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Multifocal Desmoplastic Fibromas of the Mandible

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Desmoplastic fibroma is a locally aggressive benign fibrous tumor of bone that has been reported in both the maxillofacial skeleton and the long bones. Desmoplastic fibroma of the extremities was first reported by Jaffe[1] in 1958. The first description of this entity involving the jaws was reported by Griffith and Irby[2] in 1965. In rare instances, multifocal desmoplastic fibromas have been reported in the extremities and the pelvis.[3-6] We report a case of desmoplastic fibroma that occurred asynchronously in 2 separate locations in the mandible.

Report of Case

An 11-year-old girl, otherwise in excellent overall health, presented with a 1-year history of progressive enlargement of the left mandibular angle region. There was no family history of any maxillofacial disorders nor any family history of desmoplastic fibroma. The past medical history was significant for a marginal mandibulectomy at age 5 for a desmoplastic fibroma involving the right mandibular angle-ramus region (Figs 1, 2). At that time, a costochondral rib graft was used for primary reconstruction. Since that time, the patient had been asymptomatic until recently, when a similar bony expansion involving the left angle of the mandible was noted. There was no history of trauma, infection, or temporomandibular dysfunction.

Clinical examination revealed nontender, firm, buccal expansion of the left mandible. The overlying skin was normal; there was no associated cervical lymphadenopathy, and there was neither paresthesia nor anesthesia of the left inferior alveolar nerve distribution. A cutaneous nodule was also noted in the right preauricular region.

A panoramic radiograph (Fig 3) revealed a relatively circumscribed, radiolucent-radiopaque lesion of the left mandibular angle region. More anteriorly, there was a loculated mixed radiolucency-opacity associated with 3 impacted teeth.

An inferior marginal mandibulectomy with supraperiosteal dissection, was carried out from a left submandibular gland approach (Fig 4). A costochondral rib graft was secured with a bone plate to maintain the mandibular contour. The right preauricular nodule was removed by simple excision. The patient tolerated the procedures well and was discharged from the hospital 2 days later.

Microscopic examination of both the mandibular specimen and the right preauricular lesion revealed essentially identical findings: fibroblastic-like spindle cells proliferating within a delicate mesenchymal stroma, containing intertwining collagen fascicles (Figs 5, 6). Neither cellular pleomorphism nor mitotic figures were observed in any of the sections. The lesion was noted to be locally infiltrative and unencapsulated. The findings were interpreted as diagnostically compatible with desmoplastic fibroma.

Discussion

Desmoplastic fibroma is a rare tumor that represents the intraosseous counterpart of the soft tissue desmoid tumor or fibromatosis. It comprises 0.06% of all tumors of bone and 0.3% of all benign osseous tumors.[7] Although maxillary involvement has been described,[8-10] the mandible is the most commonly affected site in the maxillofacial skeleton. In the mandible, the lesions tend to occur more posteriorly, typically with involvement of the ramus-angle region. The tumor is most commonly noted during the second and third decades and usually presents as a painless, slow-growing, firm mass.

Radiographically, desmoplastic fibromas most often appear as multilocular radiolucencies; the differential diagnosis includes benign cystic processes, benign neoplasms, and sarcomas.[17]

At the microscopic level, desmoplastic fibroma consists of a mildly to moderately cellular matrix of fibrocollageous stromal tissue, lacking cellular pleomorphism, nuclear hyperchromatism, or mitoses.[2,11,12] Increased cellularity seems to be associated with an increased tendency for recurrence.[15,16] Both the lack of a capsule and the infiltrative nature of this lesion are hallmarks of desmoplastic fibroma; these features

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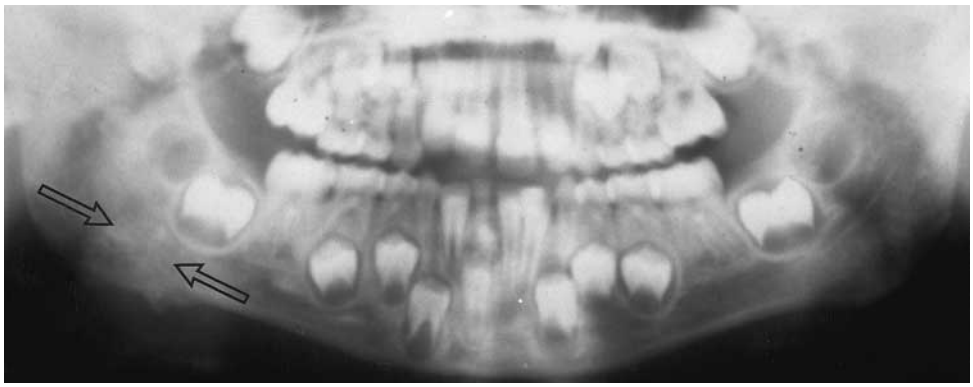

FIGURE 1. Original panoramic radiograph at age 5 reveals an ill-defined, mixed intraosseous lesion of the right mandibular angle area (*open arrows*).

can pose a diagnostic challenge. Absence of osteoid as well as prominent collagenization help distinguish this lesion from other fibrous lesions of bone, which include benign neoplasms such as nonossifying fibroma and odontogenic fibroma. It is critical to distinguish desmoplastic fibroma from low-grade or well-differentiated fibrosarcoma. The latter characteristically exhibits prominent cellular atypia and mitotic figures, and is more rapidly progressive in its clinical course.[13,14] Immunohistochemistry is generally not helpful in establishing the diagnosis of desmoplastic fibroma.

Surgical management ranging from simple curettage to segmental resection has been recommended for treatment of this tumor.[18-22] Segmental resection is preferred when a lesion displays signs of aggressive behavior and extension into surrounding soft tissues.

Significant rates of recurrence have been associated with desmoplastic fibroma, depending on the completeness of surgical removal. Lesions treated with resection or wide excision do not tend to recur. However, tumors treated with local excision or enucleation recur approximately 20% to 40% of the time, whereas those treated with curettage alone have a recurrence rate of up to 70%.[9,12,14-18]

There have been reports in the literature of multifocal desmoplastic fibromas involving the extremities. These cases typically involve a single anatomic region and suggest that the proliferative alteration of the involved connective tissue is a local phenomenon.[3] To our knowledge, this report represents the first case of multifocal involvement of desmoplastic fibroma in the maxillofacial region.

In this case, a nodule of desmoid tumor arose in the dermal soft tissue of the right preauricular skin overlying an area of previous mandibular surgery. This may have represented a new cutaneous desmoid tumor or, alternatively, it could have represented seeding or incomplete removal after the first mandibular surgical procedure. However, we believe the occurrence of a desmoplastic fibroma involving the left mandible 6 years after removal of the right mandibu-

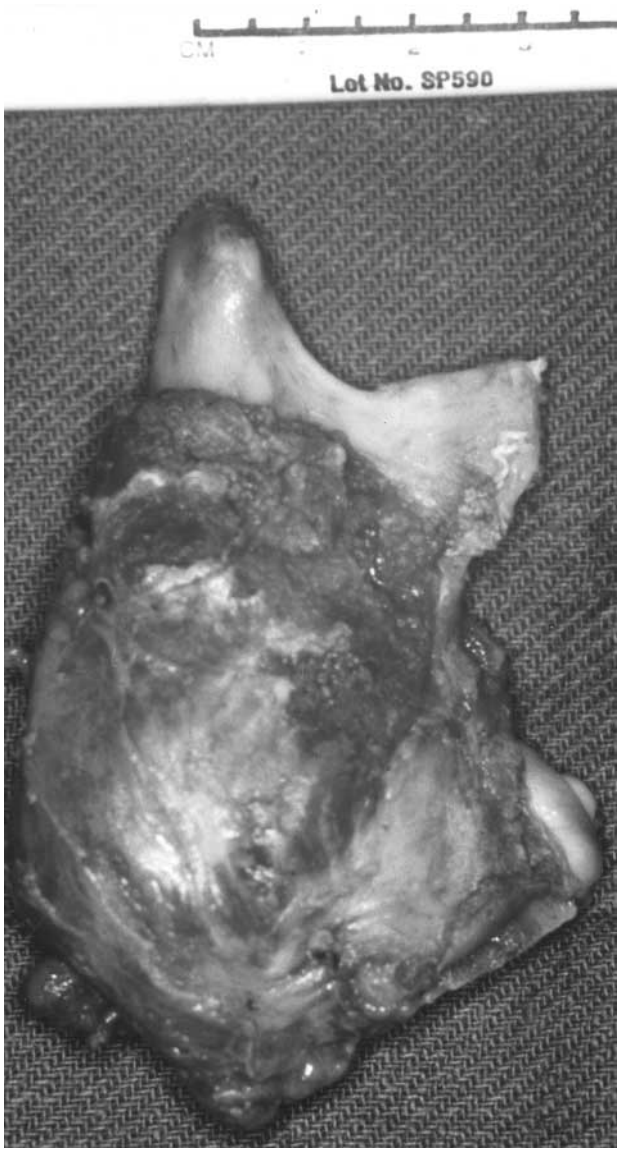

FIGURE 2. Gross view of initial surgical specimen from the right mandible.

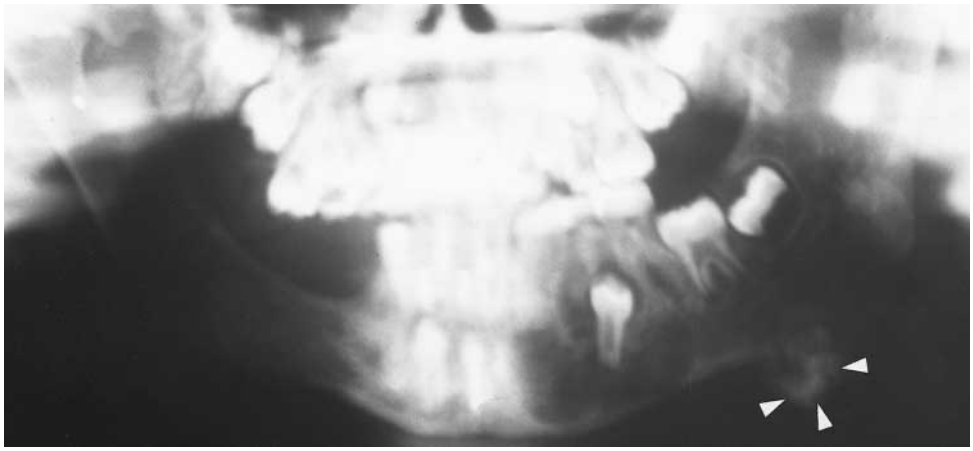

FIGURE 3. Panoramic radiograph revealing well-healed rib graft, right side and the new mixed radiolucent-radiopaque lesion of left angle of the mandible (*arrowheads*). The opacification represents residual or reactive bone. (Patient is 11 years of age.)

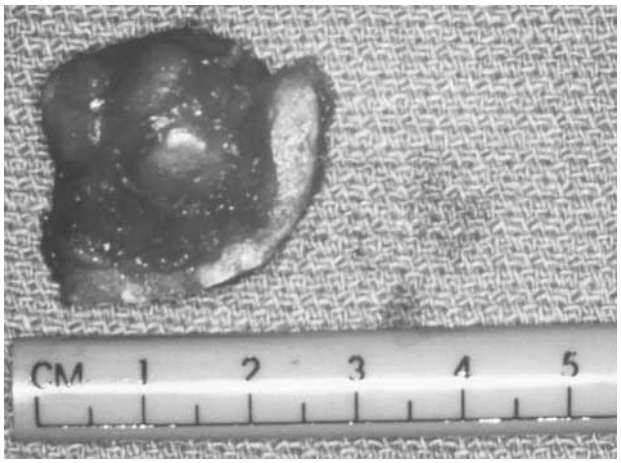

FIGURE 4. Gross specimen, left mandibular lesion.

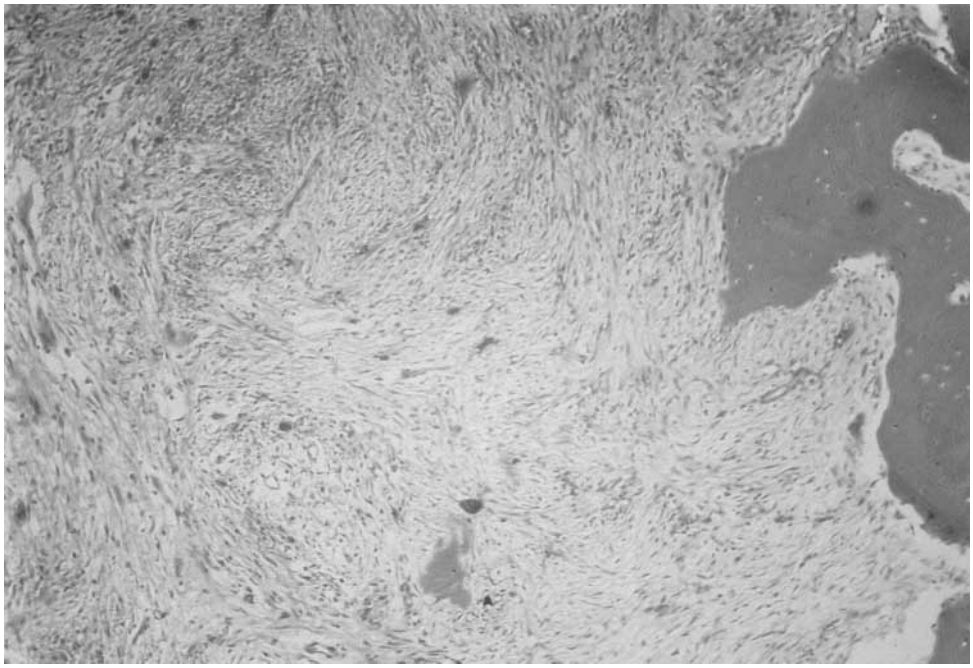

FIGURE 5. Low power (original magnification ×10) hematoxylin and eosin stained specimen showing diffuse proliferation of collagen bundles in a moderately cellular, loose stromal background. Note absence of a capsule and infiltrative nature of the lesion relative to the adjacent bone.

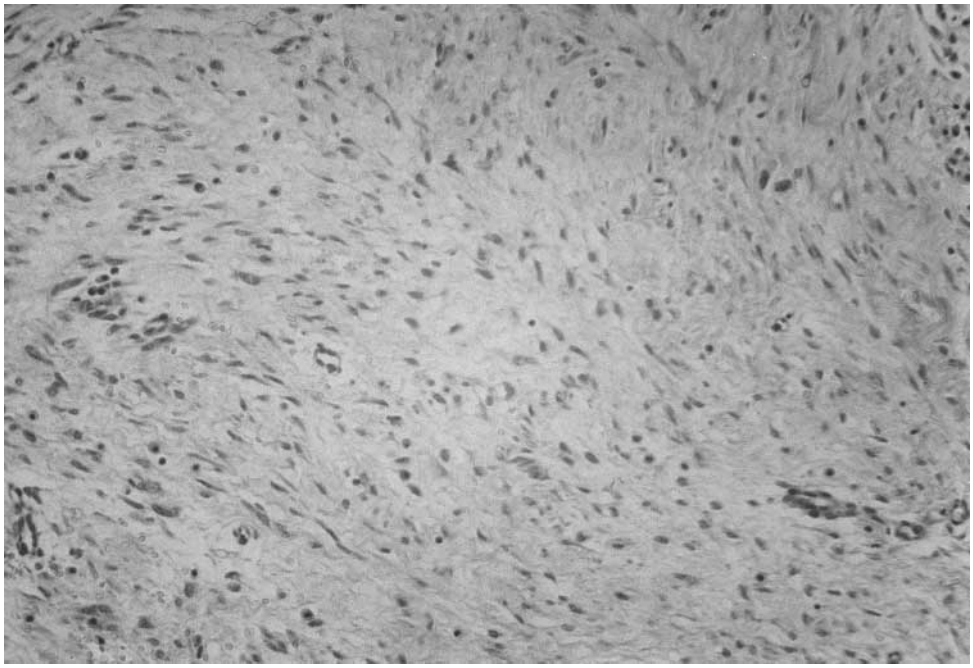

FIGURE 6. Hematoxylin and eosin stained (original magnification ×50) specimen shows sparse, innocuous-appearing fibroblasts with spindled to ovoid nuclei. Mitotic activity is absent. Scant inflammatory cells are noted.

lar lesion represents a second primary site. The presence of 2 distinct desmoplastic fibromas of bone and 1 desmoid tumor of the skin may represent a new syndrome. Because the phenomenon of multifocal desmoplastic fibromas has previously been reported in other skeletal sites, the possibility of a spontaneous mutation in all cases of multifocal disease should also be considered. In any case, because of the patient's apparent propensity to develop asynchronous and separate mandibular tumors, she will require long-term periodic follow-up examinations.

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