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Case Report

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Desmoplastic fibroma of the mandible in a child: a case report with more than 3 years of radiographic follow-up

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Running title: Pediatric mandibular desmoplastic fibroma

Abstract

Background: Desmoplastic fibroma (DF) is a rare, benign but locally aggressive intraosseous fibroblastic neoplasm. Jaw involvement is uncommon, and diagnosis may be challenging because its clinical and radiographic appearance overlaps with other odontogenic and fibro-osseous lesions.

Case presentation: We report a pediatric case of left mandibular DF in a 6-year-old boy. Imaging demonstrated an osteolytic destructive lesion involving the left mandibular body, with cortical destruction and close relationship to the inferior alveolar canal and developing permanent teeth. Histopathologic examination of an incisional biopsy showed a hypocellular spindle-cell neoplasm arranged in fascicles within abundant collagen, with mild pleomorphism, no necrosis, and fewer than 1 mitosis/10 high-power fields. Immunohistochemistry showed tumor-cell positivity for smooth muscle actin, focal desmin and β-catenin, negative H-caldesmon and S100, and a low proliferative index of approximately 1–2% by Ki-67. The overall clinicoradiologic and pathologic findings favored DF. The lesion was surgically treated and followed by serial radiographic surveillance. Follow-up imaging documented progressive mandibular remodeling without definite evidence of recurrence through approximately 3 years after treatment.

Conclusion: This case highlights the importance of clinicoradiologic-pathologic correlation in pediatric mandibular DF and the value of prolonged surveillance after surgery. 1-cm surgical margin was documented in the operative report. Published mandibular DF cases have similarly used 1-cm safety/free margins in selected resections, supporting the adequacy of this margin in the present case.

Keywords: desmoplastic fibroma; mandible; pediatric; spindle-cell tumor; β-catenin; oral and maxillofacial surgery; case report

Background

Desmoplastic fibroma is a rare primary intraosseous fibroblastic tumor that is histologically benign but may behave aggressively at the local level. It is regarded as the osseous counterpart of soft-tissue desmoid-type fibromatosis and accounts for less than 1% of primary bone tumors. In the jaws, the mandible is affected substantially more often than the maxilla, with a predilection for the posterior mandible. [1–4]

The clinical and radiographic presentation is nonspecific. Lesions may be unilocular or multilocular, expansile, and radiolucent, sometimes with ill-defined margins and cortical perforation. These findings can mimic ameloblastoma, odontogenic myxoma, aneurysmal bone cyst, fibro-osseous lesions, or low-

grade sarcomas. Consequently, diagnosis requires integration of imaging and histopathology, with immunohistochemistry used when the spindle-cell differential is broad. [2,3,5]

Surgical management remains individualized. Because the tumor is infiltrative and recurrence is associated with incomplete removal, complete excision with histologically clear margins is generally favored when morbidity is acceptable. Conservative curettage or enucleation has also been reported, particularly in very young patients when preservation of mandibular growth and developing dentition is prioritized. [1–6]

Case presentation

Patient and initial assessment

A 6-year-old boy was evaluated for a lesion involving the left mandible. The available clinical documentation identifies the lesion as an expansile/destructive mandibular lesion.

Radiographic findings

Preoperative panoramic and CBCT imaging demonstrated a lytic destructive lesion in the left mandibular body. The radiology report described portions of the lesion as ill-defined and documented destruction involving the mandibular body, lingual cortical plate, and inferior mandibular cortex. A periosteal reaction was also reported. The lesion was in close relationship to the inferior alveolar canal and to developing permanent teeth and the roots of the first molar. These findings supported a locally aggressive intraosseous process.

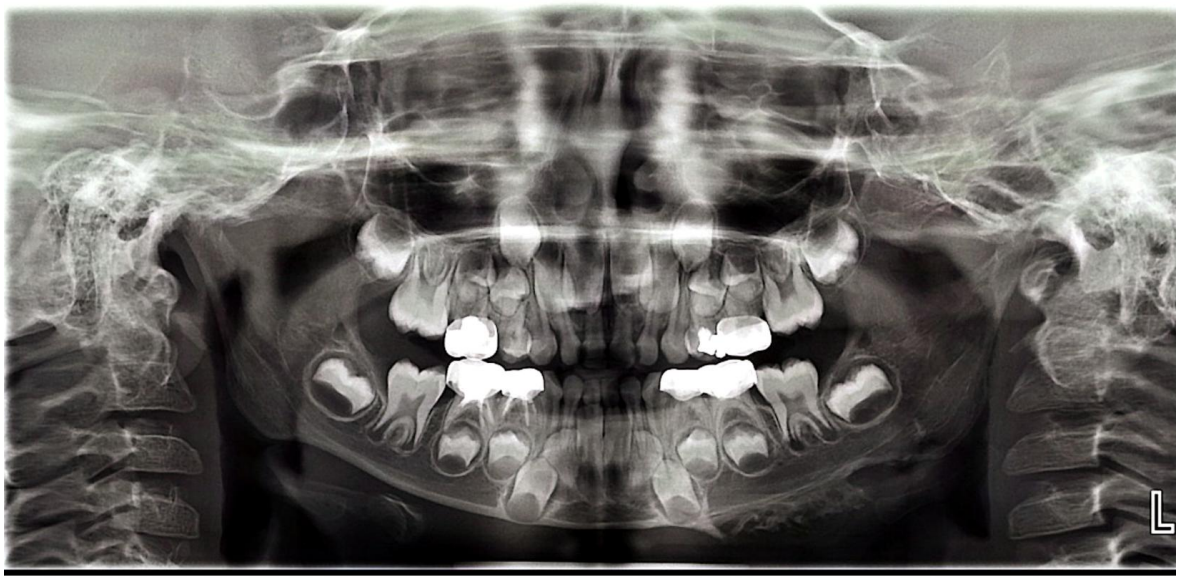


Fig. 1. Pre-treatment panoramic radiograph demonstrating the left mandibular lesion and the developing dentition.

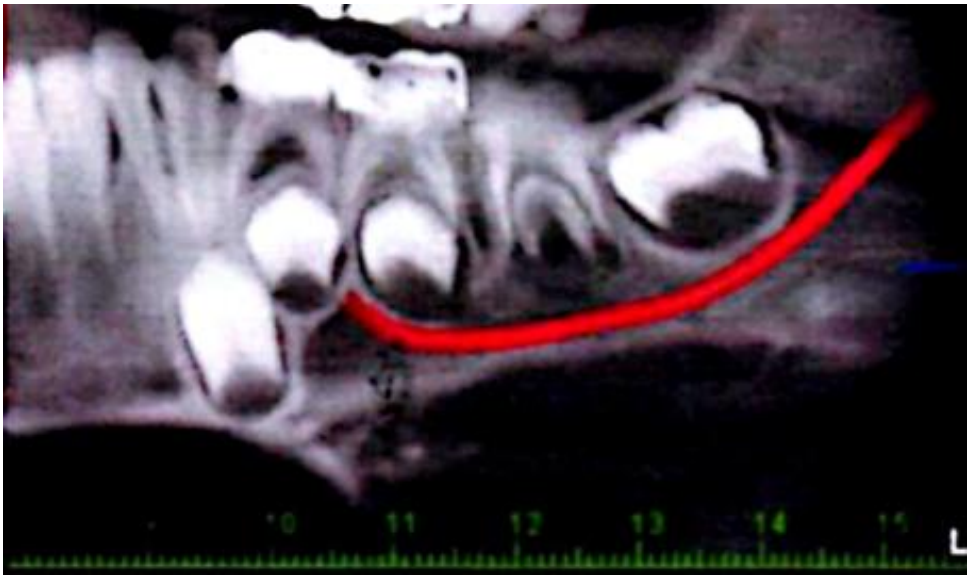


Fig. 2. Representative pre-treatment CBCT image showing the osteolytic lesion in the left mandibular body. The original radiology report described cortical destruction and close proximity to the inferior alveolar canal and developing permanent teeth.

Histopathology and immunohistochemistry

An incisional biopsy was reviewed as a second opinion with immunohistochemical assessment. Microscopically, the lesion consisted of bone trabeculae and intervening spaces infiltrated by a hypocellular neoplasm composed of fascicles of stellate-to-spindle cells with mild pleomorphism separated by collagen bundles. No necrosis was identified, and mitotic activity was reported as fewer than 1/10 high-power fields. The pathology report described an infiltrative spindle-cell tumor with approximately 1–2% proliferative activity and concluded that, in correlation with the radiologic findings, desmoplastic fibroma was the most eligible diagnosis.

Immunohistochemistry showed smooth muscle actin positivity in tumor cells, focal desmin positivity, negative H-caldesmon and S100, CD34 highlighting vascular endothelial cells rather than tumor cells, focal β-catenin positivity in tumor cells, and Ki-67 positivity in approximately 1% of tumor cells. These findings, together with the morphology and radiographic presentation, supported the diagnosis of desmoplastic fibroma.

Clinical Information: Left mandibular expansive lesion

Specimen: Received specimen for second opinion and IHC study consists of four paraffin blocks and four stained slides No: DP-19-12042 from Danesh, path lab, Tehran, accompained by a copy of corresponding pathology report.

MICROSCOPIC DESCRIPTION:
Sections show bone trabeculae and intervening spaces which is infiltrated by a hypocellular neoplasm composed of fascicles of stellate to spindle cells with mild pleomorphism, separated by bundles of collagen. No necrosis seen. Mitotic figures are less than 1/10HPF. /h2

DIAGNOSIS:
Left mandibular lesion, incisional biopsy (second opinion):
- Infiltrative spindle cell tumor with about 1-2% proliferative activity
- Necrosis: Absent
- Mitosis: Less than 1/10HPF

Note: Based on microscopic findings and correlation with radiologic report **"desmoplastic fibroma" appears the** most eligible diagnosis.

Fig. 3. Anonymized excerpt of the pathology report documenting a hypocellular infiltrative spindle-cell neoplasm, absence of necrosis, very low mitotic activity, and the radiopathologic impression favoring desmoplastic fibroma.

Treatment

The patient was initially evaluated for a left mandibular expansile lesion in November 2019. An incisional biopsy was subsequently performed, and the second-opinion histopathological and immunohistochemical evaluation dated March 10, 2020 (29 Esfand 1398) supported the diagnosis of desmoplastic fibroma. Definitive surgical treatment was performed in May 2020 (29 Ordibehesht 1399) through a submandibular extraoral approach. Segmental resection of the involved mandible was performed with a 1-cm surgical margin, followed by reconstruction and fixation with a mandibular reconstruction plate

Follow-up

Postoperative radiographic follow-up demonstrated progressive mandibular remodeling and preservation of the reconstruction plate without evidence of recurrent disease. A panoramic radiograph and CBCT obtained on September 16, 2021 (25 Shahrivar 1400) showed satisfactory postoperative changes and progressive bony regeneration. Further panoramic imaging on June 6, 2023 (16 Khordad 1402) demonstrated continued mandibular remodeling without an obvious recurrent lesion. A subsequent CBCT examination dated December 30, 2023 (9 Dey 1402) showed continued postoperative stability, with no definite radiographic evidence of tumor recurrence. Thus, the patient remained clinically and radiographically free of recurrence for more than 3 years after definitive surgery



Fig. 4. Later panoramic follow-up demonstrating continued mandibular remodeling and no definite radiographic evidence of recurrent tumor. The supplied record is dated 1402/03/16.

Discussion

DF is a rare intraosseous fibroblastic neoplasm with a predilection for the mandible in patients younger than 30 years. The lesion may be particularly challenging in children because the differential diagnosis includes several expansile odontogenic and fibro-osseous lesions, while the extent of surgery must be balanced against the effects of treatment on mandibular growth, developing dentition, facial symmetry, and neurosensory function. [1–5]

The present case is notable for several features. First, the patient was only 6 years old at diagnosis, placing him well below the typical age reported for mandibular DF. Second, the imaging showed destructive behavior, including cortical involvement and proximity to the inferior alveolar canal and developing teeth. Third, the histopathologic findings were characteristic of a low-cellularity spindle-cell lesion with abundant collagen, very low mitotic activity, and no necrosis. The pathology report appropriately emphasized correlation with the radiologic findings because spindle-cell lesions of the jaw can show substantial microscopic overlap. [2,5]

β-catenin is a useful adjunct in the differential diagnosis of fibroblastic lesions, although its expression is not uniformly strong across all DF cases. Published series have emphasized the utility and limitations of immunohistochemistry, including smooth muscle actin, S100, β-catenin and Ki-67, in separating DF from other spindle-cell tumors. [2,5]

The optimal extent of surgery remains debated. Historical and contemporary reports generally favor complete removal with clear margins when technically feasible because incomplete excision is associated with recurrence. For mandibular lesions requiring extraoral access, a submandibular/Risdon approach is a documented surgical route for segmental or block resection. Published cases have used approximately 1 cm of uninvolved bone as a safety/free margin, although no universally validated centimeter threshold has been established. [1,3,4,7,8]

Regarding the surgical margin, the literature does not provide a single universally validated centimeter threshold for mandibular DF. Nevertheless, 1 cm of apparently uninvolved bone has been used in published mandibular cases. In the present case,1-cm surgical margin was documented in the operative report, consistent with margins reported in the literature. [3,4,7]

The most important strength of this report is the relatively long radiographic follow-up in a young patient. The serial images demonstrate postoperative mandibular remodeling and continued development of the dentition without a definite recurrent mass. Long-term follow-up is particularly valuable because recurrence may occur after apparently successful initial treatment, and pediatric cases require attention to both tumor control and craniofacial development. [4,9,10]

Conclusion

Desmoplastic fibroma should be included in the differential diagnosis of an expansile or destructive intraosseous mandibular lesion in a child. In this patient, the diagnosis was supported by characteristic hypocellular spindle-cell histology, abundant collagen, very low mitotic activity, low Ki-67 proliferation, and supportive immunohistochemical findings in the context of the radiographic appearance. Surgical management should be individualized, with the goal of complete tumor control while minimizing functional and developmental morbidity. An exact 1-cm surgical margin was documented in the operative report. The absence of definite recurrence on serial imaging over more than 3 years supports the value of prolonged follow-up.

Patient perspective

A patient/parent perspective was not available in the supplied records. If available, a brief statement from the parent describing the treatment experience and current functional status should be added in accordance with the journal's reporting expectations.

Abbreviations

CBCT, cone-beam computed tomography; DF, desmoplastic fibroma; IAC, inferior alveolar canal; IHC, immunohistochemistry; HPF, high-power field; OPG, orthopantomography/panoramic radiography.

Declarations

Ethics approval and consent to participate

Written informed consent for the surgical treatment and publication of this case report, including the publication of the patient's clinical and radiographic images, was obtained from the patient's parents

Consent for publication

Written informed consent for the surgical treatment and publication of this case report, including the publication of the patient's clinical and radiographic images, was obtained from the patient's parents.

Availability of data and materials

All information supporting the conclusions of this case report is contained within the manuscript and the associated clinical records. Additional de-identified information may be made available from the corresponding author on reasonable request, subject to institutional and privacy requirements.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

FM contributed to the surgical treatment and postoperative follow-up.

AR contributed to the collection of patient data.

ZAE contributed to the collection of patient data.

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Figure list

Figure 1. Pre-treatment panoramic radiograph.

Figure 2. Representative pre-treatment CBCT.

Figure 3. Anonymized pathology-report excerpt.

Figure4. Later panoramic follow-up showing mandibular remodeling.