

Periosteal reaction as a crucial radiographic finding for desmoplastic fibroma of the jaw bone in children: A case report

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ABSTRACT

This report presents the case of a 5-year-old boy with a hard swelling on the right side of the mandible body. An important point of this case is that the primary imaging finding was fine spicules in the inferior border of the mandible on panoramic radiography without significant changes in bone density. Cone-beam computed tomography views revealed a lytic lesion on the lingual side of the right mandibular body with the destruction of the lingual cortex and periosteal reaction from the midline to the first molar area. Careful attention to this radiographic finding in the primary stage in the absence of other significant imaging findings, particularly in children, could result in the early diagnosis of desmoplastic fibroma. Therefore, a better prognosis can be expected following early surgical treatment. (*Imaging Sci Dent* 2022; 52: 319-26)

KEY WORDS: Fibroma, Desmoplastic; Diagnostic Imaging; Mandible; Odontogenic Tumors

Odontogenic tumors include a wide range of lesions, from tumor-like malformations to benign and malignant lesions originating from odontogenic tissues, which can develop with disturbances in growth and proliferation signals.^{1,2} There is also another group of odontogenic tumors, which belong to a benign category with locally aggressive behavior characterized by their inherent potential for invasion and destruction and a high recurrence rate. These lesions require special attention for early detection and effective treatment to reduce subsequent morbidity.²

Desmoplastic fibroma is the bony counterpart of desmoid-type soft tissue fibromatosis.^{3,4} Desmoplastic fibroma arising in bone is an aggressive and infiltrative neoplasm. The mandible is the most common site of involvement in the craniofacial region, mainly in the posterior region (ramus, angle, and molar area), followed by long bones and the pelvis.^{3,5-7} Causative links with trauma and endocrine or genetic factors have been suggested in

the literature, but the etiology of desmoplastic fibroma remains unknown.⁸ Asymptomatic swelling followed by tooth displacement and mobility could be considered the most representative clinical findings.⁶

Although most authors reported the occurrence of desmoplastic fibroma in patients younger than the third decade of life,^{4,6,9} some reports described a broader age range, from 6 months to 60 years at the time of diagnosis.¹⁰ Diagnosis at older ages may reflect late recognition. An equal sex distribution has been reported in most studies,^{3,6,11} but Wood et al.⁹ stated there was a female predilection (56%). The most common clinical feature of desmoplastic fibroma of the jaw bones is painless swelling; the others include facial asymmetry, tooth displacement and mobility, trismus, and pathological fractures,^{3,4,6,7,12} although the latter is more common in long bones.⁸

An accurate diagnosis depends on various factors including clinical signs and symptoms, the patient's medical history, and laboratory and radiographic examinations. Diagnostic imaging is an essential step to assess bone lesions and their extent.¹³ Panoramic radiography as a conventional method is the first step of evaluation. However, 3-dimensional techniques such as cone-beam computed

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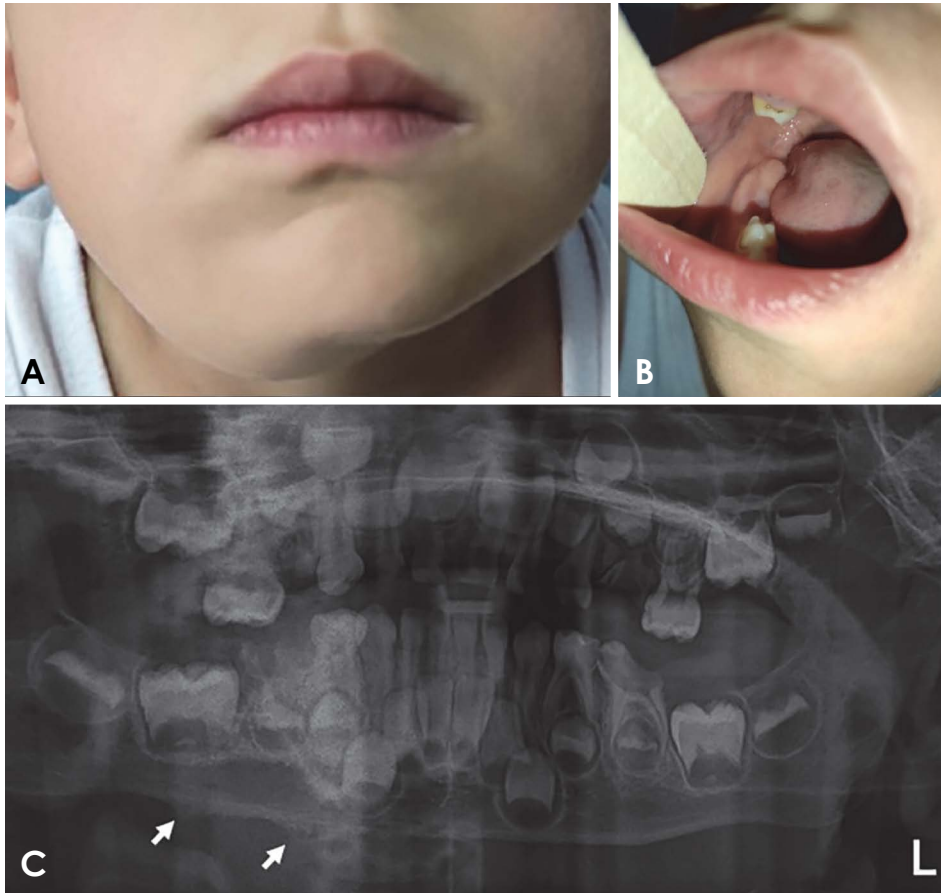


Fig. 1. A. Extraoral photograph shows swelling of the right side of the mandible. B. Intraoral photograph shows normal overlying mucosa. C. Panoramic radiograph reveals fine spicules on the inferior portion of the mandible in the right first premolar and first molar areas (white arrows).

tomography (CBCT), with its low costs and radiation dose compared to multidetector computed tomography, have prompted a revolution in the field of radiology. CBCT imaging with volumetric reconstruction and multiplanar reformation plays an essential role in the accurate assessment of osseous lesions in the maxillofacial region.^{14,15}

Regarding radiographic features, desmoplastic fibroma could present as a well- or ill-defined radiolucency with a unilocular or multilocular appearance. In addition, cortical expansion or destruction and cortical perforation due to rapid expansion may cause new bone formation with a “sunburst” appearance, mimicking osteogenic sarcoma or Ewing sarcoma.¹⁰ Histologically, desmoplastic fibroma contains spindle-shaped fibroblastic or myofibroblastic cells within a collagenous background with focal myxoid changes, but cellular mitoses or atypia are not present.^{4,11} Some authors placed it in a borderline position relative to malignancy because of its infiltrative nature.³

This report aimed to introduce a radiological finding that could alert clinicians to the presence of desmoplastic fibroma and therefore could help detect the tumor in its early stages, facilitating proper treatment and a favorable prognosis.



Fig. 2. Ultrasonography of the region reveals a hypoechoic solid mass with lobulated margins and dimensions of 32 mm × 26 mm × 20 mm, accompanied by destruction of the mandibular cortex.

Case Report

The ethical approval number of this case report is IR.GUMS.REC.1400.508. A 5-year-old boy with a tender and hard bony swelling on the right side of the mandible body that had lasted for 3 months was referred to the Department of Maxillofacial Surgery of Guilan University of

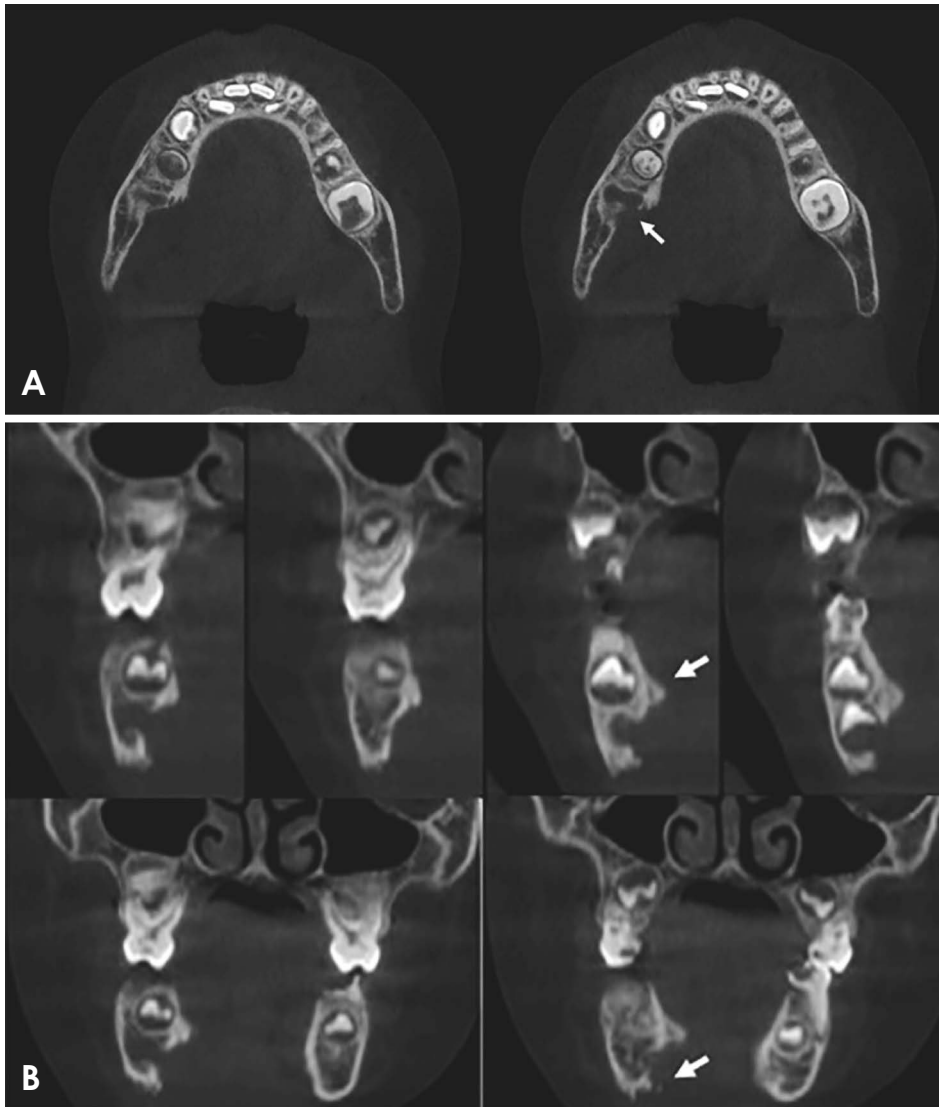


Fig. 3. Axial (A) and coronal (B) cone-beam computed tomographic images show the destruction of the lingual aspect of the right side of the mandible and periosteal reaction near the inferior border (white arrows).

Medical Sciences by a dentist. The patient had gross swelling in the right side of the mandible and a history of extraction of the second primary molar, but without improvement. There was no history of systemic disease, fever, trismus, or odynophagia. The first panoramic radiograph showed fine spicules arising from the inferior border of the mandible on the right side, in the first premolar and molar regions (Fig. 1). Unfortunately, the dentist missed this abnormal finding on the panoramic radiograph.

Despite using clindamycin, ceftriaxone, and metronidazole, the swelling persisted. Ultrasonography of the region showed a hypochoic solid mass with lobulated margins and dimensions of 32 mm × 26 mm × 20 mm with slight vascularity that had destroyed the mandibular cortex. There was no abnormality in the right submandibular gland or cervical lymph nodes (Fig. 2).

The first CBCT scan was taken 3 months after the first panoramic radiograph. Axial, coronal, and cross-sectional CBCT views revealed a lytic lesion on the lingual side of the right mandibular body with destruction of the lingual cortex, extending from the deciduous first molar to the permanent first molar. Nevertheless, in axial views, bony spicules in the lingual cortex near the inferior border of the mandible, as indicators of a periosteal reaction, were detected from the midline to the first molar area (Fig. 3). Desmoplastic fibroma, eosinophilic granuloma, Ewing sarcoma, osteosarcoma, and lymphoma constituted the first radiographic differential diagnosis list even though the histopathological report of the incisional biopsy was fibrous dysplasia. The results of the histopathological report were not convincing; therefore, the patient was followed for a few months.

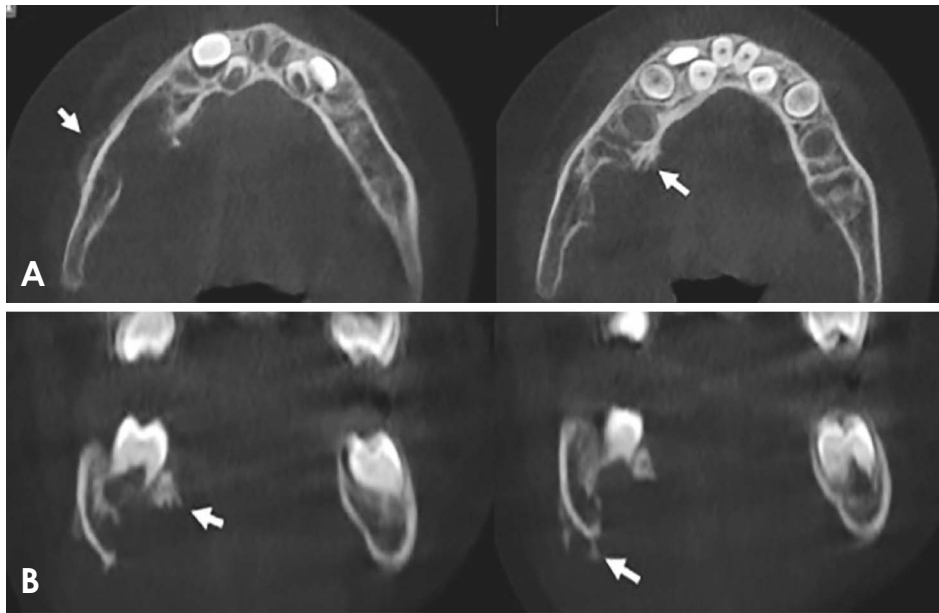


Fig. 4. Axial (A) and coronal (B) cone-beam computed tomographic images 3 months after the first biopsy show progressive bone destruction and periosteal reaction (white arrows).

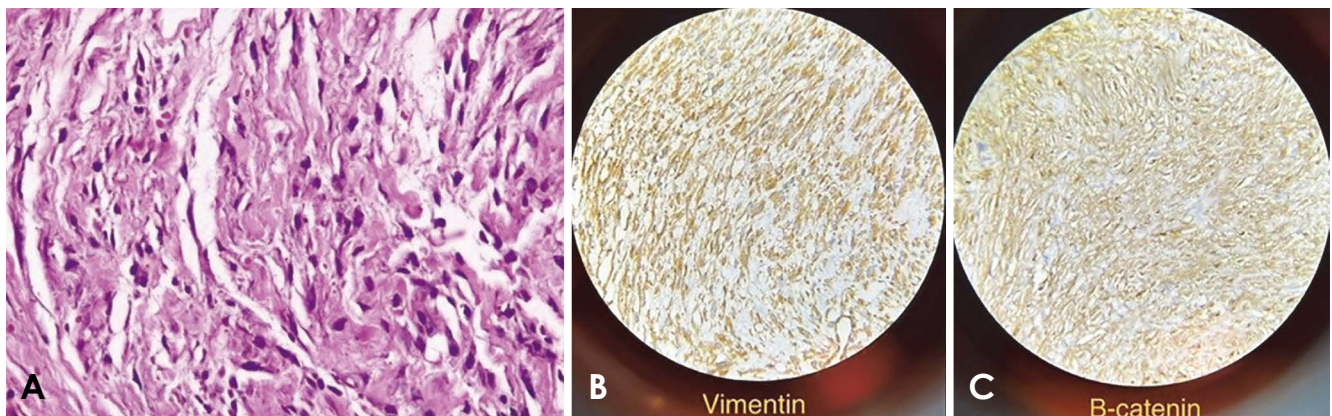


Fig. 5. A. A histopathological examination reveals spindle cell proliferation arranged in streaming fascicles, which are associated with a variable amount of collagen and partial myxoid changes. In addition, immunohistochemical tests show positive findings for vimentin (B), and β -catenin (C).

The second CBCT scan was taken 3 months after the first biopsy, which demonstrated greater extension of the lesion (Fig. 4). Unfortunately, the patient's parents refused surgical treatment. After 2 months and more exacerbation of the swelling, his parents accepted the second biopsy. The biopsy showed pieces of neoplastic tissue composed of sheets of proliferated, almost monotonous spindle cells with mild cellularity. Significant mitotic activity or tissue necrosis were not detected. A bundle of skeletal, muscular tissue was focally present at the periphery of the lesion surrounded by proliferated spindle cells, and immunohistochemical tests for vimentin and β -catenin were positive (Fig. 5). According to the histological and immunohis-

tochemical features, the lesion was reported as benign locally aggressive spindle cell neoplasm with a differential diagnosis of desmoplastic fibroma (of bone with extra-mandibular soft tissue extension), fibromatosis (of soft tissue with secondary extension into the mandibular bone), and myofibromatosis.

The third set of CBCT images and panoramic radiography, taken 2 months after the second CBCT scan and the personal decision of his parents for total surgical resection of mandible, showed marked progression of bone destruction and discrete new bone formation in the lingual and labial cortical plates, as well as the inferior border of the mandible, which represented a sunburst and layering

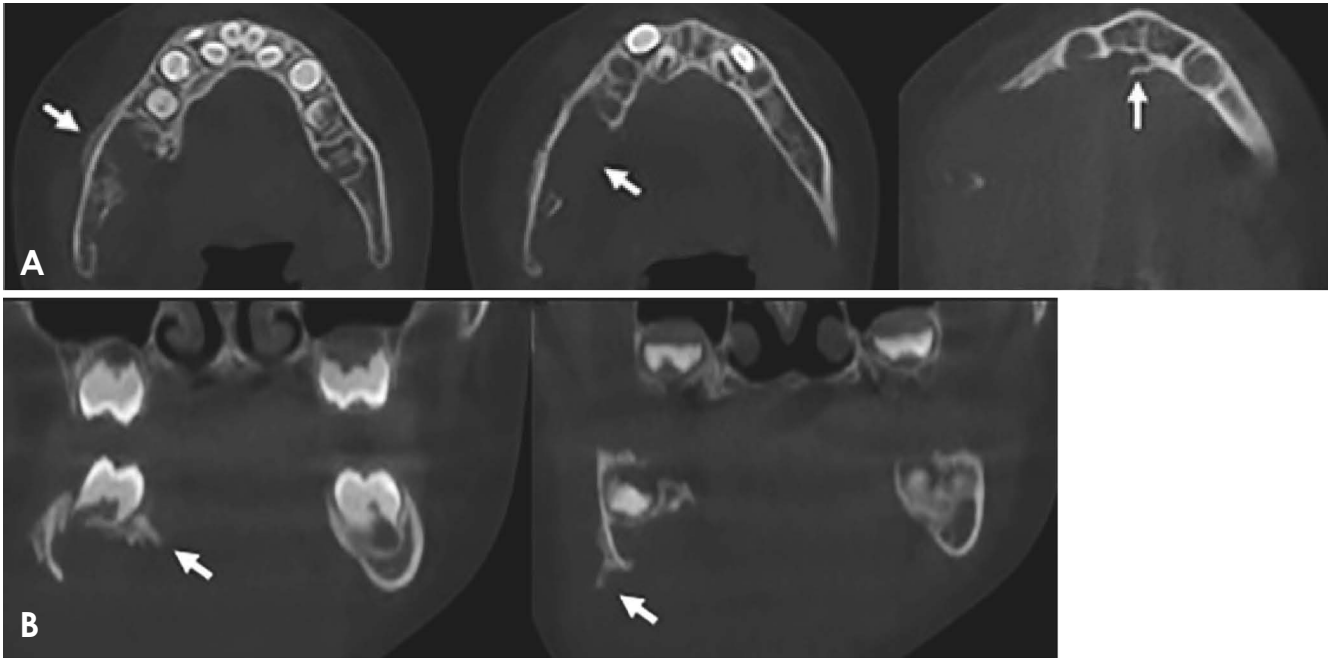


Fig. 6. A. Axial and coronal cone-beam computed tomographic images after 5 months reveal vast destruction of the inferior, lingual, and buccal cortices, periosteal reaction and root resorption of the right mandibular first molar, and displacement of the developing first and second premolars. The lesion also crosses the midline. B. Panoramic radiograph shows destruction on the right side of the mandible and root resorption of the right first molar.

form of periosteal reactions (Fig. 6). The lesion crossed the midline, and it was apparent in axial views at the level of the inferior third of the mandible and the lingual side.

After giving the information about the growth pattern of the lesion to the patient's parents, the treatment plan (e.g., initial reconstructive surgery) and the need for future dental rehabilitation, close follow-up, and a delayed second operation with an autogenous bone graft were explained to them in order to obtain their permission.

Eventually, surgical resection of the mandible with a wide margin was performed, with simultaneous reconstruction using a surgical plate (Fig. 7A). The patient was included in a close follow-up plan because of the high recurrence rate. Based on the clinical, radiological, and histopathological examinations, the final result was desmoplastic fibroma of the mandibular bone. Fourteen months after surgery, follow-up panoramic radiography revealed a safe and intact margin along the surgical site, without recurrent lesions (Fig. 7B).

Discussion

The radiological features of desmoplastic fibroma are nonspecific and variable. Desmoplastic fibroma can present as a unilocular or multilocular radiolucency, with a

well- or ill-defined margin, as a totally radiolucent or radiolucent-radiopaque lesion, and as an expansile tumor with cortical thinning or destruction. Occasionally, lesions with rapid expansion and subsequent cortical perforation imitate malignancies.^{10,15} Rapid expansion and subsequent cortical perforation can also result in the initiation of new bone formation from an osteogenic layer of periosteum, producing a sunburst appearance.^{6,9,11} Similarly, in the present case, bony spicules as an indicator of a periosteal reaction arising from the cortical plates of the mandible were visible, and layering bone formation in the labial cortex was also seen. Since the periosteal reaction originates from the periosteum, it may present only as fine spicules on conventional radiography in the initial stages, as seen in the present case and also in the case reported by Dalili-Kajan et al.,¹¹ Khatib and Pogrel,¹⁵ Said-Al-Naief et al.,¹⁶ and Shi et al.¹⁷

Based on radiographic features, malignant lesions such as osteosarcoma, Ewing sarcoma, lymphoma, and eosinophilic granuloma were considered in the differential diagnosis list. Nonconcentric periodontal space widening, called the "Garrington sign,"⁵ is a common radiographic feature in the alveolar regions in malignant lesions, absent in the present case. Although sclerotic-appearing bone can be seen in extensive osteosarcomas, other malignant

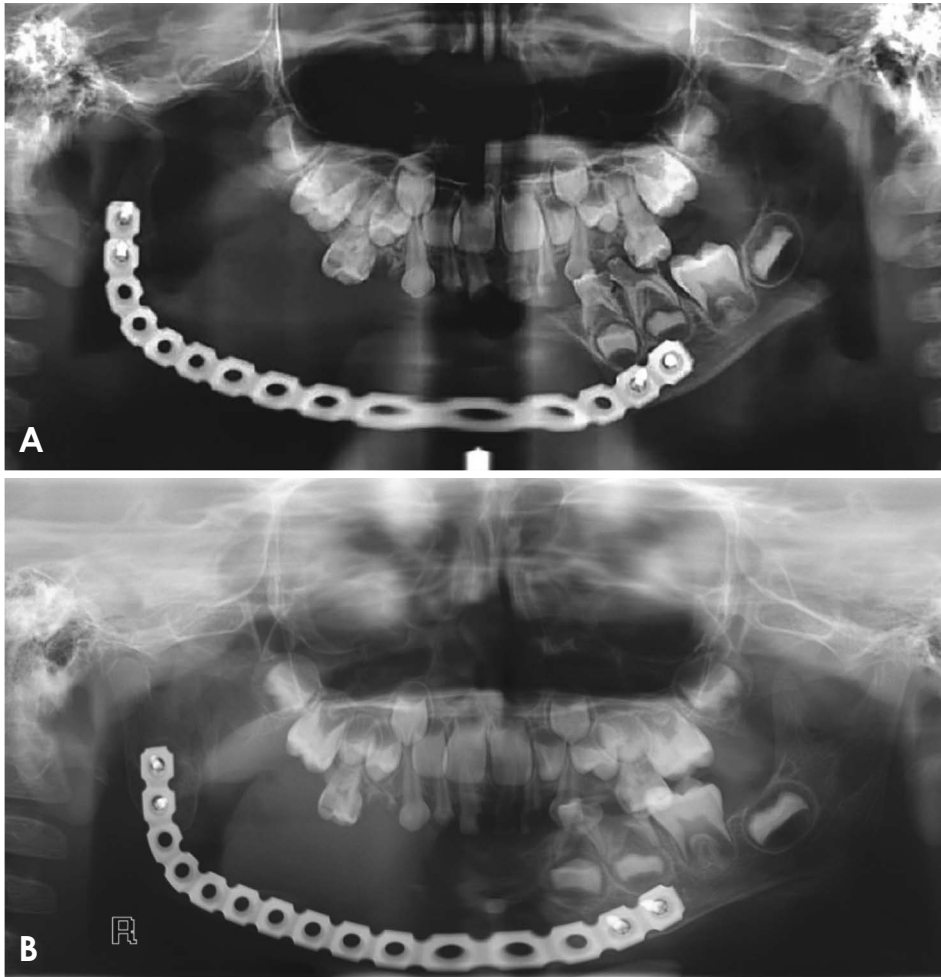


Fig. 7. A. Postoperative panoramic radiograph reveals the resection of the mandible with a wide margin. B. Follow-up panoramic radiograph after 14 months does not show a recurrent lesion.

lesions are usually completely radiolucent without any internal structure. Furthermore, eosinophilic granuloma is a multiple and well-demarcated lesion involving the mid-root epicenter of the tooth. Therefore, its early clinical finding is tooth mobility. It can also appear as punched-out lesions. Although periosteal new bone formation is not typical in eosinophilic granuloma, it could be found in the other lesions.⁵ It usually appears in their final stage when extensive osteolytic changes occur. The critical point in the present case is periosteal reaction as a primary imaging finding that could be a pivotal point in the early detection of desmoplastic fibroma.

In a histological evaluation, desmoplastic fibroma is characterized by uniform-appearing fibroblast cells in a stroma containing various amounts of collagen fibers.³ The lack of a capsule and the infiltrative nature of the lesion are hallmarks of desmoplastic fibroma. Low-grade fibrosarcoma and low-grade intraosseous osteosarcoma are the most challenging differential diagnoses.³ Hypercellularity with a herringbone pattern and higher mitotic activity in

fibrosarcoma can help to differentiate it from desmoplastic fibroma. In addition, low-grade osteosarcoma can be distinguished by osteoid formation.³ It is worth noting, in the first histopathological report, that fibrous dysplasia may be due to an inadequate depth of the biopsy specimen.

In addition, the immunohistochemical analysis could be helpful for a better diagnosis.⁸ The cells of mesenchymal origin have a variable response to vimentin.¹⁸ β -catenin is a cytoplasmic protein regulated by the *APC* gene, which is involved in tumorigenesis.¹⁹ First, β -catenin binds to E-cadherin, and this complex helps inhibit and suppress the invasion of tumor cells. Its second role is in regulating cell proliferation and maintaining the normal cellular architecture. An increase in β -catenin expression can activate oncogenes and lead to various neoplasms and cancers.^{20,21} Desmoid-type fibromatoses and adenocarcinoma have positive results for β -catenin.²⁰ Multiple cases of desmoplastic fibromas of the jaw bones have been reactive to β -catenin,²²⁻²⁴ while some have not been.^{12,22,25}

The comparative immunohistochemical findings of

desmoplastic fibroma, fibrous dysplasia, and low-grade fibrosarcoma reveal that desmoplastic fibroma is low for Ki-67, positive for vimentin and smooth muscle actin (SMA), variable for β -catenin, negative for S100 and MSA, whereas low-grade fibrosarcoma is high for Ki-67, positive for vimentin, p53, and reticulin, and negative for S100, keratin, and SMA. Meanwhile, fibrous dysplasia has none of these findings.⁹ In the present case, the lesion was positive for vimentin and β -catenin.

Various rates of recurrence have been reported, ranging from 17% to 67%, depending on the treatment method.⁶ Increased cellularity seems to be associated with an increased tendency for recurrence.¹² Various treatment approaches have been suggested for desmoplastic fibroma, including enucleation, curettage, resection, radiotherapy, and chemotherapy.⁶ Radiotherapy, because of its adverse effects such as fibrosis, paresthesia, pathological fracture, higher bone graft failure rates, and induction of osteosarcoma, is not recommended.¹⁵ In most clinicians' opinions, surgical resection of the lesion with a wide margin is the preferred method to prevent recurrence.^{6,11} Regarding the high rate of recurrence, long-term follow-up is necessary. Although there are no specific guidelines for radiographic follow-up, most reports have recommended a radiographic follow-up for a minimum of 3 years.¹⁰

Simultaneous bone reconstruction seems to be essential for patients to return to function. This issue is also essential because most patients are of growing age, and delayed or improper reconstruction may result in craniofacial abnormalities and malocclusion, which could affect their quality of life.

Although the small size of facial structures, limited donor sites, growth procedures, and small vessels are challenging factors in the reconstruction of pediatric bony defects, healthy vessels and a superior ability to heal result in a high rate of successful treatment.²⁶

Reconstruction of mandibular defects can begin immediately postoperatively, but it is logical to consider a time interval to ensure a lack of recurrence. Usually, as in the case presented herein, reconstruction plates are used for immediate reconstruction, after which a second operation with a delayed autogenous bone graft and subsequent dental rehabilitation are performed to restore function.²⁷

In conclusion, the periosteal reaction and arising bony spicules should be considered a novel, essential, and sometimes the only radiographic finding of desmoplastic fibroma of the jaw bone in children in the early stage. An early diagnosis with clinical and advanced radiographic evaluations, such as CBCT, and subsequent proper treat-

ment result in the preservation of more tissue and less morbidity, with expectations for a better prognosis. It should be noted that selecting the proper location of biopsy is very crucial in these cases. Imaging features can help clinicians avoid missing the diagnosis because histopathological findings cover a wide range of differential diagnoses.

Conflicts of Interest: None

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