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HEAD AND NECK

Clinical pathological analysis of nine cases of aneurysmal bone cyst of the jaws in a Brazilian population

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Abstract Aneurysmal bone cysts (ABCs) are benign osteolytic lesions that occur rarely in the jaws. The aim of this study was to investigate the clinical, radiographical and pathological features of ABCs of the jaws. A retrospective analysis of the content of a 39-year database, including nine cases of ABCs of the jaws diagnosed from the archives of the Oral Pathology Service. Nine patients (3 males and 6 females), ranging in age from 5 to 33 years were included. Seven (7/9) lesions were located in the mandible and two (2/9) in the maxilla. A painful swelling was the most common clinical finding ($n = 4$, 4/9). Radiologically, the lesions frequently presented as multilocular (5/9), well defined (4/9), bone expansion and perforation (2/9). Pathological analysis revealed that two cases were associated with central ossifying fibroma and one case with central giant cell lesion. Histomorphology showed a predominance of the solid type (5/9) and of sinusoidal pseudocystic spaces (4/9). Giant cells, osteoid material, calcified material, blood vessels and hemosiderin deposits were observed in 6/9, 7/9, 8/9, 9/9 and 7/9, respectively. The patients with ABCs presented clinical and radiographical features, which often posed a diagnostic dilemma. Knowledge about the most common characteristics of ABCs may contribute to the establishment of a more accurate diagnosis.

Keywords Bone cyst · Aneurysmal · Jaws

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Introduction

Aneurysmal bone cysts (ABCs) are relatively rare osteolytic lesions of uncertain etiology [1, 2] which are characterized by the presence of pseudocystic spaces filled with red blood cells and separated by fibrous connective tissue septa [3]. Most cases of ABC involve the femur and tibia, whereas 12% affect the head and neck region [1]. ABCs rarely occur in the jaws, with about 120 cases being reported so far. In the jaw bones, ABCs are more frequent in the mandible, accounting for two-thirds of cases [4].

The theories regarding the etiopathogenesis of ABC range from a post-traumatic reaction and reactive vascular malformation to a genetic predisposition to bone tumors. One of the most accepted theories is that local circulatory abnormalities result in an increase of venous pressure, with consequent dilatation of the local vascular network [5].

Radiographically, ABCs generally present as radiolucent, unilocular or multilocular lesions with well-defined or poorly defined margins, sometimes causing bone expansion and destruction and/or cortical bone perforation accompanied by a periosteal reaction [4, 6] ABCs can be present alone or associated with other conditions, such as fibrous dysplasia, central ossifying fibroma, cemento-ossifying fibroma, fibromyxoma, central giant cell lesion, osteoblastoma, chondroblastoma, and osteosarcoma [1, 2, 7]. The clinical and histopathological diagnoses are more difficult in these cases [2].

The clinical course of ABCs of the jaws ranges from slow-growing lesions that resolve spontaneously and are discovered by routine radiographical examination to fast-growing lesions that can cause cortical bone rupture [4]. ABCs sometimes show a more aggressive behavior, a fact requiring the differential diagnosis with other neoplasms, including malignant ones [2]. Therefore, knowledge about

the most common characteristics of this disease may contribute to the establishment of a more precise diagnosis and, consequently, more adequate treatment of the patients.

The objective of the present study was to analyze retrospectively the clinical, radiographical and histopathological features of nine cases of ABC of the jaws diagnosed over a period of 39 years at a referral service of the State of Rio Grande do Norte, Brazil. In addition, the data were compared with the few reports published in the literature.

Materials and methods

Case records of patients with ABC diagnosed between 1970 and 2009 were retrieved from the archives of the Oral Pathology Service. Data regarding patient age, gender, anatomical location, duration, history of cortical bone perforation, and presence or absence of swelling, painful symptoms and paresthesia were compiled from the clinical charts sent together with the biopsy records. Race was not considered in this study because the Brazilian population consists of an extensive admixture of Amerindians, Europeans, and Africans [8].

The radiographical features analyzed included a unilocular or multilocular aspect, well-defined or poorly defined margins, and the presence or absence of cortical bone expansion and pathological fracture. For histopathological analysis, the following morphological features of the lesion were analyzed descriptively by three independent and previously calibrated examiners in order to subtype ABC into the vascular, solid and vascular/solid (mixed) type: presence of cavernous or sinusoidal pseudocystic spaces and fibrous connective tissue septa. In addition, the presence or absence of multinucleated giant cells, osteoid material, calcified material, blood vessels, and hemosiderin deposits was evaluated. The data were tabulated and analyzed by descriptive statistics using the Statistical Package for the Social Sciences, version 17.0 (SPSS, Inc., Chicago, IL, USA).

Results

Among the 10,311 oral biopsies retrieved from the archives of the Oral Pathology Service comprising a period of 39 years, nine (0.087%) presented criteria for the diagnosis of ABC. Of these, six (6/9) were observed in women and three (3/9) in men (Table 1). The mean age at diagnosis was 14.3 years (range 5.0–33 years).

Information about the anatomical site was available in all cases (100%). The most common site affected by ABC was the posterior mandible (5/9), followed by the anterior mandible (2/9), anterior maxilla (1/9), and posterior maxilla (1/9) (Table 1). The duration of the lesion ranged from 2 to 60 months.

With respect to clinical manifestations, swelling was observed in four (4/9) cases and six (6/9) patients presented no painful symptoms. Data regarding paresthesia were not available in seven (7/9) cases and two (2/9) patients did not develop this condition (Table 1).

Radiographically, multilocular lesions were observed in five (5/9) cases (Fig. 1), unilocular lesions in two (2/9), and no information was available in two (2/9). Four (4/9) lesions presented well-defined margins, two (2/9) presented poorly defined margins, and no information was available in the remaining cases. Expansion and perforation of cortical bone were observed in only two lesions (2/9). Information regarding pathological fracture was not available in seven (7/9) of the cases and this symptom was absent in the other two (Table 1). We believe that these seven cases,

Table 1 Clinicoradiographic features of 9 cases of aneurysmal bone cysts of jaws

Pt No.	Age (year)	Gender	Duration (months)	Location	Swelling	Painful	Paresthesia	Locular	Border	Expansion	Perforation	Fracture
1	13	F	35	Max p	NA	No	NA	Multilocular	WD	NA	NA	NA
2	13	F	5	Mand p	Yes	NA	NA	Unilocular	WD	Yes	Yes	NA
3	33	F	60	Mand p	Yes	Yes	NA	Multilocular	ID	NA	Yes	NA
4	17	M	6	Mand a	Yes	No	NA	Multilocular	NA	NA	NA	NA
5	14	F	NA	Max a	Yes	Yes	NA	Unilocular	WD	NA	NA	NA
6	5	F	NA	Mand p	NA	No	NA	Multilocular	ID	NA	NA	NA
7	11	M	24	Mand p	NA	No	NA	NA	Na	NA	NA	NA
8	12	F	NA	Mand a	No	No	No	Multilocular	WD	Yes	No	No
9	11	M	24	Mand p	No	No	No	NA	NA	No	No	No

Pt No. Patient number, *NA* not available, *Max p* posterior maxillary, *Max a* anterior maxillary, *Mand p* posterior mandibular, *Mand a* anterior mandibular, *WD* well defined, *ID* ill-defined

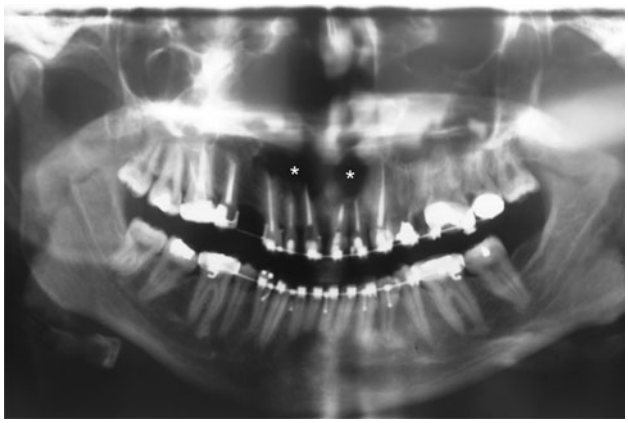

Fig. 1 Radiographically, ABC presents as a radiolucent image and with well-defined margins. Lesion located *above* the apices of the maxillary anterior teeth from the region of 13–22. There are external root resorption at the apex of 11, 12, 21 and 22

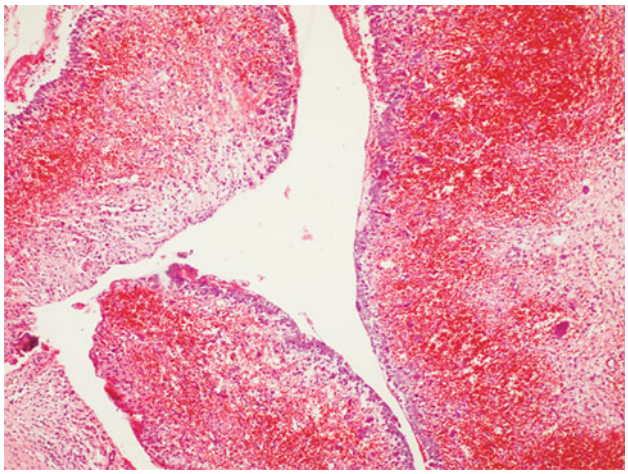

Fig. 2 Histologic section showing solid morphology with multinucleated giant cells, hemorrhagic foci and scarce blood-filled sinusoidal spaces (200X)

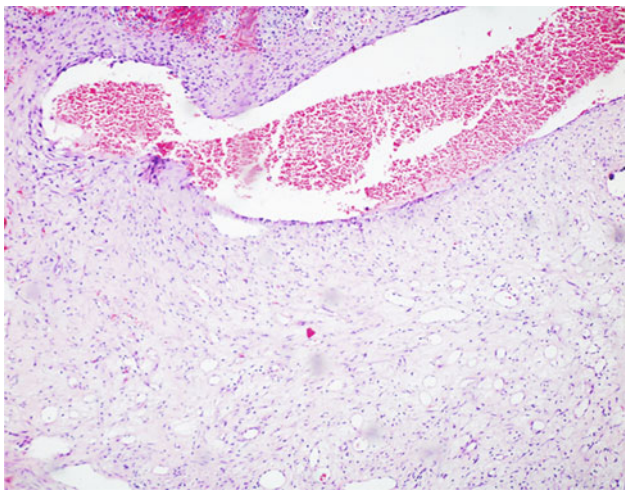

Fig. 3 Histologic section showing mixed morphology with pseudocystic space filled with red blood cells and abundant fibrous connective tissue (200X)

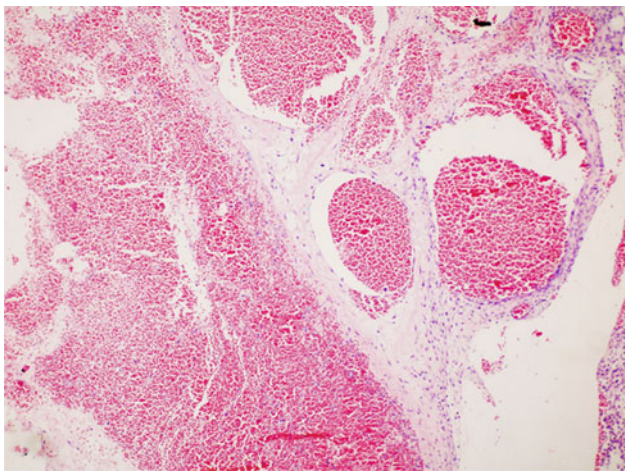

Fig. 4 Histologic section showing blood-filled spaces of variable size separated by fibrous connective tissue septa (100X)

probably, did not exhibit pathological fracture, since this finding would be a relevant signal and should be informed by the clinician.

With respect to histological subtype, five (5/9) lesions were of the solid type (Fig. 2), three (3/9) of the mixed type (Fig. 3), and one (1/9) of the vascular type (Fig. 4). Sinusoidal pseudocystic spaces were observed in four (4/9) cases, cavernous spaces in three (3/9), and cavernous and sinusoidal spaces in two (2/9). Giant cells, osteoid material, calcified material, blood vessels and hemosiderin deposits were observed in 6/9, 7/9, 8/9, 9/9 and 7/9 of the cases of ABC, respectively (Table 1). Three (3/9) cases of ABC were associated with other bone diseases, including central ossifying fibroma in two (Figs. 5, 6) and central giant cell lesion in one (Figs. 7, 8).

Discussion

The World Health Organization (WHO) defines ABC as an expansile osteolytic lesion consisting of blood-filled spaces separated by connective tissue septa that contain osteoid material and multinucleated giant cells [9]. ABC was mentioned for the first time in 1942 by Jaffé and Lichtenstein [10], who recognized and described this lesion as a distinct clinical–pathological entity [6].

The etiopathogenesis of ABC remains controversial. It is believed that the disease results from an intraosseous vascular malformation and some hypotheses have been suggested: trauma resulting in subperiosteal or intramedullary hemorrhage that is incompletely repaired [4]; an increase in venous pressure and vascular engorgement originating from hemodynamic disturbances that lead to bone

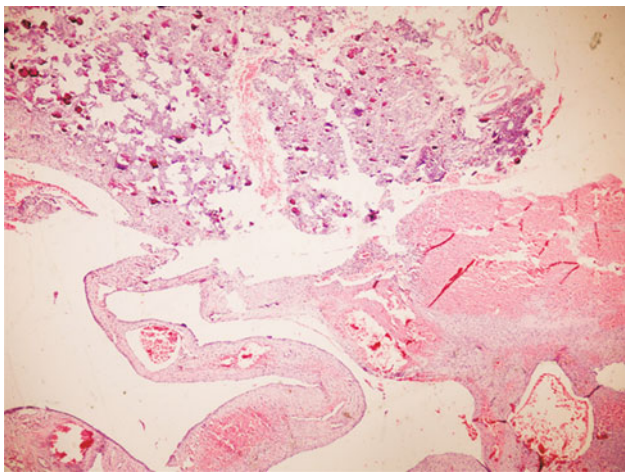

Fig. 5 Histologic section showing ABC associated with central ossifying fibroma (100X)

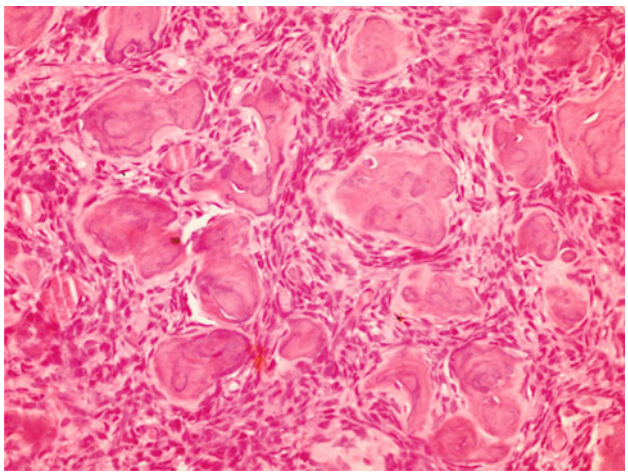

Fig. 6 Histologic section highlighting the area of central ossifying fibroma (400X)

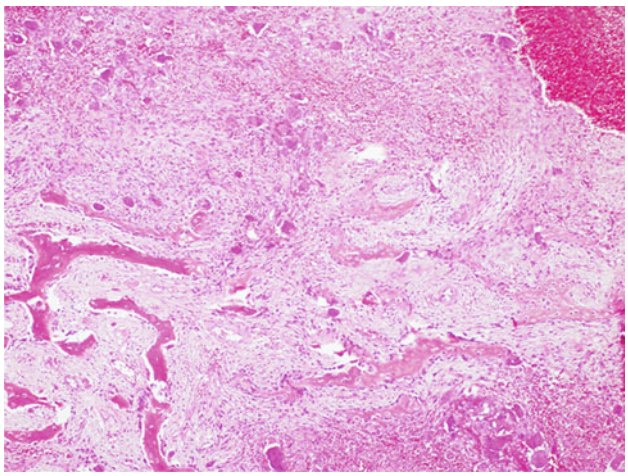

Fig. 7 Histologic section showing ABC associated with central giant cell lesion. Notice blood-filled space and hemorrhage. Dense proliferation of the mesenchymal cells and multinucleated giant cells, presence of bone in its young form are the most significant component (100X)

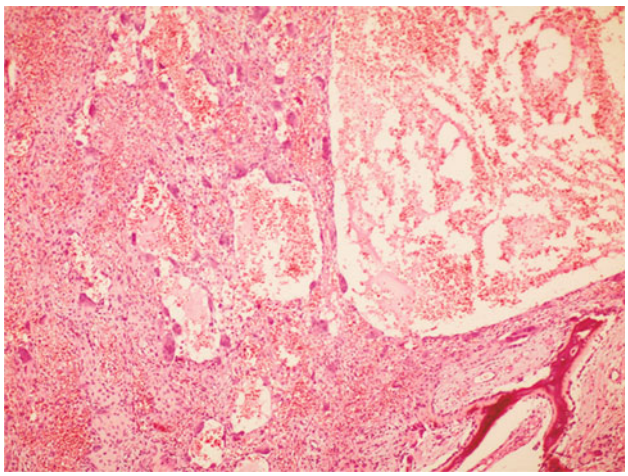

Fig. 8 Histologic section showing ABC associated with central giant cell lesion. Notice blood-filled spaces of variable size, hemorrhage, multinucleated giant cells and bone trabeculae (200X)

resorption and erosion [5, 11]; secondary phenomenon due to the presence of a preexisting lesion [12]. There is general consensus that ABC is a reactive condition of uncertain and controversial etiology [1] since it can be associated with a variety of benign and malignant bone tumors [7]. Some studies have demonstrated cytogenetic abnormalities [13, 14], a fact raising the hypothesis of a neoplastic nature of these lesions. Therefore, the reactive nature of ABC has been questioned in studies demonstrating that ABC of the extragnathic bones is associated with a chromosome translation t(16; 17)(q22, p13) involving the USP6 (an oncogene) and CDH11 (promoter) genes [13, 15].

ABCs commonly affect the metaphyseal region of long bones (especially the femur and tibia), followed by the spine [16]. Approximately 2–12% of cases involve the head and neck region [6, 16]. In the present study, only nine (0.087%) of the 10,311 specimens diagnosed in the head and neck region over a period of 39 years were ABCs of the jaws. Mohammad et al. [4] identified about 120 cases of ABC involving the maxillofacial region published until 2008, most of them individual case reports [3, 17]. Although case series are less common [2, 4, 6], they are highly relevant since the data refer to a larger number of lesions, a fact permitting to trace a more reliable clinical–pathological profile.

ABCs of the jaws can occur in patients of all ages, but most studies show a predominance in patients younger than 20 years (mean of 14 years) [2, 4, 6, 16]. In the present study, the age distribution agreed with previous studies, with young patients with a mean age of 14.3 years being mainly affected. According to Pelo et al. [11] and Sun et al.

[2], there is no gender preference, but several investigators reported a higher frequency among females [3, 4, 18, 19] in agreement with the present results.

In the head and neck region, the mandible is the site most affected by ABC, with an incidence rate of 3:1 [1, 2]. Bernier and Bhaskar [20] described the first case of ABC in the mandible. In the present study, 55.5% of the cases occurred in the posterior mandible. According to the literature, this anatomical site is affected in more than 90% of cases, with the mandibular body and ramus being most frequently involved [1, 2]. Cases involving the coronoid process and mandibular condyle are rare [4, 11, 16, 19, 21]. According to Tang et al. [17], approximately 22 cases of maxillary ABC have been reported. Although rare, in the present series two (22.2%) of the nine cases of ABC occurred in the maxilla.

ABC does not present pathognomonic clinical features [11] and its clinical course ranges from asymptomatic, slow-growing lesions that resolve spontaneously and are discovered by routine radiographical examination to fast-growing lesions that can cause expansion and destruction of cortical bone. The latter are aggressive and more frequent [4]. Although signs and symptoms depend on the anatomical site affected, pain and swelling are the main symptoms reported [2, 3, 12, 16]. In the present study, swelling was the most prevalent sign (44.4%), whereas painful symptoms were observed in only two (22.2%) cases. Facial asymmetry and cortical bone perforation are identified in the case of more extensive lesions [12, 17] and were detected in the present study in only two cases (22.2%). No data regarding the presence or absence of cortical bone expansion and perforation were available in five clinical charts.

Radiographically, the cases of ABC analyzed here exhibited a radiolucent aspect and most of them showed a multilocular image with well-defined margins, in agreement with other investigators [19]. In fact, ABCs are usually radiolucent, unilocular or multilocular lesions with a soap bubble-like or honeycomb appearance and well-defined or poorly defined margins [12]. Like the clinical features, the radiographic appearance is also not specific due to the similarity with other lesions, such as central giant cell lesion, brown tumor of hyperparathyroidism, traumatic bone cyst, odontogenic myxoma, keratocyst, ameloblastoma, and osteosarcoma [1, 11, 19]. Thus, a diagnosis of exclusion is essential and aspiration of blood from the lesion may lead the clinician to suspect a vascular lesion or ABC [19].

Histologically, ABC is characterized as an osteolytic and pseudocystic lesion due to the absence of an epithelial lining, which contains cavernous or sinusoidal blood-filled spaces of variable size. These spaces are separated by fibrous connective tissue septa that may contain osteoclast-like multinucleated giant cells, osteoid material, calcified tissue and even hemosiderin deposits [11, 19]. All of these histopathological findings were frequent in the specimens analyzed in this study.

The prominence of pseudocystic spaces and fibrous septa in ABC is variable. On the basis of this variability, some investigators have classified ABC into three subtypes: vascular (conventional), solid, and vascular/solid (mixed) [1, 4, 11]. In the present study, five (55.5%) cases were classified as the solid type, three (33.3%) as the mixed type, and one (11.1%) as the vascular type. This finding disagrees with the literature reporting an incidence of the solid type of 5–8% [22], with the vascular variant being the most common. The solid subtype is characterized by abundant fibrous connective tissue and the presence of hemorrhagic foci and scarce blood-filled sinusoidal spaces [3]. An expansile behavior without bone destruction, as well as discrete bleeding during surgical access of the lesion, is frequently observed in this variant [4]. Expansion without bone destruction was a common feature in the present series. Clinically, vascular ABC is associated with a more aggressive biological behavior, rapid and destructive growth, cortical bone perforation with invasion of soft tissues, and abundant bleeding during surgery [4, 12]. This fact might explain the less aggressive behavior exhibited by the lesions of this series since the vascular variant corresponded to the minority of cases.

In view of the presence of osteoclast-like multinucleated giant cells, the distinction between the solid variant of ABC and central giant cell lesion or central ossifying fibroma might be difficult because of the presence of osteoid and calcified material [1]. According to Roychoudhury et al. [19], the mere presence of giant cells cannot be used as a criterion to differentiate ABC from central giant cell lesion. As a distinct feature, Vergel De Dios et al. [22] reported that the stroma is more fibrogenic in ABC than in central giant cell lesion. In addition, the presence of large vascular spaces points to the diagnosis of ABC.

Some theories evidenced that some ABCs are a variant of a giant cell lesion. The term “giant cell lesion” has been used as a synonym in the pathology literature to describe this variant, mainly about the solid variant of ABC [23]. However, the ABC is considered to be as a unique entity by WHO. Contrary to what happens in giant cell tumour, ABC usually does not extend to the end of the bone, and, moreover, ABC occurs in the first two decades, while giant cell lesion is a lesion of the mature skeleton (3).

ABC can be classified as primary or secondary depending on the presence or absence of other preexisting entities. The term secondary is used in the case of association with other lesions such as fibrous dysplasia, central ossifying fibroma, cemento-ossifying fibroma, fibromyxoma, central giant cell lesion, osteoblastoma, chondroblastoma, and

osteosarcoma [1, 2, 7]. In the present study, two cases were associated with central ossifying fibroma and one case was associated with central giant cell lesion. The findings of the present study and other previously reported results [2, 24] demonstrate that central ossifying fibroma is the most common lesion associated with secondary ABC of the jaws.

The treatment of ABC depends on the size and location of the lesion and does not appear to be related to the histological variants, although vascular variety behaves more aggressively [2]. The treatment ranges from simple curettage to broad resection [2, 11]. Only in extended lesions or in case of multiple recurrences is a block resection recommendable [2]. Curettage shows a recurrence rate of 20–70%, whereas recurrence rates range from 11 to 25% for surgically excised lesions [1, 3, 11]. In this study, the treatment provided in all cases was curettage and only one case had recurrence. Block resection was instituted in the recurrent lesion. In the study of Sun et al. [2], the recurrence rate of ABCs removed by curettage was 11.8%. This fact was attributed to the incomplete removal of the lesion, mainly due to technical difficulties encountered in the case of extensive tumors [25]. The choice of a more conservative treatment such as curettage demonstrates the concern of clinicians with function and esthetics since ABCs generally affect young patients.

ABC of the jaws is an enigmatic pseudocyst due to its variable presentations. The diagnosis should be based on a combination of clinical, radiographical and histopathological features. Therefore, the analysis of the characteristics of ABC of the jaws may help understand its development and thus contribute to the establishment of a more accurate diagnosis and the most adequate therapeutic management of patients with this disease.

Conflict of interest We have no conflict of interest to declare.

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