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Case report

Ameloblastic fibrosarcoma following a tooth extraction: a case report and literature review ☆,☆☆

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Introduction

Ameloblastic fibrosarcoma (AFS) is an extremely rare odontogenic sarcoma, in which the epithelial component is benign and mesenchymal component is malignant [1]. About 50% of AFSs are described as malignant transformations of recurring ameloblastic fibroma (AF), while the other 50% are deemed to arise de novo without definite history of preexistent AF [1]. Due to its rarity, AFS still remains a poorly characterized orphan disease with <110 cases reported so far [2].

In this article, we describe a 29-year-old male patient diagnosed with AFS 2 months after a tooth extraction, to provide a better understanding of the rare odontogenic tumor.

Case report

A 29-year-old male presented with occlusal pain in the left lower molar region, and the third mandibular molar was removed at local hospital. Unfortunately, there was no imag-

Abbreviations: AFS, Ameloblastic fibrosarcoma; AF, Ameloblastic fibroma; CT, Computed tomography; H&E, Hematoxylin and eosin.

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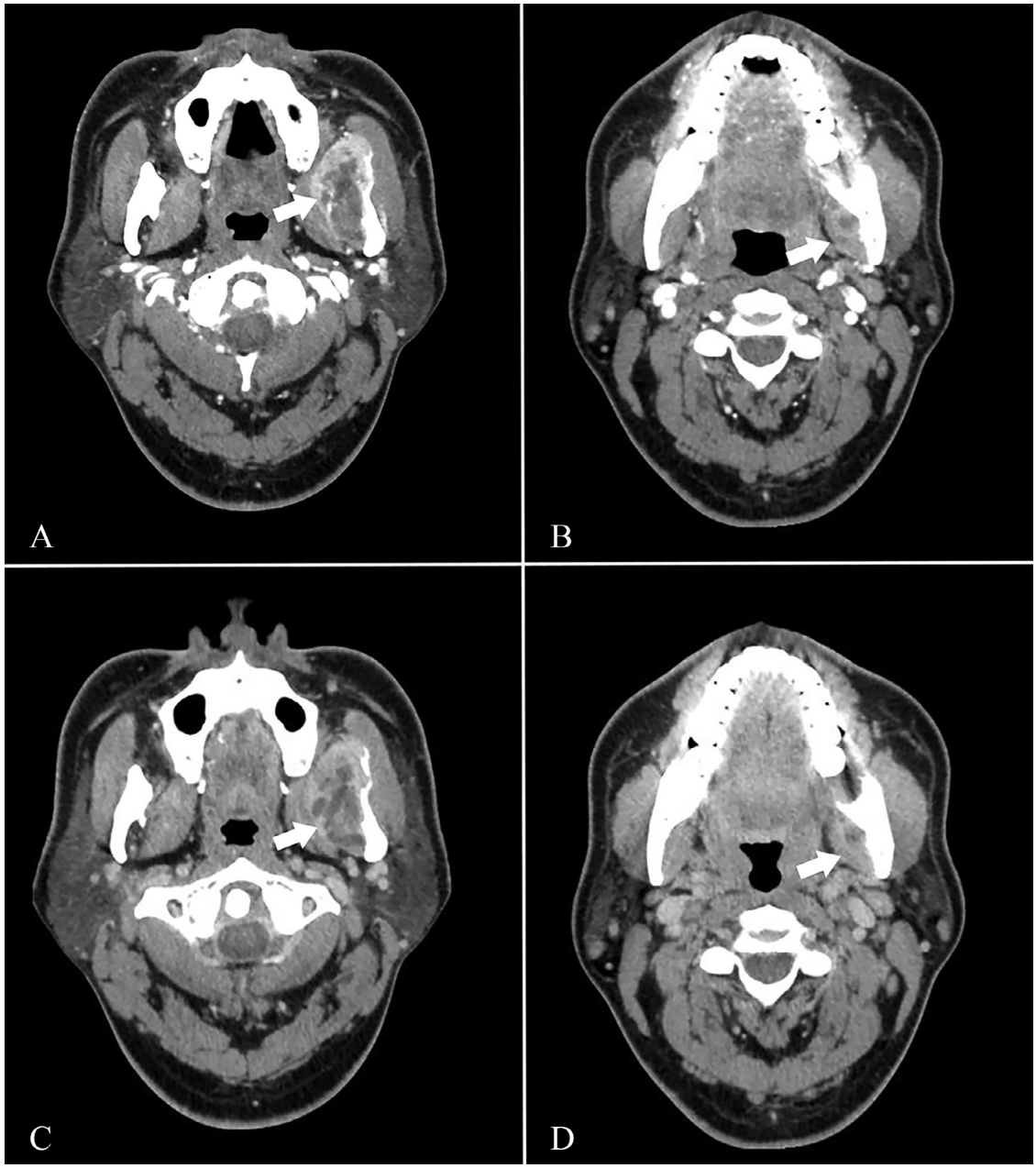

Fig. 1 – Arterial phase (A-B) and venous phase (C-D) CT images, soft-tissue window (window width: 300HU, window level: 35HU). A massive lesion straddled the left mandibular ramus (arrows in A, C) and angle (arrows in B, D). A poorly-defined soft tissue mass with cystic components in it.

ing at the local hospital before extraction. After that, the molar cavity had been remained unhealed for 2 months, with neoplasm formation. Therefore, the patient presented to our hospital for further treatment. Local examination showed lobulated mass in the molar cavity. Contrast-enhanced computed tomography (CT) of the maxillofacial region revealed a mass located in the left mandibular angle and ramus (Fig. 1). It was a poorly-defined soft tissue mass with cystic components in it (Fig. 1). The lesion straddled the left mandibular angle and ramus, extending laterally and medially, causing bone expansion and cortical bone perforation (Figs. 2A-B). Two enlarged lymph nodes were observed in the left IB and II region (Figs. 2C-D).

Fine needle biopsy of the mass was performed, and a diagnosis of AFS was rendered. Therefore, the dental surgeon chose the therapeutic approach of segmental resection of the left mandibular body and cervical lymph node dissection. Simultaneous reconstruction featured the transfer of an iliac-crest free flap fixed to a prebent reconstructive plate was performed. Microscopically, there was a biphasic pattern composed of epithelium and mesenchymal components in the mass. The mesenchymal portion of the tumor showed features of malignancy (marked cellularity, nuclear atypia and occasional mitoses), whereas the epithelial component was rather bland (Fig. 3). The benign epithelial component showed uniform positivity for pan-cytokeratin,

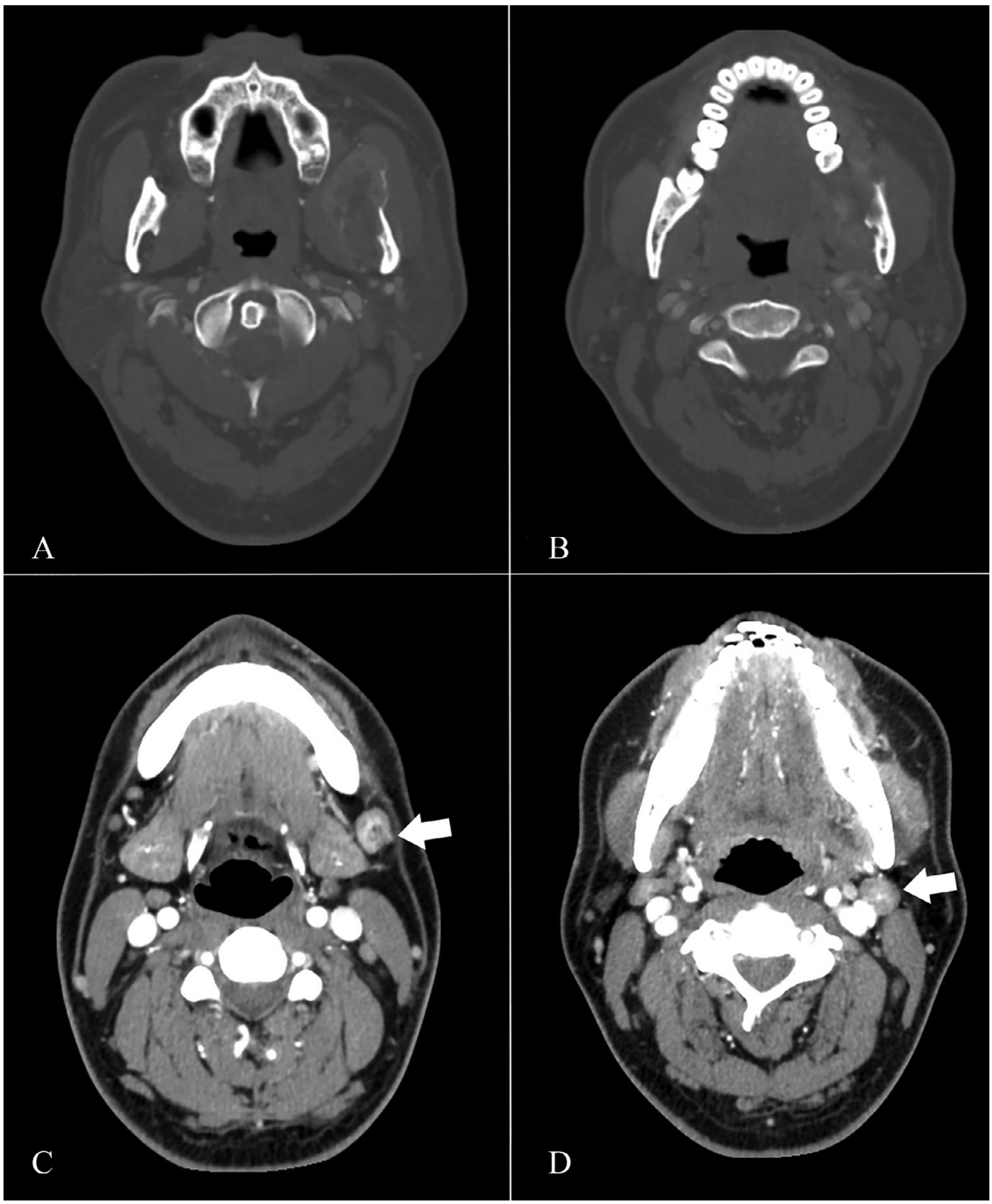

Fig. 2 – (A, B) CT images of bone window (window width: 2000HU, window level: 350HU) showing bone expansion and cortical bone perforation adjacent to the lesion. (C, D) Two enlarged lymph nodes were observed in the left IB and II region (arrows).

while the malignant mesenchymal component was positive for p53 and showed a high proliferation index (>70%) for Ki67 (Fig. 4). Based on the morphology and immunohistochemical staining pattern, a diagnosis of AFS was made. No neck lymph nodes metastases were observed. After surgery and supportive therapy during his 2-week hospitalization, the patient's general condition improved and was released from the hospital. During one-year follow-up, no recurrence was evident.

Discussion

AFS is a rare malignancy with few cases having been reported up to date. According to the World Health Organization classification, it is categorized as odontogenic sarcoma [1]. About 50% of AFSs arise from previously diagnosed AF [1]. Surgical trauma to recurring AF may result in its malignant transformation to AFS [3]. Previous studies show no gender prefer-

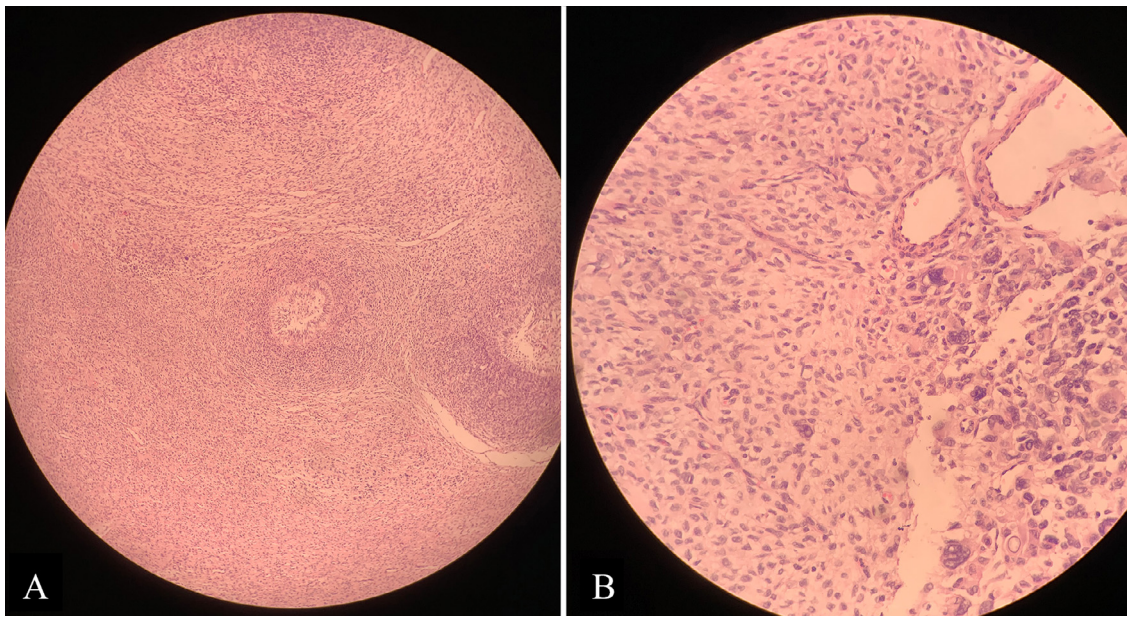

Fig. 3 – Hematoxylin and eosin (H&E) staining. A. Biphasic pattern with benign odontogenic epithelium and malignant mesenchymal components (H&E × 100). B. Malignant mesenchymal components with marked cellularity, nuclear atypia and occasional mitoses (H&E × 400).

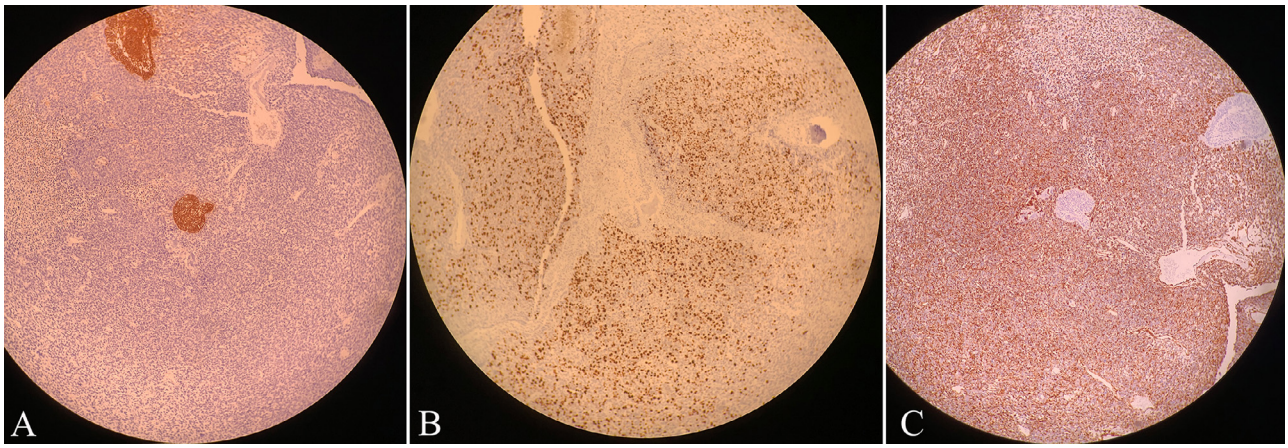

Fig. 4 – A. Cytokeratin AE1/AE3 expression in benign epithelial component (× 100). B. Positive expression for p53 in malignant mesenchymal component (× 100). C. Ki-67 expression in the tumor showing high proliferation index of >70% (× 100).

ence of AFS [2]. The mean age of disease presence is about 27 years [4], although the age range of AFS is quite large (from 3 to 83 years) [5]. In those cases that topographic information was recorded, approximately four fifth of AFS were in the mandible, with the majority of tumor involving the posterior area of mandible [2]. Rarely, AFSs arising from maxilla and skull base were also reported [6,7]. The clinical manifestations of AFS are relatively nonspecific, patients usually experience an expansible mass accompanied with pain and nerve deficit [1].

The characteristic pathological finding of AFS is a biphasic pattern consisting of an epithelial component and a mesenchymal component [1]. The epithelial component may range from lamina-like strands to epithelial islands, and the mesenchymal components can exhibit various mitotic activity [1]. Currently there are no specific immunohistochemical marker recognized as diagnostic for AFS, therefore the pathological diagnosis of AFS is mainly based on hematoxylin and eosin (H&E) staining. Furthermore, histological study provides information on the malignancy, such as Ki-67 and P53 [1, 8]. Moreover, some investigations have suggested that certain biomarkers may be correlated to survival rate of patients with AFS, such as BCL-2 and c-KIT, and may also play a role in the malignant transformation of the disease [6].

In our case, the age at presentation was 29 years old, similar to predilection age [4]. The tumor located in the mandibular

angle, straddling the mandibular angle and ramus, accounting for local pain. Pathologically, in addition to characteristic findings on H&E staining, the mitotic activity was high (>70%), further strongly suggesting the diagnosis of AFS. However, in our case, the relationship between previous tooth extraction and AFS formation remains unclear. On his first presentation at local hospital for occlusal pain, no imaging examination was performed. Consequently, it was unclear whether a preexistent AFS or AF was present at that time. Therefore, a thorough examination on first presentation is suggested, especially for patients with symptoms at specific location. It could also be speculated that tooth extraction might be a surgical trauma, stimulating AFS formation or overgrowth.

For patients with AFS, local invasiveness and recurrence are commonly seen, whereas lymph node or distant metastases are seldom identified [9]. In our case, although two enlarged lymph nodes were detected on pre-operative CT images, only reactive inflammation was observed pathologically, further suggesting that AFS is a low-grade fibrosarcoma [9]. Due to its local invasiveness and recurrence, an aggressive initial treatment of radical surgery with clear margins is highly recommended.

During our clinical work, imaging examination is crucial for diagnosis and surgery planning. For patients at stomatology department, imaging examinations are usually restricted to panoramic radiography, whereas CT could provide more detailed information. However, very sparse data is available on the CT features of AFS. In imaging studies, most AFSs present as radiolucent, poorly marginated lesions, often consisting of multiple cysts (or multilocular), and bone destruction is common [4-7,9,10]. Although non-specific, these imaging features should be paid more attention to, especially for tumorous lesions in posterior area of mandible.

Conclusion

In conclusion, we described a 29-year-old male patient diagnosed with AFS two months after a tooth extraction, with focus on its radiological manifestations. A thorough imaging examination and a better knowledge of its manifestations on CT may prove our diagnostic ability of the rare entity.

Ethical approval and consent

Ethical Approval was waved for this study. Written informed consent was obtained from the participant.

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