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Fibrous dysplasia and ossifying fibroma of the paranasal sinuses

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Abstract

Fibro-osseous lesions involving the paranasal sinuses, the mid-face and anterior skull base are uncommon. In addition, there appears to be no clear pathological or clinical classification that embraces the variety of lesions that exhibit such diverse pathological and clinical behaviour, yet may still be referred to as a fibro-osseous lesion. The diagnosis of fibrous dysplasia and ossifying fibroma is made on a combination of clinical, radiological and pathological criteria.

This paper emphasizes the clinical and pathological differences between fibrous dysplasia and ossifying fibroma. The more aggressive clinical behaviour of the latter is highlighted and a more radical surgical approach is recommended. In contradistinction, fibrous dysplasia can exhibit a more benign behaviour and radical surgery is not always justified.

A clinicopathological distinction between these two conditions is important from a management perspective despite the fact that they both may be encompassed under the 'umbrella' term fibro-osseous lesion.

Key words: Fibrous dysplasia of bone; Ossifying fibroma

Introduction

A fibro-osseous lesion is a generic term encompassing several conditions. Makek (1987) described a classification of these lesions which have a characteristic histological appearance. Mature bone is replaced by tissue composed of fibroblasts and collagen in a calcified matrix. The pathological classification of these lesions remains ill-defined and nomenclature, in particular, is determined to a large degree on the subjective interpretation of the pathologist concerned.

Montgomery (1927) first coined the term ossifying fibroma in describing three cases that were probably fibrous dysplasia. A description of a mandible tumour in a 35-year-old female consistent with ossifying fibroma had been reported earlier (Menzel, 1872). Fibrous dysplasia was described later (Lichtenstein and Jaffe, 1942). They recognized it as the same bony condition associated with McCune Albright syndrome (skin pigmentation, endocrine abnormalities and polyostotic dysplastic changes) (Albright *et al.*, 1937; McCune and Bruch, 1937). Oral pathologists have traditionally classified these lesions on the basis of whether they are odontogenic or non-odontogenic in origin. There are pathologists who think that there are no absolute differentiating features between ossifying fibroma and fibrous dysplasia, regarding ossifying fibroma merely as a variant of the monostotic form of fibrous dysplasia. The controversy has persisted for decades (Schlumberger, 1946).

Acknowledging present controversies, the following paper attempts to provide a clinicopathological distinction between ossifying fibroma and fibrous dysplasia supported by case reports.

Fibrous dysplasia

This idiopathic benign condition is a slow progressive disorder where normal bone is replaced by fibrous tissue and immature woven bone. Osteoblastic mesenchymal tissue undergoes abnormal development resulting in a condition which has a variable growth rate. It usually presents in the first two decades and the clinical features vary according to the site and potential complications. It is more common in females and the maxilla is most commonly affected. Involvement of the ethmoid and sphenoid sinuses is uncommon and thought to be due to the fact that these ossify in cartilage. In general, fibrous dysplasia occurs more readily in membranous bones.

Three clinical variants are recognized:

(1) The monostotic form accounts for 70–75 per cent of cases. It presents as painless swelling and craniofacial involvement occurs in approximately 30 per cent of cases.

(2) The polyostotic form accounts for 30 per cent of cases. These lesions occur at an earlier age and are of longer duration. Craniofacial involvement occurs in 50 per cent of cases.

(3) McCune Albright syndrome (polyostotic fibrous dysplasia, skin pigmentation and endocrine disturbances). There is a female predominance and skin lesions often occur ipsilaterally.

Painless swelling and facial deformity are the most common presenting symptoms in craniofacial fibrous dysplasia. Orbital involvement is common with proptosis, diplopia and loss of visual acuity resulting from 'tumour' encroachment on the optic canal or infective complications. Growth is variable and although cessation of growth

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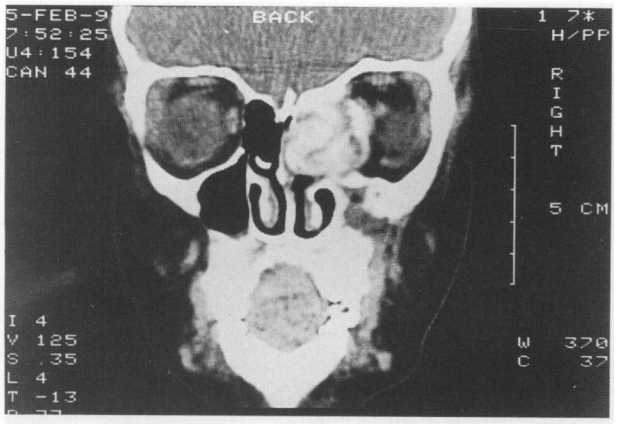

FIG. 1

Coronal CT scan of paranasal sinuses. Ethmoid lesion with mixed sclerotic and lucent bone densities – the pagetoid type of fibrous dysplasia.

usually occurs following puberty this is not universal (Chen and Fairholm, 1985). There is a 0.5 per cent risk of malignant transformation (Schwartz and Alpert, 1964).

Macroscopically, the tumour is grey-yellow with a thin bony cortex and gritty centre. Histologically normal bone is replaced by fibrous tissue with varying degrees of osseous metaplasia. Fibrous connective tissue is found with islands of spindle cells and foci of cartilage and metaplastic bone. Fibroblasts are less plump and the stroma less cellular when compared with ossifying fibroma. Occasional multinucleated giant cells are found in randomly arranged trabeculae of woven bone.

The radiological features are variable but occasionally characteristic. The plain X-ray features were well described by Fries (1957). The ground glass appearance is characteristic. Computed tomography (CT) scanning is a helpful adjunct for diagnostic and surgical planning purposes and is still the most commonly performed radiological procedure in fibrous dysplasia. It is useful for measuring growth rate in relatively asymptomatic patients. Magnetic resonance (MR) imaging, however, in addition to differentiating frontal bone fibrous dysplasia from meningiomas, will probably become the procedure of choice for routine follow-up of patients treated conservatively. Imminent involvement of the optic nerve can be detected and the true extent of the condition mapped preoperatively (Yano *et al.*, 1993).

Ossifying fibroma

Although found predominantly in the mandible (75 per cent), ossifying fibroma may arise within the paranasal sinus and skull base. Ossifying fibroma usually presents between the second and fourth decades. The male to female ratio is 1:5. They are characteristically monostotic and found predominantly in the craniofacial bones. Tumours within the mandible tend to behave in a more indolent manner compared with the paranasal sinuses. Ossifying fibroma may present as a painless cheek swelling. Involvement of the orbit or nasal cavity may be signified by proptosis, visual acuity loss, epiphora, nasal obstruction and epistaxis. Malignant transformation is extremely rare. Histologically, ossifying fibroma has a cellular stroma with plump fibroblasts and osteoblastic rimming is characteristic. Osteoclasts are found at the periphery. Lamellar bony trabeculae are found interposed with woven bony osteoid. There can be areas within the tumour which may be difficult to distinguish from fibrous dysplasia. The need for adequate biopsies is, therefore, emphasized by Harrison (1984). There are many subtypes described. Juvenile, aggressive, active, psammamatoid and cementifying ossifying fibroma are a few examples all of which are encompassed by the term ossifying fibroma. They do not have obvious predictive or prognostic value. Juvenile ossifying fibroma is not apt as these tumours often present in the fourth decade with gross bone destruction. Psammoma bodies may be seen but are not pathognomonic as they are found in other tumours such as meningioma and thyroid tumours.

Radiologically, a well-circumscribed mass with eggshell rimming may be seen with surrounding bone destruction giving a ballooned appearance. This varies depending upon the degree of ossification as in the case of fibrous dysplasia. MRI complements information obtained by CT by outlining the extent of tumour and skull base involvement particularly relating to dura.

Case reports

Case 1

A 16-year-old female presented with a three-month history of frontal headache and left-sided cheek swelling. Examination revealed 'polyps' within the left nasal cavity but there was no proptosis or loss of visual acuity. On CT there was a mass within the left ethmoid sinus and an opaque left frontal sinus. The combination of the clinical and radiological findings and histological examination of the polyps suggested a diagnosis of fibrous dysplasia.

A left external ethmoidectomy was performed and a frontal sinus pyocoele drained. Unfortunately, CT performed three months later revealed 'tumour' recurrence which extended to the cribriform plate. This was treated with local excision and diamond burring of the plate. Three years later the patient required a further left fronto-ethmoid exploration to treat recurrence of sinus symptoms. No evidence of fibrous dysplasia was found at surgery and the patient has remained disease-free since.

Case 2

A 12-year-old girl presented with a one-year history of diplopia, deteriorating visual acuity and right-sided cheek swelling. Examination revealed a proptosed right eye and the presence of unilateral nasal 'polyps'. CT revealed a mass within the right ethmoid sinus with extension to but not through the cribriform plate (Figure 1). Radiological and histological criteria of fibrous dysplasia were present. Excision of the lesion was performed by a lateral rhinotomy approach. The patient has remained well two years after the procedure with no radiological evidence of recurrence.

Case 3

A 27-year-old female presented with a four-month history of deteriorating vision, retro-orbital discomfort and proptosis. Examination revealed proptosis of the right eye and a suggestion of unilateral nasal polyps. CT revealed an extensive predominantly right ethmoid mass (Figure 2). Histological examination confirmed the radiological suspicion of a diagnosis of ossifying fibroma. As the tumour involved the skull base and extended across the midline as well as encroaching on the optic canal, a craniofacial approach was performed. The patient made an uneventful post-operative recovery and remains well and disease free two years later.

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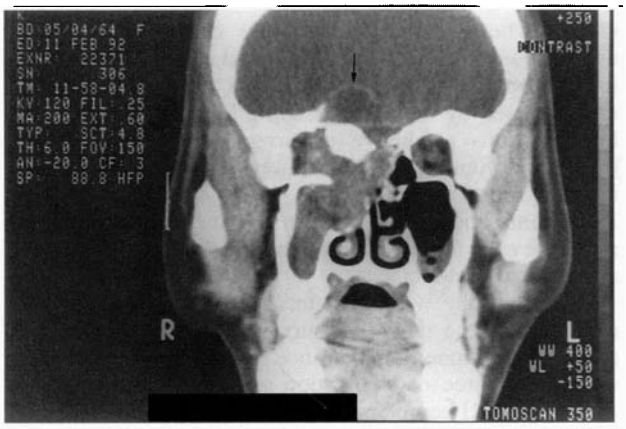

FIG. 2(a)

Coronal CT of paranasal sinuses. Characteristic eggshell rimming of lesion (arrow) extending well above the cribriform plate.

Case 4

A 34-year-old female presented with a nine-month history of right epiphora and proptosis, deteriorating vision, painful cheek swelling and nasal obstruction. A nasal mass medializing a concha bullosa-like middle turbinate and displacing the septum to the left was found on nasendoscopy. CT revealed a huge midfacial lesion which was well demarcated with an eggshell rim (Figure 3).

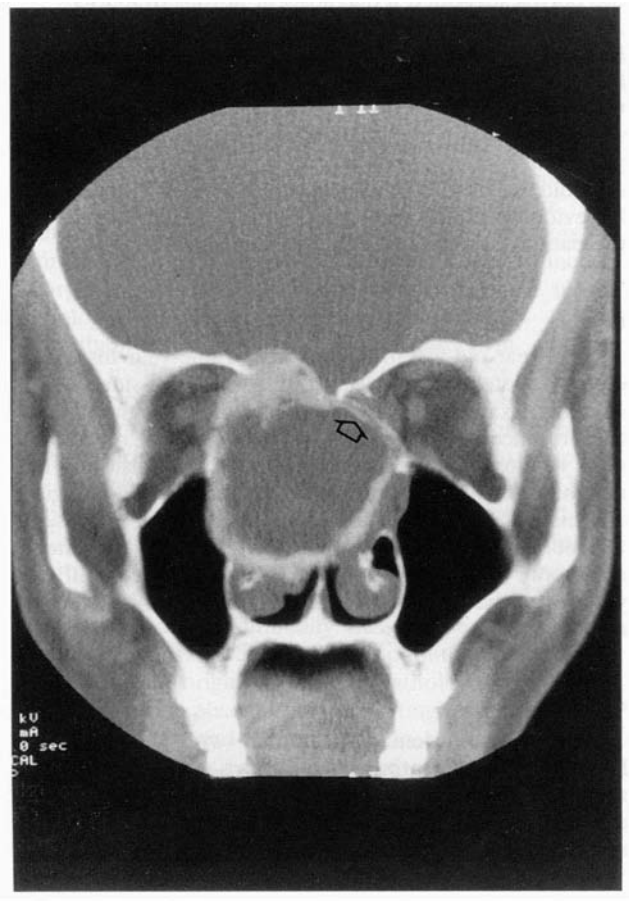

FIG. 3

Coronal CT. Massive midface ossifying fibroma. Osteoblastic activity at the periosteal surface along with tumour stimulated osteoclastic activity at the inner aspect of cortical bone gives rise to the characteristic radiological appearance.

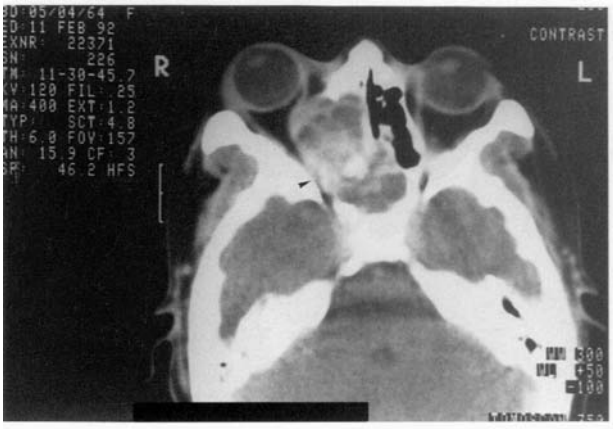

FIG. 2(b)

Axial CT. Note encroachment on optic canal (arrow).

A biopsy confirmed the radiological evidence for a diagnosis of an ossifying fibroma. The patient underwent a craniofacial resection with disarticulation of the nasal bones to facilitate complete excision of the tumour. The post-operative course was uneventful and the patient remains asymptomatic two years post-operatively.

Case 5

A 54-year-old male presented with an indolent history of progressive cheek swelling, proptosis and diminishing visual acuity in the left eye. Nasendoscopic features suggested the presence of polyps (Figure 4). An osteotome was required to obtain the biopsy however which

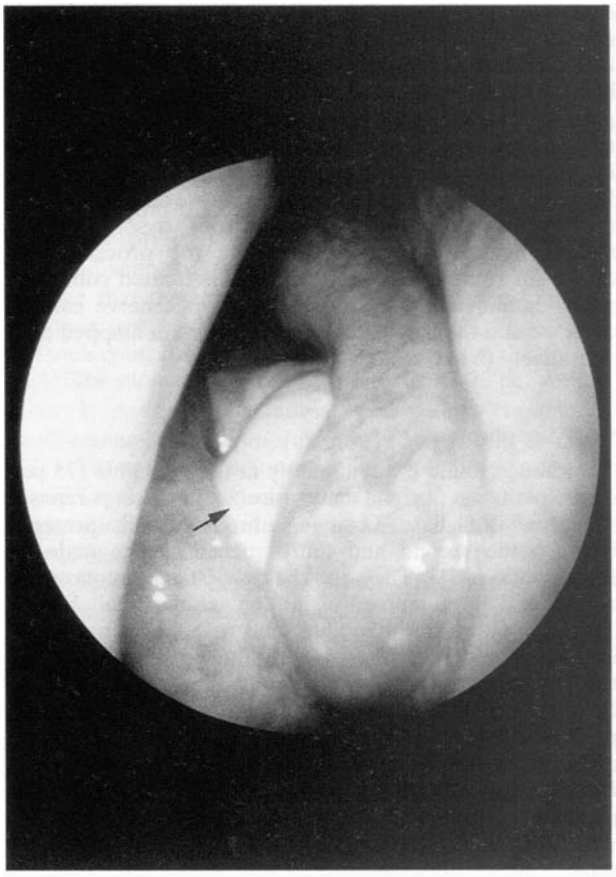

FIG. 4

Displaced middle turbinate by polyp-like lesion.

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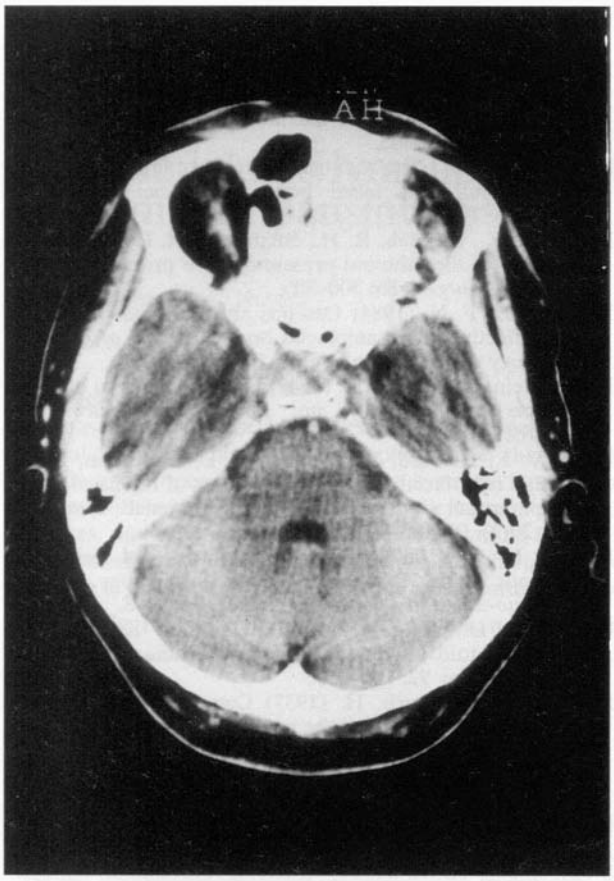

FIG. 5(a)
Axial CT showing extensive dysplasia with encroachment on the optic canal.

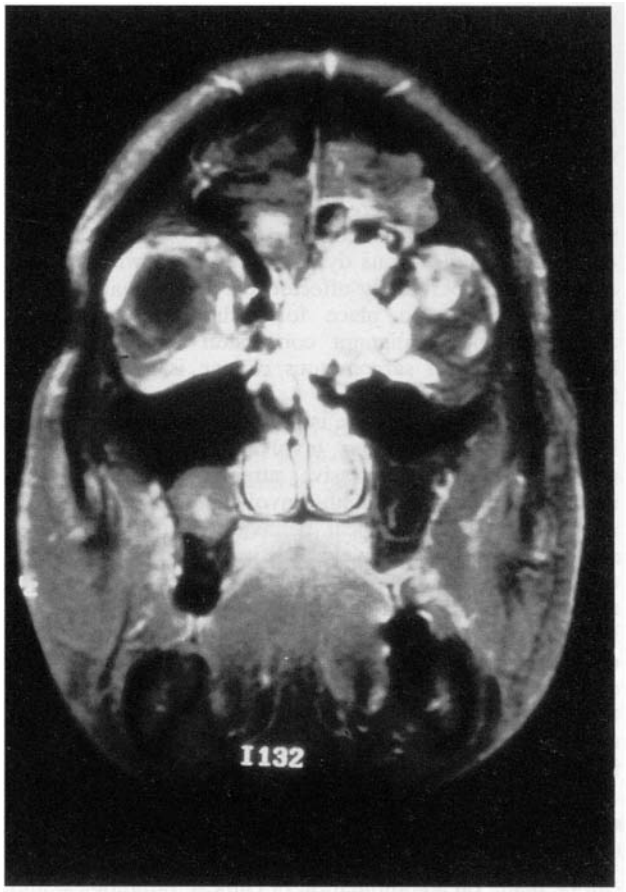

FIG. 5(b)
Contrast enhanced T1W MRI showing lateral displacement of left globe by the lesion.

confirmed the CT features of fibrous dysplasia (Figure 5). A conservative management approach was adopted as the patient had recently been diagnosed as having stage IV non-Hodgkin's lymphoma and there had been no deterioration in visual acuity over the preceding six months.

Discussion

At present many pathologists regard ossifying fibroma as a variant of fibrous dysplasia. This is further complicated by the finding that within a histological specimen of 'ossifying fibroma' there may be areas of tissue which have many of the features of fibrous dysplasia. An erroneous diagnosis may, therefore, be made and the necessity for adequate biopsies has been stressed by Harrison to enable the clinician to establish a diagnosis (1984).

Nomenclature will continue to confuse the issue until a specific cell marker is found which can accurately differentiate the two conditions on a histopathological basis. Until such time, clinicians, radiologists and pathologists remain dependent on one another to help establish the diagnosis in those cases where the features are equivocal.

Frazer *et al.* (1969) described three cases of fibrous dysplasia of the ethmoid sinus. There would be several arguments as noted by the authors then for naming these lesions ossifying fibroma. Radiologically, one case described had a well-circumscribed margin with eggshell rimming. The histological features were those described by Margo *et al.* (1985) as being consistent with ossifying fibroma.

Pathologists and radiologists are of the opinion that there are no specific biological or radiological features to predict behaviour in either fibrous dysplasia or ossifying fibroma (Smith and Zavaleta, 1952). The two cases of ossifying fibroma presented certainly behaved in an aggressive manner but were histologically dissimilar.

Management of fibrous dysplasia is controversial. Several authors advocate early surgery in all cases of cranio-facial fibrous dysplasia on the basis that optic nerve complications and surgical morbidity are thereby reduced (Munro and Chen, 1981; Chen and Fairholm 1985; Margo *et al.*, 1985; Edgerton *et al.*, 1986). Fronto-orbito-sphenoidal fibrous dysplasia causes visual impairment in 18 per cent of cases (Chen and Fairholm, 1985). The condition merges imperceptibly with surrounding normal bone and although complete resection is the treatment of choice, radical resections are not always warranted and should not create a deformity worse than expected from the condition.

Lesions confined to the ethmoid can be approached conservatively in prepubescent patients. Partially excised and curettaged lesions recur in 25 per cent of cases therefore warranting radiological follow-up. MRI will play an ever increasing role in this regard. A cranio-facial resection is advocated for recurrence of growth.

Massive midface and fronto-ethmoido-orbital fibrous dysplasia is usually managed by a craniofacial team. 3D CT scanning of the deformity facilitates the planning of resections and offers patients the option of more conservative contouring procedures with the possibility of recurrence when the lesions are extensive (Ricciardelli *et al.*, 1992). The volume and shape of the bone grafts can

be planned pre-operatively. Extensive autologous grafting requirements have been greatly diminished by the use of re-contoured dysplastic bone and methylmethacrylate implants. Re-contoured dysplastic bone tends to have a high take rate with no risk of re-growth due to disruption of the blood supply (Edgerton *et al.*, 1986). Some units advocate complete resection and primary reconstruction using split rib and iliac crest cortical grafts. Iatrogenic optic nerve loss is a potential complication in extensive fronto-ethmoido-orbital fibrous dysplasia resection where visual acuity is pre-operatively affected by optic canal involvement. There is no place for radiotherapy which is associated with malignant conversion up to 13 years later. The risk of sarcomatous change is increased 400 fold by radiotherapy.

In ossifying fibroma, site is all important with regard to management. Mandibular involvement can be treated conservatively but aggressive surgery is warranted for midface and paranasal sinus involvement because of its more aggressive biological behaviour. Both cases presented behaved aggressively. It is probably the site of the tumour that is more predictive of aggressiveness rather than the specific histological subtype as suggested by Margo *et al.* (1985). Management involves complete excision where possible in the paranasal sinuses. A craniofacial approach offers good access for tumours in the fronto-ethmoid region and facilitates complete excision (Lund *et al.*, 1998). The recurrence rate is high with incomplete resection.

Conclusion

The diagnosis of fibrous dysplasia and ossifying fibroma can be made on a combination of clinical, radiological and pathological criteria. Ossifying fibroma tends to exhibit a more aggressive biological behaviour justifying an early and more radical surgical approach. In contradiction, fibrous dysplasia can behave in a benign fashion when limited to the ethmoid sinus and a conservative surgical approach can be adopted with careful radiological follow-up. Aggressive fibrous dysplasia with gross deformity and importantly optic canal compromise warrant early surgical intervention by a team skilled in optic canal decompression.

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