The dilemma of fibrous dysplasia versus chronic osteomyelitis of the posterior mandible: a case report

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Abstract

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Key Clinical Message

Dental professionals should be familiar with the radiographic appearance of fibro-osseous lesions and different types of osteomyelitis to distinguish between them and not rely solely on histopathological appearance.

Introduction

Fibrous dysplasia (FD) is a type of hamartoma in which the medullary bone is replaced by immature and poorly calcified bone,[1] whereas osteomyelitis is an infection. Distinguishing between the two conditions is difficult when there is no trauma and no systemic disease. Various authors have reported that despite numerous similarities, useful radiographic differences do exist that help distinguish chronic osteomyelitis from FD.[2-4]

Here we present a case in which we highlight this clinical and radiographic dilemma and describe the differences used to establish diagnosis and administer treatment.

Case Report

A 4-year-old female patient was referred to a maxillofacial surgeon for swelling over the left angle of the mandible, suspected to be an abscess or a tumor. The parents reported that the whole family was recovering from flu-like symptoms likely to be COVID-19 and that the swelling started after the patient became infected.

It was possible, however, that the parents only noticed the swelling at that time, although it was, in fact, there beforehand.

Extraoral and intraoral clinical examination

The extraoral examination showed a painless unilateral bone swelling and facial asymmetry due to swelling of the body and ramus of the left mandible. The swelling was bony hard with no clear signs of paresthesia or inflammation such as fever or redness. Intraorally she had intact and sound primary dentition with no signs of pulpal or periodontal inflammatory lesions.

A solitary palpable lymph node, movable and tender, was noted in the submandibular area measuring approximately 1 cm in diameter.

A panoramic x-ray radiographic examination was made with difficulty due to patient movement, computed tomography (CT) scan were obtained to rule out malignancy or infection, as well as to monitor craniofacial or soft tissue involvement.[5]

Radiographic interpretation

The panoramic X-ray radiograph (Figure 1a) showed overall enlargement of the left side of the mandibular body and ramus with an abnormal bone pattern and an ill-defined lesion extending from the area of the developing mandibular left first permanent molar to the ramus and reaching the coronoid process. The internal structure was heterogeneous, with mixed radiolucency and radiopacity, or a ground glass appearance, having a granular bone pattern with a haphazard distribution. The lesion did not seem to be centered in the alveolar ridge and involved bone inferior to the mandibular canal; the teeth appeared to be intact. The 3D CT image (Figure 1b) showed expansile disease affecting the left side of the mandible (body and ramus), causing asymmetry in the lower third of the face. The multiplanar view (Figure 1c) showed a well-defined radiopaque expansile lesion from the buccal and lingual aspects (axial view) and a moth-eaten appearance accompanied by ground glass attenuation occupying the left ramus and part of the body of the mandible (sagittal view), as well as thinning of cortices and an abnormal bone pattern (axial and cross-sectional views). The expansile lesion in the medullary space showed variable attenuation with mixed radiolucency and radiopacity, loss of trabecular bone architecture, and periosteal new bone formation on the lingual side of the mandible (Figure 1c, cross-sectional view).

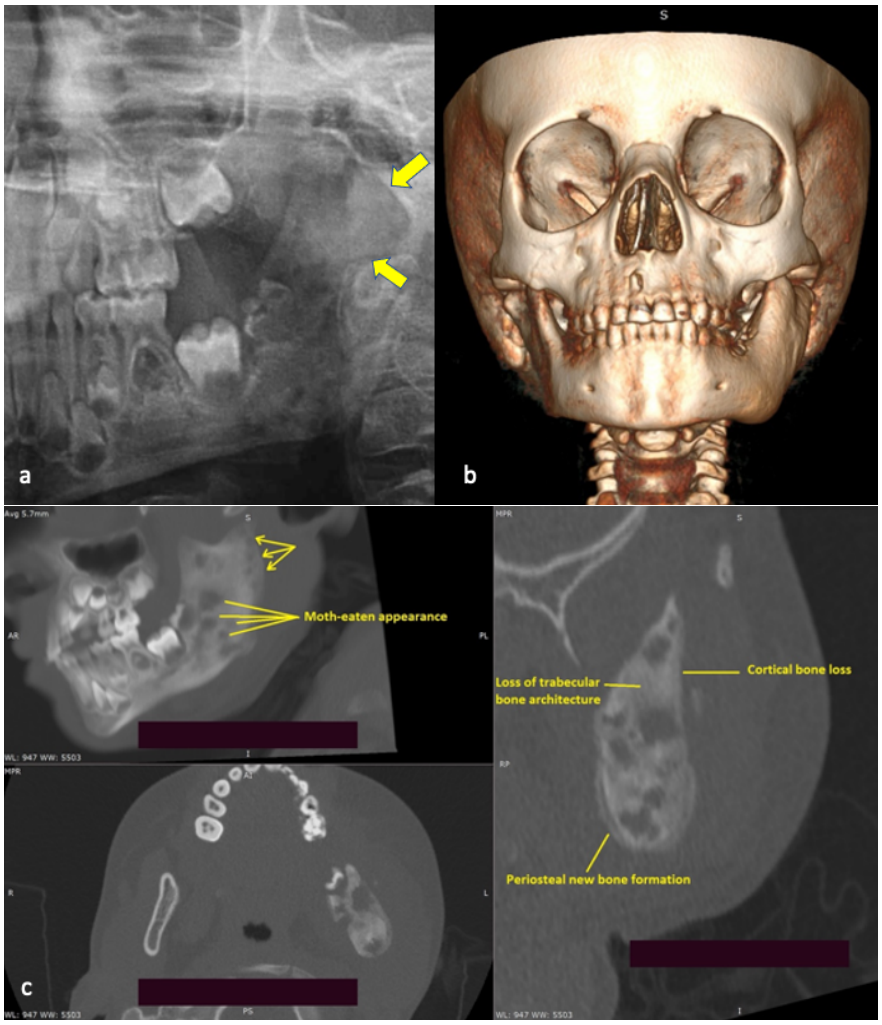

Figure 1 (a) Panorama radiograph :ill-defined lesion extending from the area of the developing mandibular left first permanent molar to the ramus, reaching the coronoid process. (b) The volumetrically rendered CT image showing left mandible enlargement. (c) Multiplanar views: sagittal, axial, and cross-sectional views with radiographic characteristics of osteomyelitis.

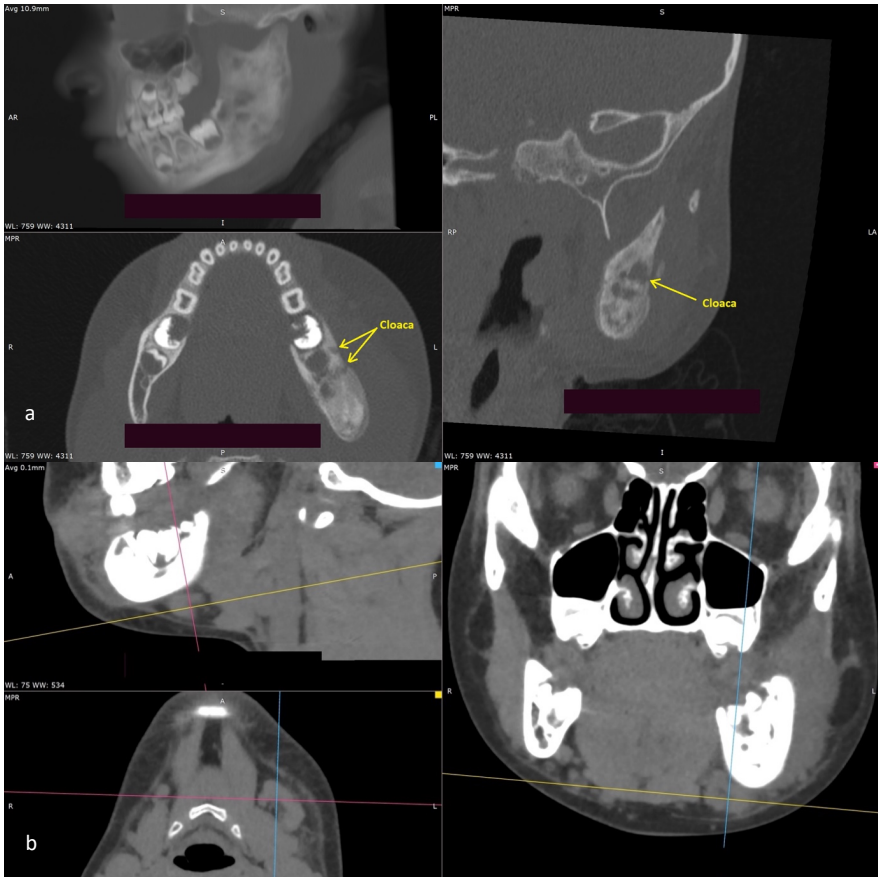

Figure 2 (a) Multiplanar view of the "bone window": sagittal, axial, and cross-sectional views showing a cloaca, which is characteristic of osteomyelitis. (b) Multiplanar view of the "soft tissue window": sagittal, axial, and cross-sectional views showing submandibular lymph node involvement.

The multiplanar view of the "bone window" (Figure 2a) showed a cloaca in the cross-sectional and axial views, which is the beginning formation of a draining fistula; the well-defined break in the outer cortex is an involucrum, which allows drainage of purulent and necrotic material out of dead bone. The multiplanar view of the "soft tissue window" (Figure 2b) showed submandibular lymph node enlargement in the sagittal, axial, and coronal views.

All of these radiographic features are associated with osteomyelitis and may sometimes be associated with severe fibro-osseous lesions.[6] The extensively dysplastic bone and limited blood supply in a compact mandible that already has less collateral circulation than the maxilla will produce an ischemic condition that may result in such a presentation, especially in a very young patient. In light of the chief concern, patient history, extra- and intra-oral examination, and radiographic investigations, a diagnosis needed to be confirmed histologically. The patient's parents were informed about the surgical plan, and informed consent was obtained. Inflammatory markers were measured, and the results of blood investigations were all well within the normal range. Mandibular shaving was performed, and a biopsy sample taken; there was no pus and no culture was done.

Histopathological examination

The following results of the histological examination were obtained (Figure 3 a and b):

- bone tissue exhibiting branching and anastomosing irregular trabeculae of woven bone with no conspicuous osteoblastic rimming
- intervening fibrous stroma containing cytologically bland spindle cells
- no mitotic figures

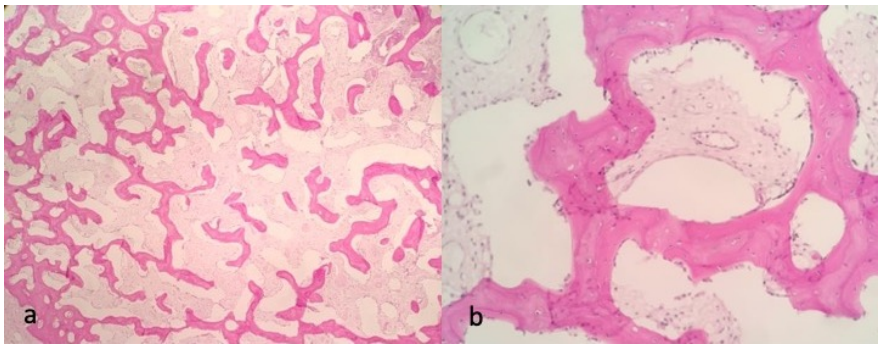

Figure 3 (a) Trabecular pattern in fibrous stroma seen in (5X) magnification. (b) The (30X) magnification shows "Chinese letter" trabecular pattern

The final diagnosis was fibrous dysplasia. A few months later, the patient returned with a relapse of the lesion accompanied by pain. Another CT scan showed the same radiographic appearance that was seen in the first and second CT scans: a sequestrum, an involucrum, and periosteal new bone formation surrounding the sequestrum, in addition to periosteal reaction thickening (periostitis) and fistula formation (cloaca), highly suggestive of osteomyelitis and inconsistent with FD. Also this later CT (after 3 months) showed wider involvement of the mandible that is getting closer to the condylar head (Figure 4a), an increase in the size and number of lytic lesions, more sequestrum formation (Figure 4b), and swelling and blurring of normal soft tissue planes (Figure 4c). The patient has been followed up since October 2020, her most recent follow-up being in January 2021.

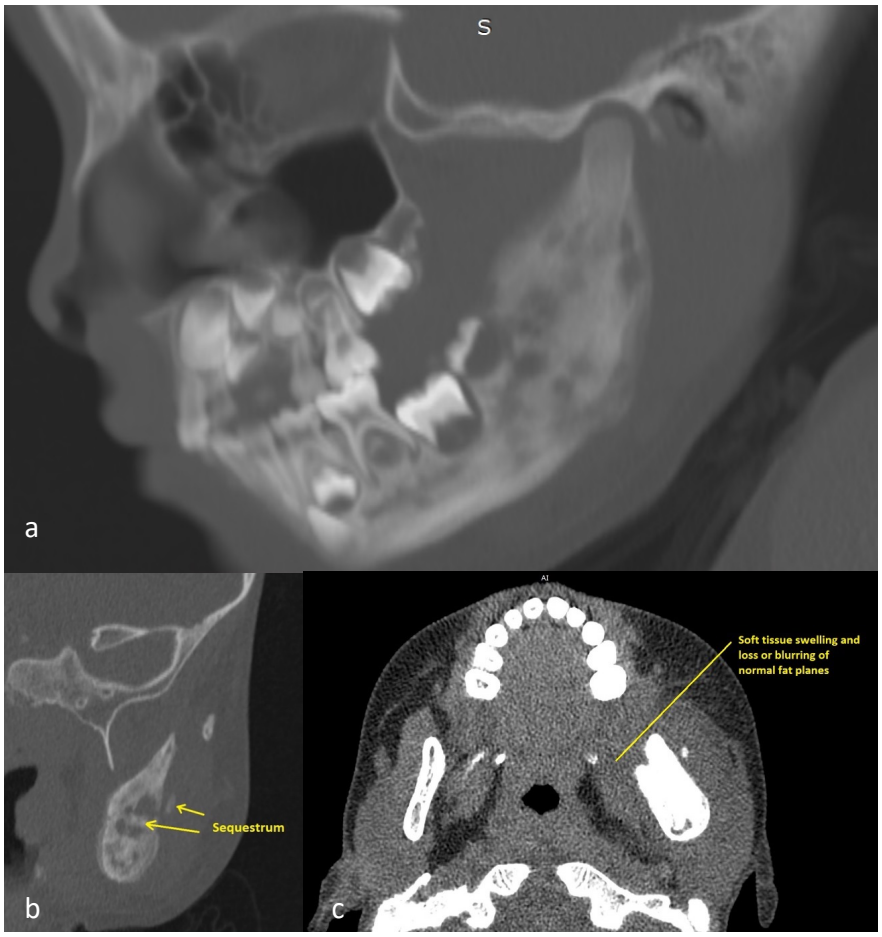

Figure 4 (a) Sagittal view showing the spread of the lesion to the condyle. (b) Cross-sectional view showing the sequestrum. (c) Axial view of the "soft tissue window," showing soft tissue swelling and loss or blurring of normal fat planes.

Discussion

FD lesions typically manifest during the first and second decade of life, and cases have been reported as early as 3 years of age.[7] Monostotic FD affects the mandible, typically the unilateral posterior area with no specific sex predilection.[8] The bone becomes enlarged but maintains its overall shape, an ill-defined border gradually blends with surrounding bone, and the lesion bone pattern may have a ground glass, cotton wool, or dense amorphous appearance. The lesions usually begin growing at a young age and stop at the end of somatic growth. In the present case, the radiographic appearance of the lesions was typically mixed radiolucency and radiopacity, producing a characteristic ground glass appearance, but this depends on the stage of the disease, as early lesions tend to be radiolucent and become more opaque as they progressively calcify.[9,10] Although the FD margins were generally poorly defined on CT for the mandibular lesions, one factor could be the relatively thick (5 mm) sections. CT images of FD on bone windows can display a range of opacification observed on plain radiographs, such as radiolucency, ground glass, and cotton wool.[11]

Chronic sclerosing osteomyelitis, an inflammatory bone condition of uncertain origin, was suggested in the differential diagnosis, as it is generally believed to be an infection of etiological significance, and recurrent pain and swelling of long duration are commonly reported symptoms. However, on occasion, mild and often nonspecific symptoms make a definitive diagnosis difficult. A study reported that chronic sclerosing

osteomyelitis involves the ramus in 16.9% of cases and both the body and the ramus in 14.3% of cases.[12] Painful swelling was reported in only 13.8% of patients,[13-15]trismus in 15.1%,[16] and fever in 7.7%.[12] The left mandibular side seems to be slightly more affected (54.0% of cases) than the right side (41.5% of cases). Systemic signs such as body temperature, white blood cell count, and erythrocyte sedimentation rate are either within normal range or slightly elevated,[17] similar to those our case. Obtaining a positive culture for the responsible microorganisms from the lesion may be either difficult or nonrepresentative because of contamination with skin or oral microflora.

In our case, the histopathological appearance was of a typical "Chinese letter" trabecular pattern. However, studies have shown that the histopathological appearance of osteomyelitis can feature replacement of normal marrow with fibrous connective tissue, sometimes accompanied by an inflammatory cell infiltrate, new bone formation, and the presence of bony sequestra. The inflammatory cell infiltrate is sparse or difficult to detect. Occasionally, the appearance is similar to that seen in FD or osteogenic sarcoma.[17,18]

Chronic non-suppurative osteomyelitis with proliferative periostitis (PP) was also suggested in the differential diagnosis, as there was no demonstrable cause.[12,19] A hypothesis was therefore raised to explain our case: that the infection was caused by a local low-virulence bone infection, such as COVID-19 . However, investigations could not confirm this hypothesis. Suggestive of osteomyelitis of unknown origin were sequestra and laminated "onion-skin" periosteal new bone as seen in Figure 1c; mixed sclerosis and bone destruction as seen in Figure 1c, the presence of a cloaca and sinus tract as seen in Figure 2a, and the presence of a submandibular lymph node on the same side as seen in Figure 2b. loss of normal fat as seen in Figure 4c.

Many classifications of chronic osteomyelitis have been proposed, depending on clinical aspect or origin (bacterial or inflammatory). Newly formed periosteal bone was organized, at early stages of the disease, in different aspects, either with an "onion skin" appearance (33.8% of cases), lamellae duplication (12.3% of cases), or sunburst aspect (3% of cases).[12] PP occurred predominantly on the buccal side of the mandible (85% of cases). In the present case, PP was present buccally and lingually.

According to Bisseret et al,[20] the appearance of the underlying cortex in a periosteal reaction is an important criterion to assess the aggressiveness of osteomyelitis and is more useful than the pattern of the periosteal reaction. Thus, the destruction of the cortex and a "moth-eaten" appearance, as seen in Figure 1c, denotes aggressive disease.

Differential diagnoses

Before making a diagnosis of chronic non-suppurative osteomyelitis with PP, numerous other causes of periostitis should be considered, in particular if no obvious etiological factor exists. Juvenile mandibular chronic osteomyelitis and chronic osteomyelitis should be excluded. A differential diagnosis must then be made with isolated periostitis associated with osseous benign lesions, such as giant cell granuloma, FD, or osteoid osteoma.[20] Malignant tumors, such as Ewing's sarcoma, chondrosarcoma, and osteosarcoma usually present more aggressive symptoms and soft tissue involvement. In these pathologies, a periosteal reaction radiographically resembles a "sun ray" or an "onion skin" pattern.[20]

Systemic diseases that may be responsible for secondary periostitis in pediatric patients should be excluded such as metabolic disorders, hematologic malignancies (leukemia, lymphoma, Langerhans cell histiocytosis), sickle cell anemia, and vasculitis.[21]Bacterial mandibular osteomyelitis lesions are characterized by suppuration, osteolytic radiographic changes with lamellar-type periosteal reaction, and an impressive response to antibiotic administration.

Although primary chronic osteomyelitis is not age related, most of the relevant published data refer to adult patients, and only case reports or small series of patients with early onset of the disease during childhood or adolescence are encountered in the literature. Heggie et al[22] and Baltensperger et al[23]observed a high incidence and uniformity of features of the disease among children and adolescents, respectively, proposing that the pediatric "variation" should be regarded as a separate clinical entity and suggesting the term "juvenile chronic osteomyelitis" for its description.[24]

Conclusion

Dental professionals should be familiar with the radiographic appearance of fibro-osseous lesions and different types of osteomyelitis to distinguish between them and not rely solely on histopathological appearance. The distinction can aid patient diagnosis, care, and treatment in accordance with the different pathogenic processes involved.[25]

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Conflict of interest

The authors have no conflict of interest to declare.

Proof of consent

I confirm that the patient consent has been signed and collected in accordance with the journal's patient consent policy . I will retain the consent form and will provide it if requested**.**

Authors' contributions

Below are all authors of this work and a brief description of how each contributed towards the submission:

Authors names	Contribution
Ebtihal Zain-Alabdeen	Introduction, Discussion, Data collection
Ashraf Abdelfattah	Clinical and Pathological examination, biopsy
Osama Kurdi	Past medical history and histopathology
Ra'ed Al-Sadhan	Radiographic interpretation and figures selection

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