JUVENILE OSSIFYING FIBROMA: REPORT OF A CASE

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SUMMARY : *Juvenile ossifying fibroma is an uncommon benign fibro-osseous lesion containing different amounts of calcified tissue scattered in a cellular fibroblastic stroma. Controversy exists in classifying these benign lesions, and neither the clinical presentations nor the histologic criteria alone are distinctive. A child with a rapidly growing mandibular mass, causing severe facial asymmetry, was seen. After a biopsy confirming the diagnosis as a benign fibro-osseous lesion, the entire mass was curetted and the cavity was reconstructed. Combining the clinical and pathologic data, our case resembled an aggressive juvenile ossifying fibroma. Recurrences are encountered in 30 to 50% of the cases, and patients must be under close follow-up for early detection, before facial structures are destroyed by these benign lesions.*

Key Words: *Benign Fibro-Osseous Lesions, Cementum, Cemento-Ossifying Fibroma, Juvenile Ossifying Fibroma, Aggressive.*

INTRODUCTION

Cemento-ossifying fibroma (COF) is a benign fibro-osseous lesion containing different amounts of calcified tissue, composed of bony trabeculae, cementum or both, scattered in a cellular fibroblastic stroma (1-7). These benign fibro-osseous lesions present problems in their diagnosis and classification. COF is most frequently seen in the mandible and maxilla. There is a 5:1 female to male ratio and the patients are generally in the third or fourth decades. Although a small number of patients with aggressive tumors and recurrences have been reported in the literature (1,5); usually the lesion grows slowly and curettage or enucleation is the treatment of choice (1-3). Histologic criteria are not helpful in distinguishing these aggressive lesions from the more common slow growing lesions and the biologic behaviour of COF is somewhat difficult to predict at the onset.

The term active juvenile ossifying fibroma (JOF) is considered when the lesion behaves in an aggressive manner and the patient is under the age of 15 years Recurrence rates of up to 30% and 50% are encountered in this aggressive type of lesions (7-10).

We present a fast growing fibro-osseous lesion located in the mandible of a 6-year-old female patient, with its histopathological diagnosis as well as its preferred treatment.

CASE REPORT

A 6-year-old girl with a rapidly enlarging

mass, causing jaw asymmetry was presented to our clinic. She had no dental or medical problems until a hard mass began to enlarge on the left side of her jaw two months ago. On physical examination the left side of her face was considerably swollen and a hard mass of 4x4 cm was palpated on the premolar and molar region of her mandible A panoramic mandible X-ray film revealed a radiolucent and radiopaque appearing multilobulated mass expanding the cortex with 3 unerupted teeth inside it (Fig. 1). The computerised tomography also showed similar findings. An incisional biopsy, which was performed to decide on the definitive treatment, revealed a benign fibro-osseous lesion. In a second operation, a window was made in the mandibular cortex and the whole mass was curetted until the healthy bony margins were reached (Fig. 2a). The three teeth lying inside the mass were also extracted (Fig. 2b). The mandibular defect was filled with spongious bone chips (Fig. 2c).

Microscopically, the gray/white curettage material consisted of calcified tissue either in the form of cementum like spherules or bony trabeculae in a fibrous stroma. Cementum was seen as rounded psammoma-like masses with a prominent osteoblastic rimming, and bony trabeculae was recognized by irregular isolated nonlinear lamellar and woven bone areas. Stroma surrounding these calcified structures was

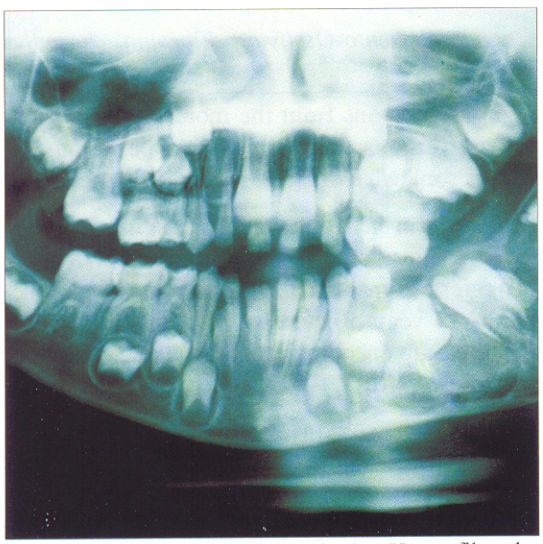

Fig. 1: On a panoramic mandibular X-ray film the lesion extends from the body of the mandible up to the subcondylar region, significantly expanding the cortex.

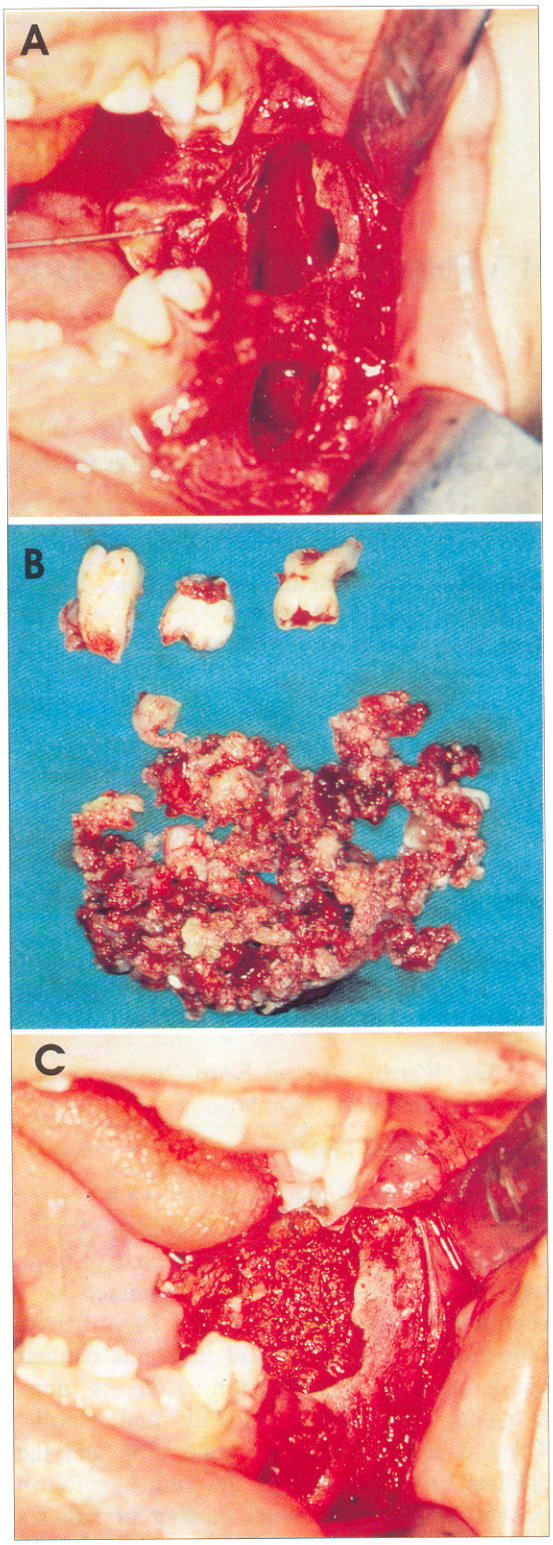

Fig. 2: Through an intraoral approach a cortical window is opened and the entire mass is curretted (A). The macroscopic appearance of the lesion with the three extracted teeth surrounded by the mass (B). The created mandibular cavity is reconstructed with spongious bone chips to achieve structural continuity (C).

bimorphic in appearance and highly hypercellular for the former and hypocellular for the latter (Fig. 3 a,b). No mitotic activity was observed in any of the areas. Immunohistochemically stromal cells showed strong reactivity for vimentin but not for actin (Fig. 3c). In the light of these histopathological findings, and taking the age and clinical presentation of the patient into consideration, the lesion was diagnosed as "juvenile aggressive ossifying fibroma".

The patient had an uneventful recovery period without a significant problem. At postoperative 2 months the cortical expansion and soft tissue edema had largely subsided and it was seen that the bone graft fitted the defect well (Fig. 4). Six months postoperatively the patient was under our close clinical follow-up, and there was still no sign of recurrence.

DISCUSSION

The term 'fibro-osseous lesion' refers to a diverse process in which normal bony architecture is replaced by fibrous tissue containing mineralised structures. It covers a broad spectrum of distinct lesions with different clinical presentations, microscopic appearances and growth potentials (7, 8, 10). Problems in classification, diagnosis and management exist and it has been difficult to distinguish cementifying fibroma from ossifying fibroma by any criteria. Until recently, the lesion has been called cementifying fibroma when spheroidal calcifications were seen and considered to be of odontogenic origin. When bone tissue predominated, the term ossifying fibroma was used to describe a tumor of bony origin. In 1992, these two conditions were classified as cemento-ossifying fibromas (COF) under a single designation, but this also did not clear the controversies in nomenclature (2,3,10). The concept is further complicated by the extracranial appearance of lesions showing histomorphologic similarities to COF (11), and some authors are reluctant to agree to the common decision that periodontal membrane was the origin of these tumors (6,12). It is also questioned whether cementum-like calcifications seen in the stroma of these tumors truly represent cementum, because similar bodies are also seen in extragnathic bony lesions. There are sporadic reports of various extragnathic bones affected by the disease, such as the ethmoid sinus, the

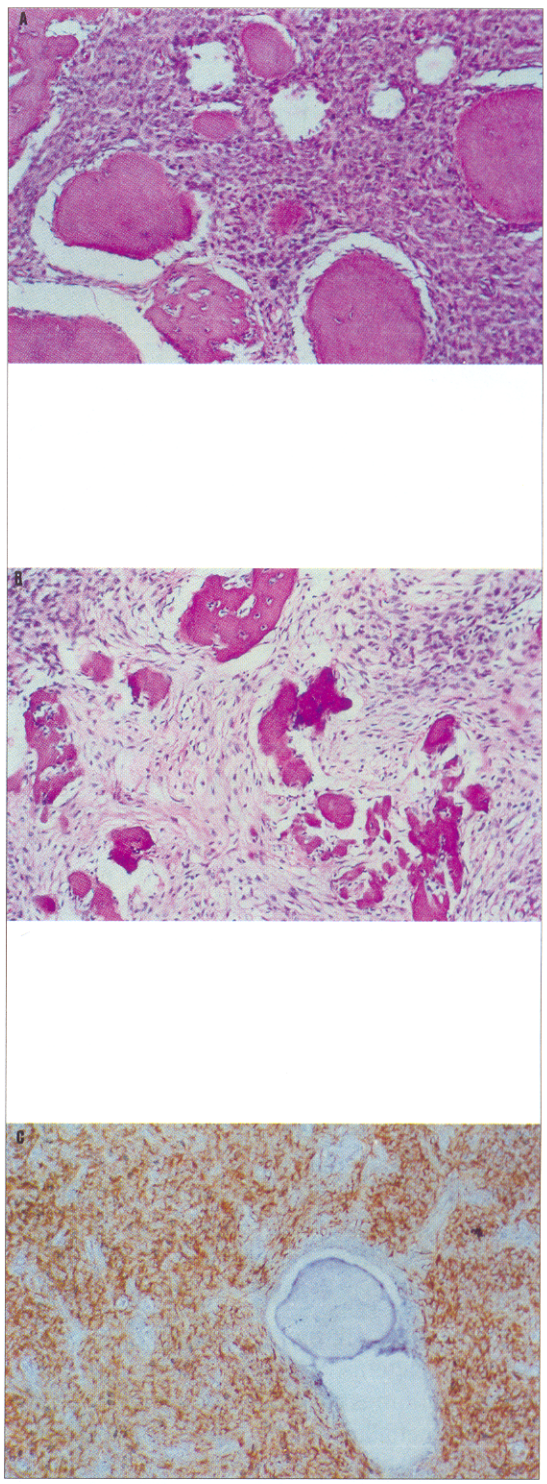

Fig. 3: Cementifying areas surrounded by hypercellular stroma and osteoblastic rimming (H&EX 200) (A). Bony trabecula without an osteoblastic rimming in a hyposellular stroma (H&E X 200) (B). Fibrocellular stromal cells showing strong vimentin positivity around a cementum like spherical calcified mass (Immunoperoxidase vimentin staining X 200) (C).

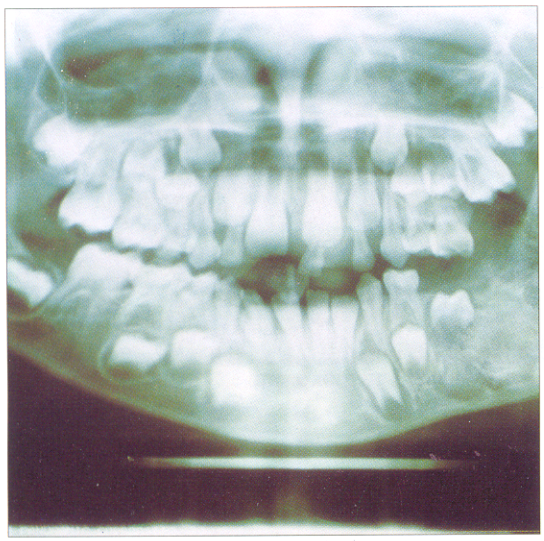

Fig. 4: Radiographic study shows good fit of the bone grafts.

sphenoid and the frontal bones (13-16).

Most benign fibro-osseous lesions of the jaws are asymptomatic or slowly progressing. On the other hand, when the lesion is encountered in a younger patient, especially below the age of 15, an unusual clinical presentation with apparent aggressive and destructive growth may be expected (5-10). Although no definitive predictor variables with regard to radiological features exist among the silent and aggressive types of these lesions, any such lesion necessitates a full radiographic work-up. Plain X-ray and tomographic films demonstrate whether the lesion is demarcated or not and give some information about the radiodensity of the lesion. Both cemento-ossifying fibromas and juvenile aggressive ossifying fibromas appear to be unilobulated or multilobulated radiolucent lesions in their early stages of development. At a later stage they are radiopaque and surrounded by a uniform radiolucent rimming (5-8, 10).

Ossifying fibromas possess different histologic patterns. The soft tissue component consists of fibrous connective tissue with varying degrees of cellularity and contains small amounts of collagen. The calcified component consists of either bony trabeculae or cementum-like bodies or even both and these histopathologic findings are generally shared by all fibro-osseous lesions (6). The histologic features ascribed to JOF are also not strict. The presence of a mineralised component composed of numerous spherical cementum-like bodies surrounded by osteoblastic rimming and a cellular stroma is characteristic (17,18). Hypocellularity of the stroma accompanying bony trabeculae without osteoblastic rimming suggests the maturation of the lesion, which correlates with the radiological findings.

It is not possible to differentiate these two by histopathological criteria alone, and without sufficient clinical data the pathologist can only report that a given biopsy is consistent with a benign fibro-osseous lesion (10). Although a young patient with a fast growing lesion indicates an aggressive lesion, this data is not sufficient for a definitive diagnosis. However, when good clinical and radiological information are combined with pathological findings, juvenile aggressive ossifying fibroma can be distinguished from its counterparts.

Before the definitive treatment is considered, we preferred to obtain a pathologic confirmation that the lesion was not malignant and performed an incisional biopsy. As noted in the literature, the lesion could also be recognised by fine needle aspiration (FNA) (19). However, firmer tumors may prevent the collection of enough material with FNA.

When the initial pathological assessment reveals a benign fibro-osseous lesion, the clinician must plan the definitive surgical excision and reconstruction. For a benign fibro-osseous lesion, whether it is aggressive or not, the appropriate treatment is either curettage or enucleation of the lesion until the healthy bony margins are reached (3, 5). Since benign fibro-osseous lesions are radioresistant and radiotherapy has malignant and transforming effects, this type of treatment is not considered as an option for these tumors (4, 5). Although recurrence rates of 30% to 50% are reported for JOF (9, 10), procedures such as hemimandibulectomies are not justified bearing in mind that these are benign tumors. It is not possible to predict the biological behaviour of the lesion either by clinical or histopathological assessments and if recurrences are encountered, instead of aggressive procedures, the clinician must choose local surgical excision (1, 8, 10). However, a 15-year-old patient reported by Pui-Chee Wu et al. (1), having an aggressive fibro-osseous lesion of the mandible, required partial

mandibulectomy after repeated conservative surgical procedures.

Considering the case presented here despite its non-specific pathological evaluation revealing cemento-ossifying fibroma, when this is combined with the clinical picture of a 6- year-old girl having a fast growing, aggressively destructive bony lesion of the mandible, juvenile aggressive ossifying fibroma presented as our diagnosis. Regardless of the subtype of the benign fibro-osseous tumor, it is wise to perform curettage and reconstruct the created defect. However, one should keep in mind that this girl may suffer a recurrence of the tumor, necessitating close periodical follow-up and decide on the future surgical procedures if required.

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