

Original Article

Aggressive mandibular aneurysmal bone cyst misdiagnosed as simple bone cyst: a case report and literature review

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Abstract: In this report, we present a case of an aggressive aneurysmal bone cyst (ABC) in the jaw that presented with a challenging and puzzling diagnosis. A comprehensive diagnosis was made based on the clinical, radiographical, and pathological features. The present case highlights the importance of surgeons' knowledge of and careful attention to maxillofacial giant cell lesions, particularly the rare and aggressive ABCs. Diagnosis can be challenging owing to inadequately detailed biopsy and atypical pathological presentation. Further, because of the rapid growth of the cyst, early diagnosis is important for wide excision, successful bone reconstruction, and good prognosis. As ABC is a rare entity, oral and maxillofacial surgeons, radiologists, and pathologists should be more alert to and aware of this lesion for early and accurate diagnosis and curative treatment.

Keywords: Aggressive, aneurysmal bone cyst, simple bone cyst, central giant cell granuloma

Introduction

An aneurysmal bone cyst (ABC) is a rapidly growing and destructive benign bone tumor mainly seen within the long bones. However, ABCs are extremely rare in the jaw, with an incidence of just 1-2%; further, they represent only about 1.5% of all non-odontogenic and non-epithelial cysts of the jaws [1]. ABCs are significantly more frequently seen in the mandible than in the maxilla, and in more than 90% of patients, they affect the posterior regions of the jaws [2]. While the majority of lesions are clinically similar to odontogenic cystic lesions, with painful or painless swelling being one of the main symptoms [3]. ABCs are more aggressive than common odontogenic cystic lesions and tend to reoccur after inadequate surgery. Reoccurrence rates for ABC are relatively high and exceed 30% [4]. While the majority of ABCs exist as a primary bone lesion, they may also occur secondary to other osseous conditions such as central giant cell lesion, fibrous dysplasia, chondroblastomas, osteoblastomas, and fibromyxomas [1, 2, 5]. This makes the diagnosis of ABCs more complex and challenging.

In this report, we present the case of an aggressive mandibular ABC that was misdiagnosed as a simple bone cyst because of the surgeon's ignorance of the condition and inadequate pathological examination. Consequently, a surgical curettage was performed, which resulted in recurrence shortly thereafter. After a complete resection and thorough pathological examination, we diagnosed the lesion as an ABC based on the clinical, radiographical, and pathological presentation. The present case highlights the importance of surgeons' knowledge of and attention to giant cell lesions in the jaws, especially in case of the rare and aggressive ABCs.

Case report

This case report was approved by the Ethics Committee of Affiliated Stomatology Hospital of Medical School, Nanjing University. Informed consent was obtained from the patient included in the case.

In November 2012, a 10-year-old girl was referred to our hospital with a painless swelling

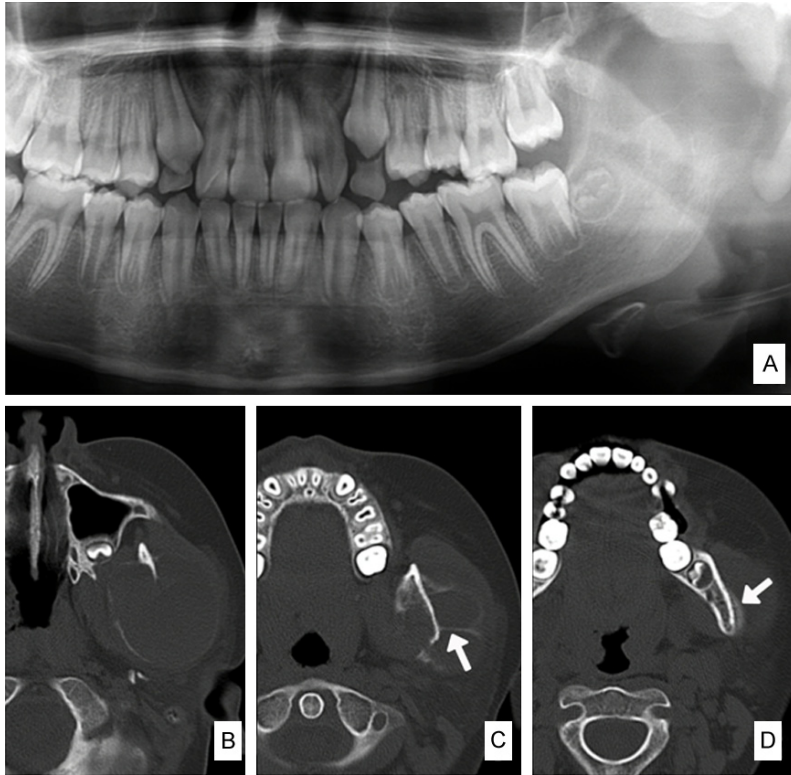


Figure 1. Images depicting the primary lesion. A: A panoramic radiograph showing swelling of the left condyle and coracoid. B: Spiral computed tomography showing a balloon-like cystic lesion with an eggshell-thin cortex. C: Straight and thin septa in the lesion (arrow). D: Periosteal reaction (arrow).

on the left side of her face since 10 days ago. The patient recalled a history of a fall, which occurred several years ago. Extraoral examination revealed an obvious swelling of the left pre-auricular region and considerable reduction of mouth opening. The lesion was painless to palpation, and the submandibular and cervical lymph nodes were not palpable. A panoramic radiograph taken in our hospital revealed an obvious swelling of the left condyle and coracoid (**Figure 1A**). Computed tomography (CT) showed the following characteristics: (1) The lesion was considerably expansible and occupied the majority of the left ramus, entire condyle, and coracoid. (2) The cortex was eggshell thin and discontinuous in some regions. (3) Onion-skin-like bone apposition was observed at the periphery of the cortical bone. (4) The lesion contained straight and thin septa (**Figure 1B-D**). Central giant cell granuloma (CGCG) was considered based on the radiographical features.

Blood routine examination showed unremarkable results. The lesion was biopsied via a sub-

mandibular approach. Abundant bloody fluid was observed intraoperatively. A small amount of the soft tissue attached to the bone was resected for frozen-section examination, and intraoperative pathology revealed a jaw cyst; therefore, surgical curettage was performed. Subsequent routine histopathological diagnosis was a simple bone cyst (SBC) without epithelial lining.

In January 2013, the patient again presented with facial swelling on the left side and was referred to our hospital for treatment. The short interval between the first surgery and recurrence suggested an aggressive lesion, and a surgical exploration was performed under general anesthesia. This time, more tissues were resected for frozen-section examination. Histo-

pathological examination revealed that the lesion consisted of spindle cells and multinucleated giant cells (**Figure 2A**) and showed some “vascular” form (**Figure 2B**). The pathological diagnosis was inclined to a central giant cell granuloma (CGCG) with secondary ABC. However, the patient’s parents wanted to have more certain diagnosis and take the patient to other hospitals for consultation, and the patient did not have further treatment in our hospital.

In June 2013, the patient was again referred to our hospital. No treatments were performed since the patient left our hospital in January 2013. The pre-surgical CT revealed some new characteristics (**Figure 3**): (1) The lesion was extremely expansible with a wave-like pattern of cortical bone. (2) Calcified foci were seen within the lesion and granular bone pattern at the periphery of the expanded bone. (3) Fluid levels (~20 HU) and gas levels (~860 HU) were present. (4) Resorption of the maxillary tuberosity was detected.

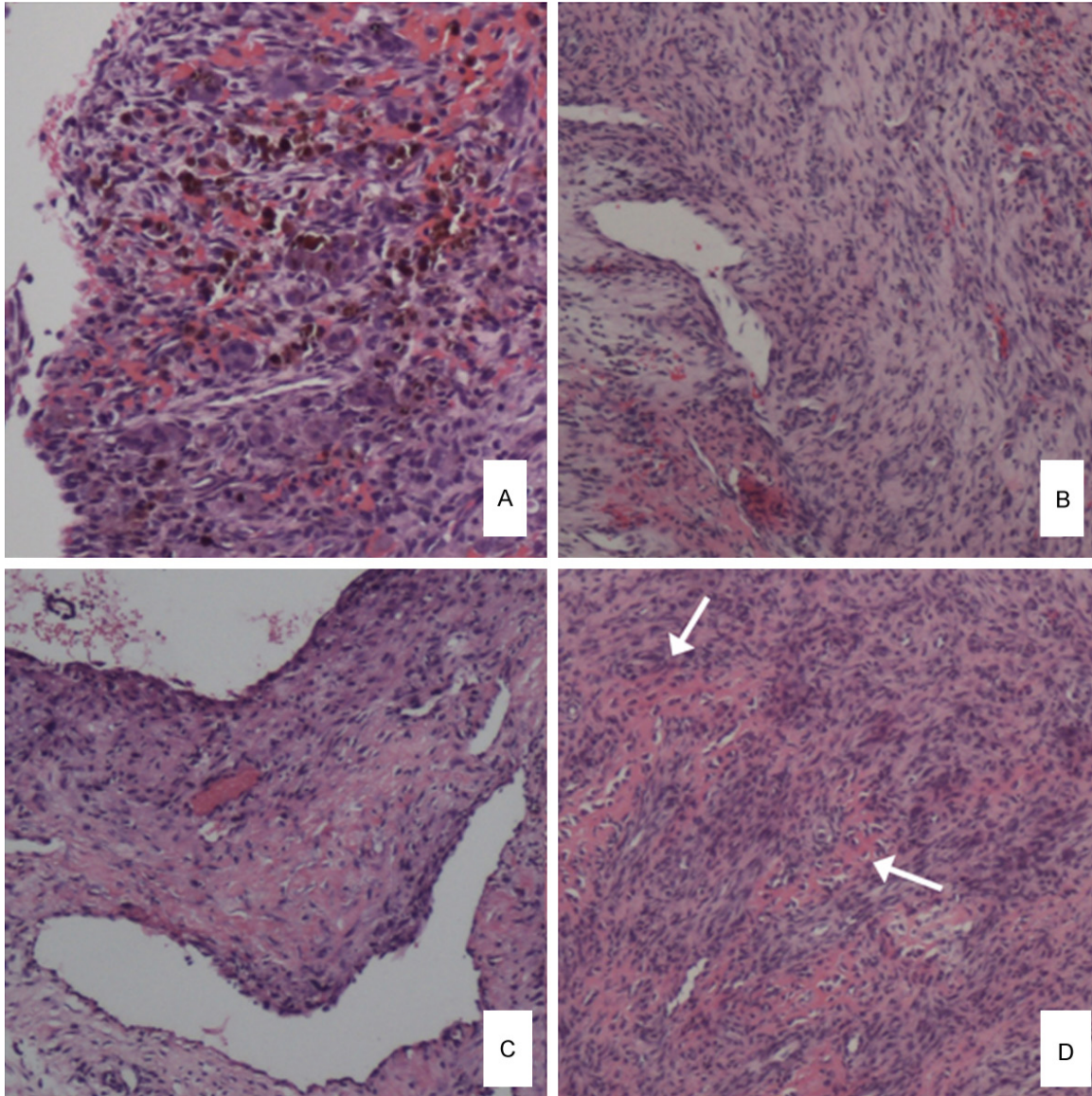


Figure 2. Pathological examination of the recurrent lesion. A and B: Specimen in January 2013 showing spindle cells and multinucleated giant cells and “vascular” form in the lesion. C: Specimen in June 2013 showing “vascular” form and fewer multinucleated giant cells. D: Osteoid formation in the cyst wall (arrows) (A-D: Hematoxylin-eosin stain, original magnification, 200×).

A segmental resection from the left mandibular second premolar to the entire ramus, condyle, and coracoid was performed under general anesthesia. This time, a thorough histopathological examination was performed after complete resection of the lesion. The cross-section of the specimen presented a typical cystic appearance (**Figure 4A**). Smear examination of an aspiration biopsy specimen obtained during the surgery showed numerous blood cells with scattered giant cells (**Figure 4B**). On microscopy, fibroblasts, scattered osteoclast-like giant

cells, and “vascular” forms and osteoid tissue were found in some regions (**Figure 2C** and **2D**). And the specimen was sent to another hospital for a consultant. Finally, a comprehensive diagnosis of ABC was made owing to the clinical, radiographical, and pathological presentations. Immunohistochemistry of the tumor specimen revealed positive staining for CD68 (+) and Ki-67 (+, 20%) which suggested more active cell proliferation than common cystic lesions. The patient has not had another recurrence until July 2015.

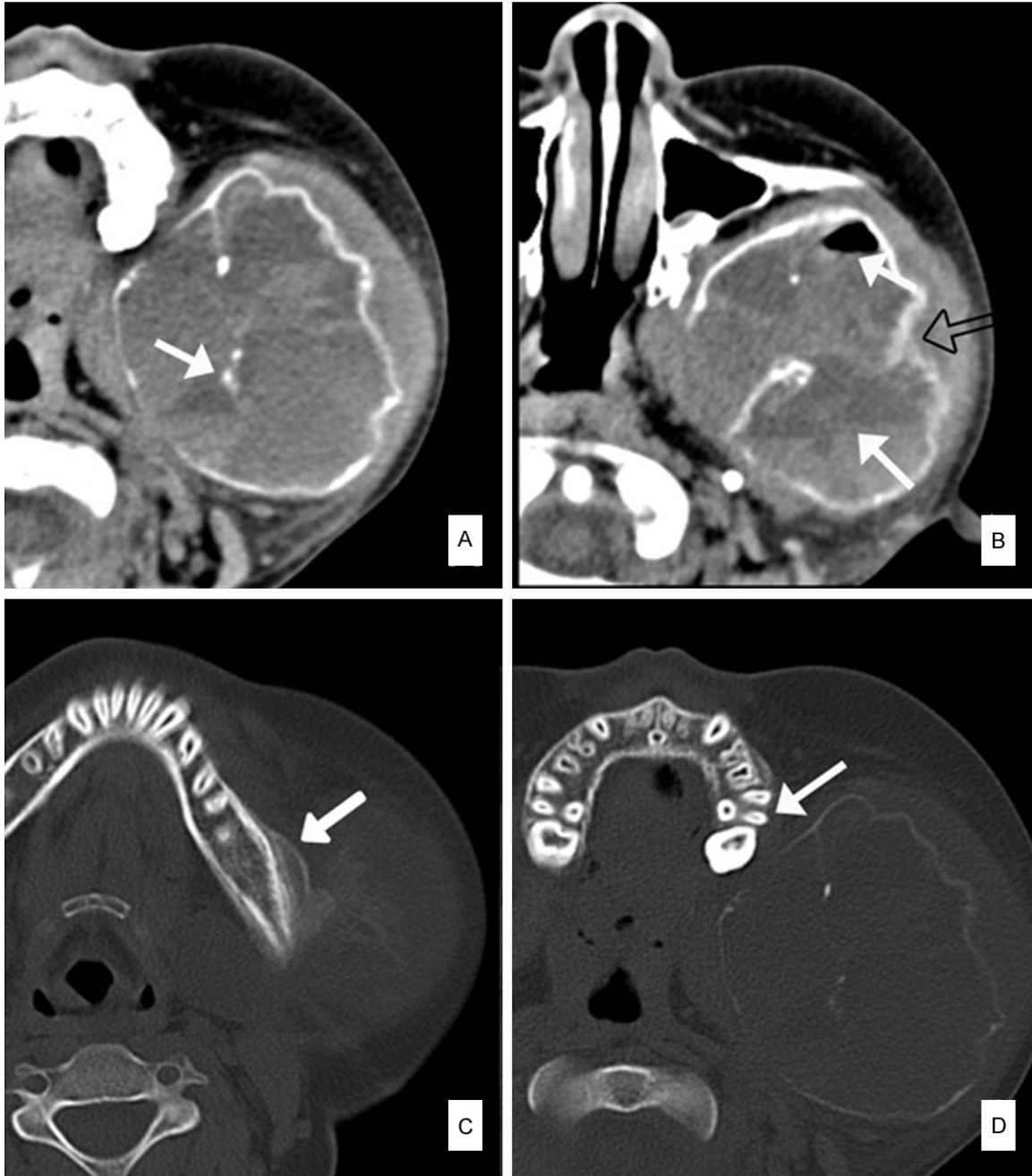


Figure 3. Images depicting of the lesion (June 2013). A: Wave-like pattern of cortical bone and calcified foci (arrow). B: Fluid and gas levels (arrows) and granular bone pattern at the periphery of the expanded bone (black arrow). C: Periosteal reaction (arrow). D: Resorption of the maxillary tuberosity (arrow).

Discussion

The diagnosis of an ABC in the jaw is sometimes challenging and complex, or even not straightforward. During the first surgery, the surgeon overlooked the specific radiographic presentation of the lesion and little tissue was resected for biopsy examination, which likely

led to a misdiagnosis of simple bone cyst at that time. For the recurrence in January 2013, surgical exploration showed abundant multinucleated giant cells and “vascular” form in some regions; therefore, a modified diagnosis of CGCG secondary to ABC was made. In June 2013, smear examination during the surgery showed numerous blood cells. The pathological

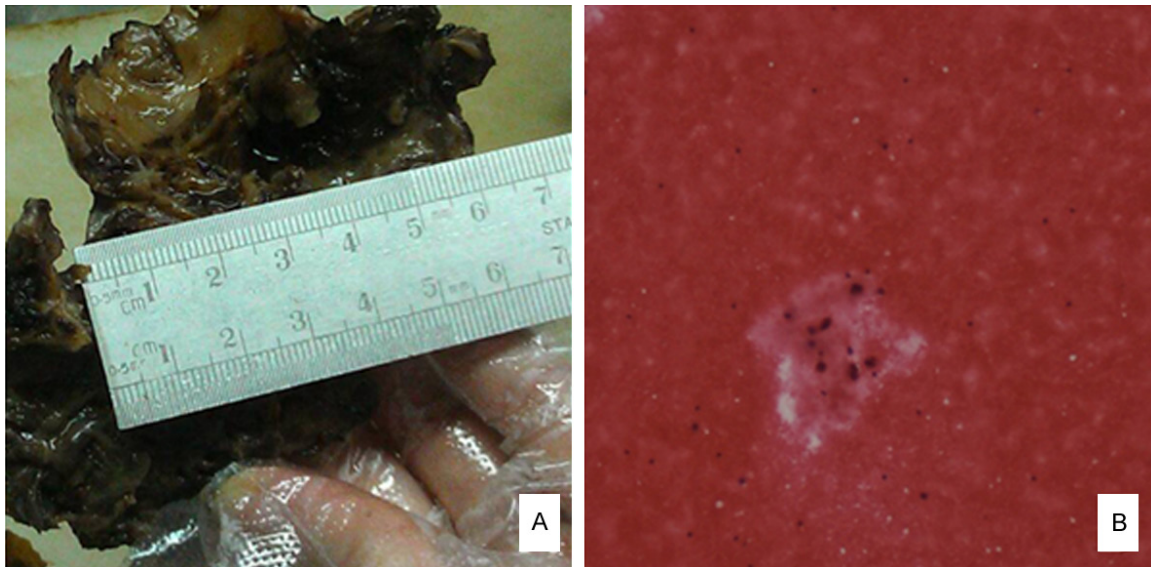


Figure 4. June 2013 A: Surgical specimen showing a cystic appearance. B: Cytology smear showing numerous blood cells with scattered giant cells (Original magnification, 200×).

examination showed fewer multinucleated giant cells and more typical “vascular” form. Moreover, And owing to en-bloc resection performed at this admission, the gross specimen presented a multi-cystic appearance that also supported the diagnosis of ABC. Additionally, rather typical radiological signs of ABC were found on the CT images, which finally helped make a conclusive diagnosis of ABC.

In the oral and maxillofacial region, ABC, SBC, and CGCG are the three kinds of non-odontogenic lesions that present with similar clinical, radiographical, and pathological characteristics. All of these occur more frequently in the mandible and show a predilection for young people (<30 years) [1, 6, 7]. On radiography, all 3 types present as radio-transparent, expansive lesions with thinning of the cortical bone. Pathologically, the 3 types do not have an epithelial lining, and their fibroblastic stroma may contain calcified bone matter and multinucleated giant cell [8, 9]. Furthermore, ABCs may also occur secondary to CGCG [1, 2]. In clinical practice, the differential diagnosis of ABC from SBC is very important for the prognosis is quite different. While SBCs have a good prognosis and usually undergo spontaneous remission, ABC is more aggressive and sometimes requires more radical treatment to prevent further recurrences [8]. For this case, the pre-surgical CT examination of the primary lesion was not consistent with the diagnosis of an SBC, as

SBCs are usually unicameral, well-contoured, with fine bony margins and are not as expansible as the lesion in this case. Our patient showed a balloon-like swelling with eggshell-thin cortex on the first CT examination, which became more obvious in the recurrence. Moreover, the lesion contained fluid and gas levels that are considered characteristic features of ABCs [10, 11]. In addition, resorption of the maxillary tuberosity observed in June 2013 further suggested the aggressive nature of this lesion.

According to the World Health Organization, ABCs are considered a benign intra-osseous lesion, characterized by blood-filled spaces of varying sizes associated with a fibroblastic stroma containing multinucleated giant cells, osteoid, and woven bone [9]. Histologically, the lesion has no similarity to either a cyst or an aneurysm. The classic numerous blood-filled sinuses that make up the typical “vascular” form are a major characteristic for the differential diagnosis of ABCs [1]. In this case, “vascular” form was found in some region in June 2013, although was scattered, and this finally supported the diagnosis of ABC.

The present case highlights the importance of rapid and thorough diagnosis in cases of ABC. Diagnosis can be challenging owing to inadequately detailed biopsy and atypical pathological presentation. Further, because of the rapid

growth of the cyst, early diagnosis is important for wide excision, successful bone reconstruction, and good prognosis. As ABC is a rare entity, oral and maxillofacial surgeons, radiologists, and pathologists should be more alert to and aware of this lesion for early and accurate diagnosis and curative treatment.

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Disclosure of conflict of interest

None.

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