor and are surrounded by a narrow radiolucent ring ^(4, 5, 17).

It is important to note that odontomas in an intermediate stage of development, as in the present case, were previously classified as ameloblastic fibro-odontoma by the World Health Organization; however, with the classification update in 2017, this lesion was removed and is now classified as a more immature stage of odontoma development ⁽¹⁾.

Multiple odontomas must be differentiated from bone lesions and odontogenic tumors with hard tissue formation ⁽¹⁷⁾. The identification of tooth enamel is important because it is not present in bone-related lesions such as fibrous dysplasia and ossifying fibroma. The possibility of involvement of an impacted tooth and displacement to the peripheral border of the tumor also indicate an odontogenic origin. Malignant tumors such as osteosarcoma can be excluded because of the long history, limited and slow growth, and well-delimited borders. Odontogenic tumors with hard tissue formation such as adenomatoid odontogenic tumor and calcifying epithelial odontogenic tumor may not be easily distinguishable from odontomas ^(5, 19). In these cases, histopathological evaluation is necessary to consolidate the diagnosis. In view of these findings, the diagnostic hypotheses of the present case agree with the literature.

Histopathologically, mature complex odontomas are composed mainly of tubular dentin that surrounds areas of the enamel matrix; reduced enamel epithelium, with occasional scattered phantom cells, may surround cracks or hollow circular structures corresponding to mature

enamel that was removed during demineralization. A thin layer of cementum is frequently found at the periphery of the mass. The soft tissue capsule, if present, generally consists of immature connective tissue with cords or islands of ameloblastic epithelium^(17, 20). In the present case, the microscopic features are characteristic of a developing odontoma, including tissue resembling a developing tooth germ and scarce mineralized material.

The present patient was treated with enucleation followed by curettage of the bone defect. Surgical management of odontoma should be conservative and enucleation of the tumor is the treatment of choice. For extensive tumors, stepwise removal may be considered in order to preserve mandibular function and to reduce the risk of pathological fracture⁽⁵⁾. According to the literature, once enucleated, recurrences are not expected⁽¹⁹⁾.

Despite their high incidence, benign nature and indolent behavior, bilateral odontomas are uncommon in the posterior mandible, as described here. In view of the rarity of MOs, this case report contributes to the worldwide literature by describing the 16th diagnosed case of this condition. The findings will help to establish the epidemiological profile and clinical-radiographic and histopathological features of multiple odontomas, as well as to discuss possible differential diagnoses.

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