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Juvenile Aggressive Psammomatoid Ossifying Fibroma: An Interesting, Challenging, and Unusual Case Report and Review of the Literature

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Case Presentation

A Caucasian 12-year-old girl was referred to the Oral and Maxillofacial Department of University College London Hospital with a slowly enlarging right-sided mandibular swelling of 2 years' duration, with a previous diagnosis of aggressive trabecular variant of ossifying fibroma. Two previous attempts at excision had failed. Extraorally there was right-sided facial swelling. There was anesthesia along the whole distribution of the right inferior alveolar nerve. On intraoral examination, there were no teeth in the right mandibular quadrant. The mandibular swelling was not tender on palpation, bony-hard in consistency, covered by normal mucosa and extended from the ramus to the mental region, and had caused expansion of the lingual and buccal plates of the right mandible. No other bones were involved and no other abnormalities were found in a general physical examination. The medical history was noncontributory. There was no family history of skeletal disease.

IMAGING

An orthopantomogram showed a multilocular expansile lesion of the right side of the body of the mandible. The lesion had in part a ground glass appearance, with endosteal scalloping and a narrow transitional zone with adjacent normal bone (Fig 1A). Computed tomography (CT) scans confirmed a multilocular lesion present in the right mandible with heterogeneous attenuation (Figs 1B,C). It extended from just posterior to the third molar to the mental foramen.

Treatment Recommendation

Harry Papadopoulos, MD, DDS

Juvenile aggressive ossifying fibroma is an uncommon, benign neoplasm of bone that typically presents in children and adolescents. The World Health Organization describes it as a lesion affecting individual below the age of 15.[1] There are 2 histopathological variants of this lesion: a psammamatous and trabecular pattern. The psammamatous type mainly involves the bones of the orbit and paranasal sinuses whereas the trabecular type commonly involves the jaws, although there is controversy as to which jaw has a greater predilection, maxilla[2] or mandible.[3] These lesions exhibit a slight male predilection[2-4] and a more aggressive behavior compared to the more common ossifying fibroma seen in adults, although not always. El-Mofty[4] in an extensive literature review showed that not only was there a difference in site predilection between the 2 variants, but an age difference as well, with the psammamatous type occurring in an older and wider age range, compared to the trabecular type which occurs in children and adolescents.

Radiographically they present as well-circumscribed radiolucencies that in some cases contain central radiopacities.[5] Lesions that are more aggressive may show cortical thinning and perforation.[6] Being well-demarcated distinguishes it from other fibro-osseous lesions such as fibrous dysplasia, which may have similar histological features as well. Histologically, both variants tend to be unencapsulated, composed

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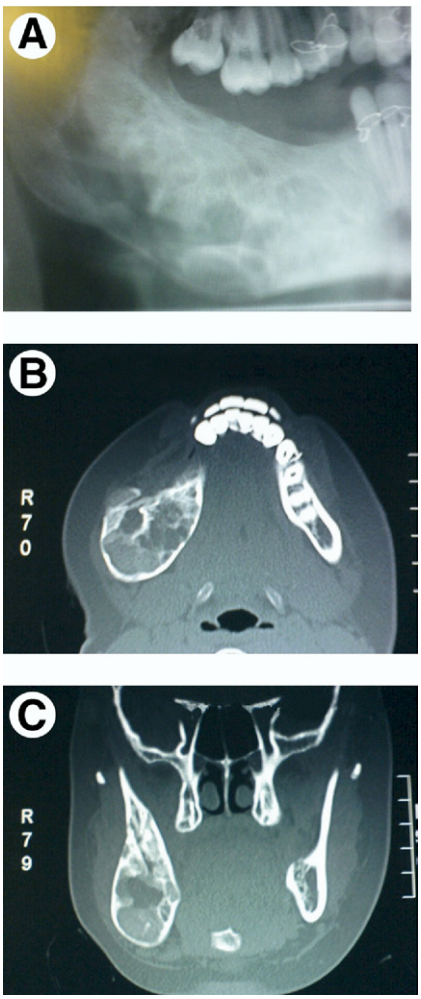

FIGURE 1. *A*, Orthopantomogram. *B*, Axial section of CT. *C*, Coronal section of CT, showing the multilocular swelling of the right body of the mandible.

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of a cellular fibrous connective tissue, and have small areas of giant cells. Both differ in their mineralized components with one showing trabeculae of woven bone (trabecular type) and the other showing concentric lamellated and spherical ossicles that vary in shape, and a myxoid component with aneurysmal bone cyst-like areas (psammamatous type).

Because of their slow, asymptomatic growth, they may be quite large on initial presentation; however, some lesions do demonstrate rapid enlargement, especially in younger children.[7] Complications occur when there is impingement on adjacent structures such as the orbits,[8] sinonasal cavities,[9] and rarely the cranial vault.[10] Unlike ossifying fibromas whose recurrence rates are very low, recurrence rates for juvenile ossifying fibroma vary, but have been reported as high as 30% to 56%.[5,11] Therefore it is not unusual for these lesions to require another operation for complete removal.[12-14] This in some cases may be due to incomplete excision of the tumor.[3,4,11,14] Smaller tumors may be treated by enucleation and curettage successfully. Although some authors advocate this type of conservative management for these tumors,[14,15] larger, more aggressive tumors will require resection with 5-mm margins, since infiltration by the tumor into adjacent bone is minimal.[16] Resection should be considered also in cases where there is recurrence, invasion of adjacent bony cavities, or where preserving the inferior border is not feasible, as in this case.[16,17] The procedure is undertaken via modified Risdon approach, and the resection accomplished with 5-mm margins. Taking age into consideration, attempting to preserve the periosteum is prudent, not only for its osteogenic potential, but it may also serve to prevent prolapse of soft tissues.[18] Attempting to preserve the inferior alveolar nerve may be difficult, but not contraindicated since this lesion does not invade nerve. The free segments of the mandible are then stabilized with a 2.7-mm reconstruction plate, after placing the remaining dentition into maxillomandibular fixation. There is no standardized follow-up protocol in the literature; however, because of the fairly high recurrence rate, immediate reconstruction is not advised. Secondary reconstruction may be undertaken sooner for slow-growing lesions (<1 year), and be delayed for fast-growing, aggressive lesions (>1 year). Although there has never been any reported case of malignant transformation,[19] rapidly growing lesions should raise suspicion for this, and add reason for delaying reconstruction.

Actual Treatment Rendered

A 3D, computer-aided design, computer-aided manufacturer (CAD CAM) model was constructed, using the preoperative CT. The mirror image of the left unaffected side of the mandible was transcribed onto the model, so an accurate symmetrical model could be used to aid reconstruction of the mandible. A titanium reconstruction plate was manipulated to fit the model perfectly and sterilized before surgery.

The surgical treatment was carried out in 2 stages, a week apart. At the first operation, the lesion was excised completely by a segmental resection of the jaw. The mandible was divided posteriorly with a vertical cut through the ramus passing through the sigmoid notch and anteriorly dividing the bone through the canine region. A neck dissection was carried out to access vessels for the following week's surgery, and to aid delivery of the tumor. Multiple cervical lymph nodes were dissected and sent to histopathology. The reconstruction plate was inserted.

In the second procedure, a right-sided deep circumflex iliac artery free flap was raised and inserted to

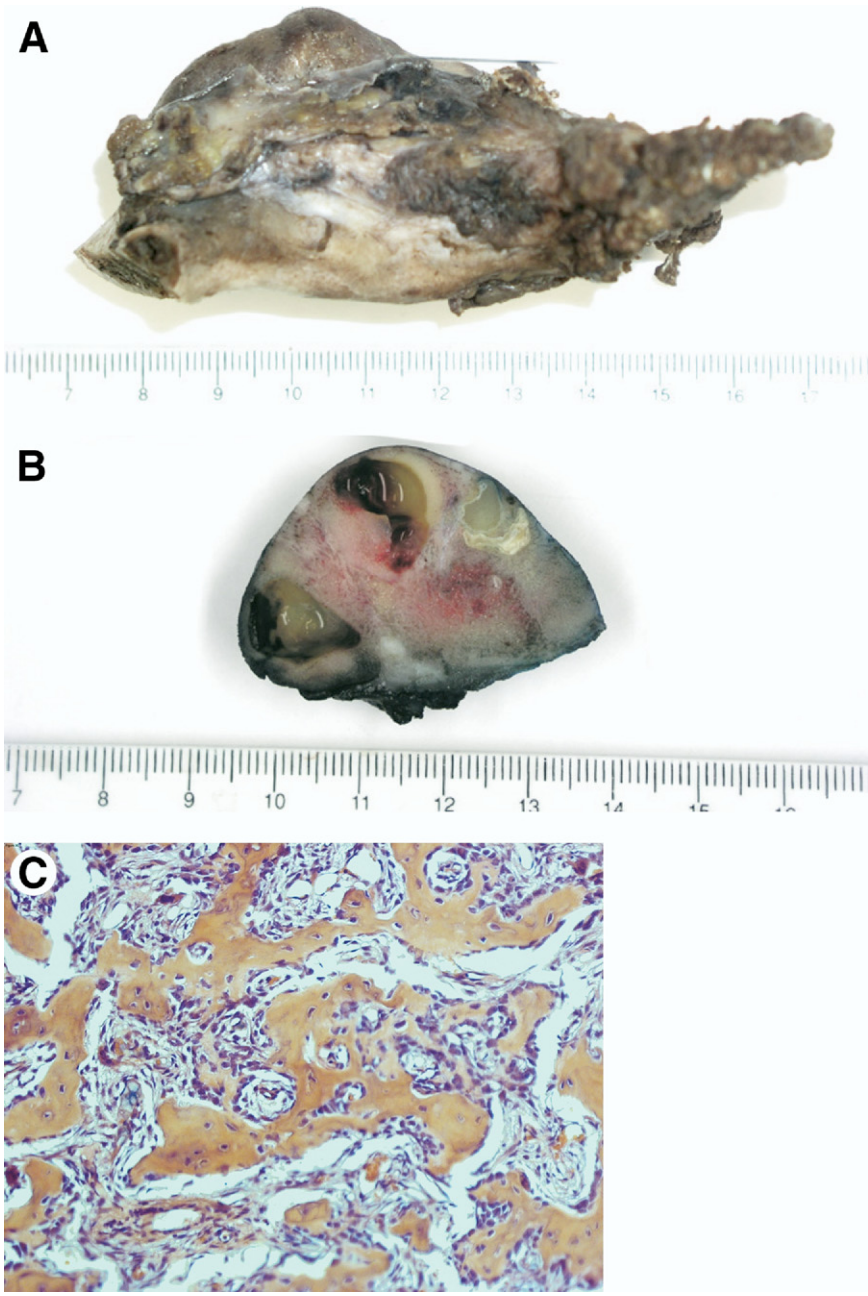

FIGURE 2. *A*, Gross appearance of the resected segment of the mandible and attached sulcular mucosa, seen from its buccal side, showing the expansion of the jaw. *B*, Stromal cysts in the sawn cut surface of the specimen. *C*, Histology of the ossicles in the psammomatoid component of the tumor. **(Figure 2 continued on next page.)**

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reconstitute the mandible. The patient recovered well from her surgery.

PATHOLOGY

Macroscopically, the tumor measured 68 × 40 × 38 mm (Fig 2A) and its cut surface displayed large cysts (Fig 2B) alternating with solid lesional bone and fibrous tissue. Microscopically 2 patterns were present. In the predominant psammomatoid component, approximately spherical ossicles with irregular seams of osteoid had been laid down in a highly cellular fibrous tissue. The second pattern was trabecular in type, characterized by trabeculae of woven bone lined by osteoblasts (Fig 2D). The lesional bone was separated incompletely from the surrounding normal bone by a cellular rim of fibrous tissue (Fig 2E). The tumor appeared to have been excised completely. There was no evidence of malignancy.

At 9 months postoperatively, 3 osseointegrated dental implants were inserted into the deep circum-

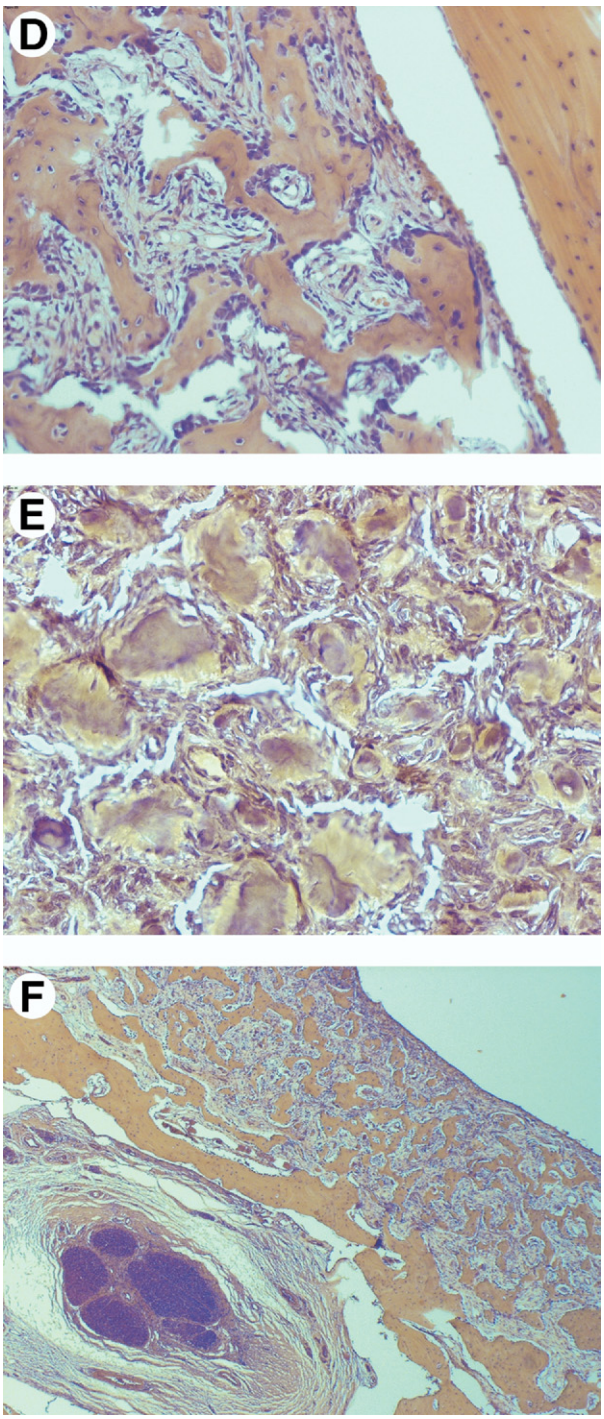

FIGURE 2 (cont'd). *D*, A rim of fibrous tissue separated the normal cortical outer layer of bone from the lesional bone. *E*, Lesional bone was incompletely separated from the surrounding normal bone by a cellular rim of fibrous tissue. *F*, Edge of a stromal cyst, psammomatoid lesional bone and the inferior alveolar nerve sacrificed in the resection. No perineural spread is apparent.

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flex iliac artery bone, on which a bridge was made, to replace the missing teeth (Fig 3). There has been no recurrence of the swelling 16 months postoperatively.

Discussion

Ossifying fibromas (OFs) are well-demarcated benign tumors of the craniofacial skeleton, most often in the jaws. The lesions feature fibrous tissue, osteoid and mineralized material comprising bone, and cementum-like deposits.[20,21] It is believed that these mesodermal jaw tumors arise from cells of odontogenic origin, probably from the periodontal ligament or alternatively from a primitive mesenchymal cell nest or from cells remaining after incomplete migration of the medial part of the nasal anlage.[21-23] A variety of chromosomal abnormalities have been described in the small number of OF tumors submitted to cytogenetic analysis.[24,25] Two subgroups, juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF) have been delineated on the basis of their histology.[4] They are uncommon, can grow rapidly, and may recur after attempted excision. The patients are usually but not invariably, children.

Ossifying fibromas are seen more often in blacks than whites,[23] occurring most commonly in the second to fourth decade. These lesions occur more often in males.[4] Ossifying fibromas arise most commonly in the mandible,[26-30] particularly the premolar-molar region,[31] followed by the maxilla.[26-30] They may occur in more unusual sites, however, such as the tibia,[32,33] ethmoidal,[34-38] maxillary and sphenoid sinuses,[34,39,40] petro-mastoid region,[41] temporal bone,[42] orbit,[43,44] and occasionally extraosseously.[45]

More than 50% of OF patients display jaw expansion that can lead to marked deformity of the face. The tumors had a mean diameter of 3.8 cm in 1 series.[31] They are usually painless, well circumscribed,[46] slow growing, and benign.[27] They can be destructive locally[43] and very aggressive.[41]

The histopathology of OFs is distinctive although there are significant variations. The lesions are well defined, often together with a surrounding outer border of cellular fibrous tissue, whereas the lesional bone of fibrous dysplasia (that usually features in the differential diagnosis) usually shows points of continuity with adjacent normal bone. The lesions of OF contain fibrous tissue that may be highly cellular or alternatively zones of almost acellular collagen, even in the same tumor.[4] The mineralized tissue comprises woven and lamellar bone, and acellular, approximately spherical, calcified deposits resembling cementum. Consequently these tumors previously were termed cemento-ossifying fibromas; however, cementum can only be distinguished reliably from bone when it covers tooth roots. The uncertain nature of this material in tumors at other sites in bones prompted the World Health Organization to rename them ossifying fibromas.[24]

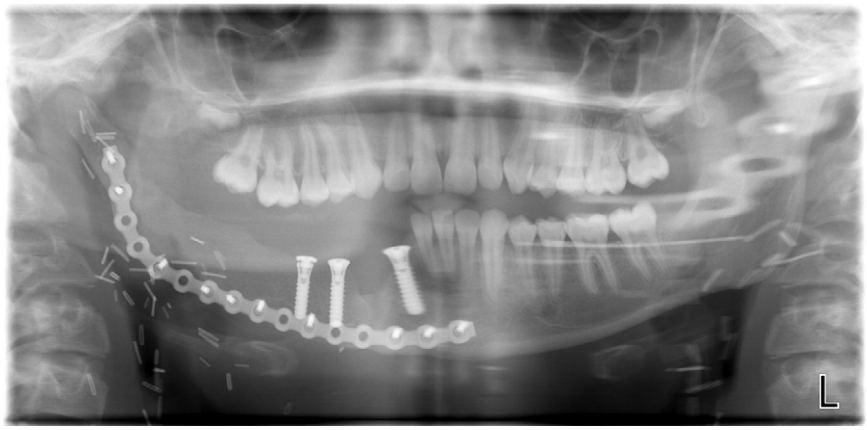

FIGURE 3. Postoperative orthopantomogram showing the position of the DCIA flap, the reconstruction plate, and dental implants.
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Ossifying fibromas typically feature stellate or spindle-shaped fibroblasts, with a widely variable degree and many patterns of cellularity and collagen formation.[21,43,46] A storiform pattern of cellularity in the stroma is present in more than 50% of cases. When giant cells are present, they are clustered in the center of the cellular stroma.[21]

The types of mineralized deposits within the tumor also vary widely.[46] Trabeculae of woven bone with osteoblastic rimming often are prominent and may form a reticular pattern.[21,46] The acellular or paucicellular calcifications resembling cementicles that also occur regularly[43,46] are minute at first but gradually fuse and ultimately form a dense mass. Ossifying fibromas usually present as a swelling but alternatively they may be diagnosed as a chance finding on a radiograph. A well-defined border is seen in 85% of cases,[31] with also a radiolucent rim in some tumors. Ossifying fibromas predominantly are radiolucent with various degrees of calcification, particularly near their center[46]; 53% of cases present purely as a radiolucency[31] and 86% of OF show no relationship with tooth apices or previous extraction sites. Tooth roots occasionally may be displaced or resorbed.[46]

The diagnosis of juvenile forms of OF is made on the distinctive histology. JPOF which is more frequent, is characterized by roughly spherical ossicles resembling the psammoma bodies of meningiomas set in cellular fibrous tissue. Unlike the smoothly contoured OF particles, the ossicles in JPOF have a thick irregular collagenous rim. JTOF is recognized by its trabeculae of woven bone with coarse lacunae, swollen osteocytes and a lining of plump osteoblasts. Bands of cellular osteoid are also found.[24] As in our case, stromal cysts of aneurysmal type and hemorrhage occur in JPOF and JTOF. Both JPOF and JTOF may occasionally lack a distinct fibrous rim.[4] Although initially categorized on the histology of the previous specimens as a trabecular form of juvenile OF, the recurrence in our patient removed in the resection specimen predominantly was psammomatoid in type. JPOF and JTOF seem to behave similarly, however, and their management is the same.

JPOF mainly arises in the bony walls of the paranasal air sinuses whereas the JTOF tends to affect the maxilla. Our case belongs to the minority of juvenile OF that occur in the mandible.[47] In contrast to OF, there is a slight male preponderance in both forms of juvenile OF.[4] The mean age also differs in that in JPOF, it is approximately 20 years, compared with an age range of 8.5 to 12 years in JTOF.[24] The term juvenile refers to this lower age range in both groups compared with the 35 years of age in OF, but patients as young as 3 months and as old as 72 years have been diagnosed with JPOF.[48] In addition, although the term aggressive is appropriate for most cases of juvenile OF on account of their recurrence rate ranging from 30% to 56%[4,24,48] and rapid growth, there are cases on record in which the lesions have only enlarged slowly.[48]

Although the progressive, painless swelling resulting in an obvious facial asymmetry in our patient is typical of OF, the involvement of the inferior alveolar nerve causing anesthesia is rare. There was, however, no histologic evidence of perineural spread (Fig 2F). Malignant change has not been recorded in OF or its juvenile subgroups.[47]

In contrast to its juvenile variants, recurrence of OF is considered rare after complete removal of the mass,[31,45] although it may follow after even a long interval.[22,27] Sudden expansion after trauma has been reported,[22] but the lesion simply may have not been detected until investigations for the injuries began.

Almost all cases of OF should be treated surgically.[49] Radiotherapy seems contraindicated because the tu-

mors are believed to be radio-resistant and because of the adverse effects of radiotherapy.[41,45] In view of the previous recurrences, a more radical operation to remove the juvenile OF in our patient was decided on, involving segmental resection of the jaw. More conservative excisions in the plane of the cellular fibrous outer rim that may possibly be part of the margin of the tumor, rather than merely a capsule of compressed surrounding stroma, may risk a recurrence. The mandible was reconstructed with a vascularized deep circumflex iliac artery bone graft, achieving a good cosmetic result and without any significant postoperative complications. The patient is left with persisting anesthesia in the distribution of the right mandibular nerve, however, as a consequence of the decision to sacrifice this structure in the jaw resection. Insertion of endosseous implants in the right body of the mandible replaced the missing teeth and restored normal masticatory function.

In conclusion, the juvenile forms of ossifying fibroma are uncommon but they need to be recognized and managed appropriately in view of their distinctive behavior. Although the lesion in the present case seems to have been excised completely, our patient will need to be followed up carefully, particularly as her tumor has proved to be aggressive, recurring after 2 previous attempts at excision.

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