Skeletal Radiol (1982) 8:141–143

Skeletal Radiology

Fibrous Dysplasia of the Facial Bones and Mandible

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Abstract. Five cases of fibrous dysplasia isolated to the facial bones and mandible are presented. A review of the clinical and roentgen features of facial fibrous dysplasia are described.

Key words: Fibrous dysplasia – Mandible – Maxilla – Facial bones

Since fibrous dysplasia (FD) was first described by Lichtenstein in 1938 [3], much has been written on the clinical, pathologic, and radiographic findings of fibrous dysplasia of the skeletal system. Nonetheless, the radiologic literature contains few examples of FD isolated to the facial bones and mandible. Involvement of the mandible is often diagnosed by oral surgeons with Panorex radiographs; thus radiologists may not be familiar with the mandibular changes of FD. FD of the facial bones, although much less common than cranial and long bone involvement, is more likely to be seen by radiologists for diagnosis and determination of extent of disease. Five cases of maxillary and mandibular fibrous dysplasia are presented below.

Case Reports

Case 1

A 55-year-old white woman presented with a five year history of progressively worsening right-sided nasal obstruction. Physical examination revealed septal deviation and a large polyp attached to the lateral wall of the right nasal cavity (Fig. 1).

Case 2

A 48-year-old black woman was aware of painless overgrowth of her right maxilla for 20 years. Physical examination revealed the right maxillary alveolus to be twice the normal size (Fig. 2).

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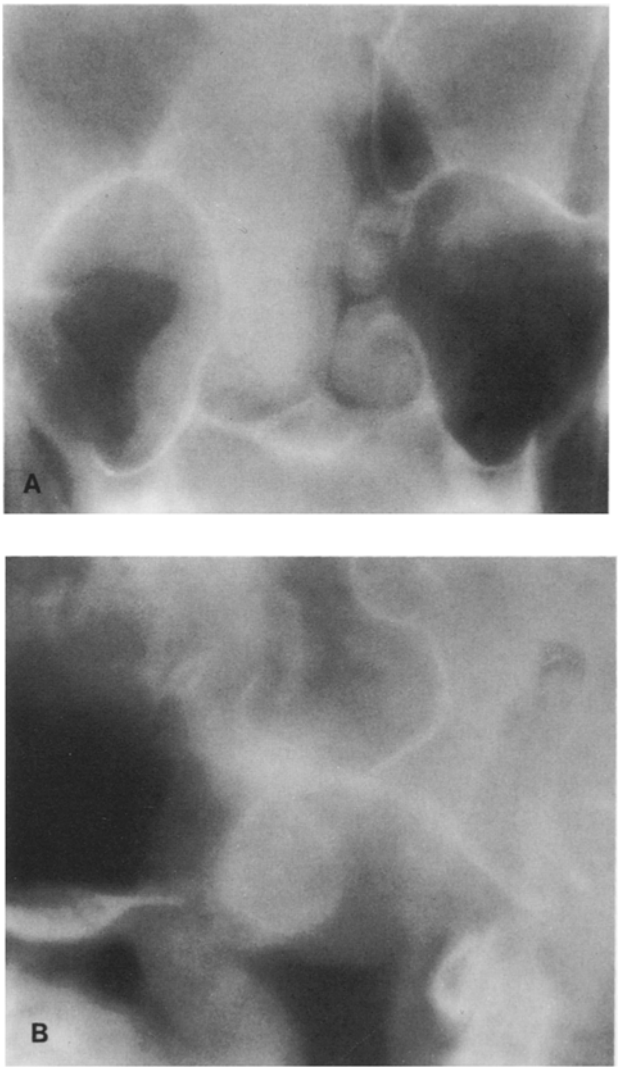

Fig. 1 A, B. *Case 1.* **A** Anteroposterior (AP) polytomogram of the nasopharynx demonstrates opacification of the right nasal fossa and ethmoid air cells by a homogenous expansile mass of osseous density. **B** Lateral polytomogram: Note that the nasopharyngeal mass is composed of a bony core with a fibrous tissue periphery

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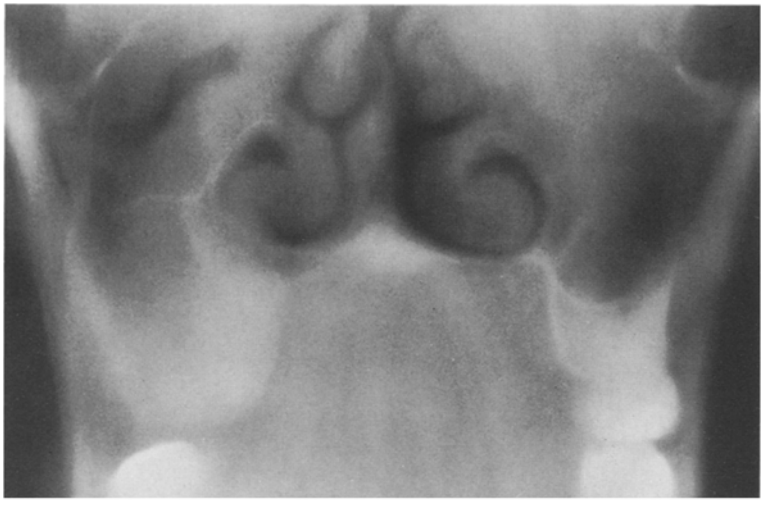

Fig. 2. *Case 2.* AP polytomogram of the maxilla shows expansion of the right alveolar ridge and inferior aspect of the right maxillary antrum by a homogenously sclerotic mass

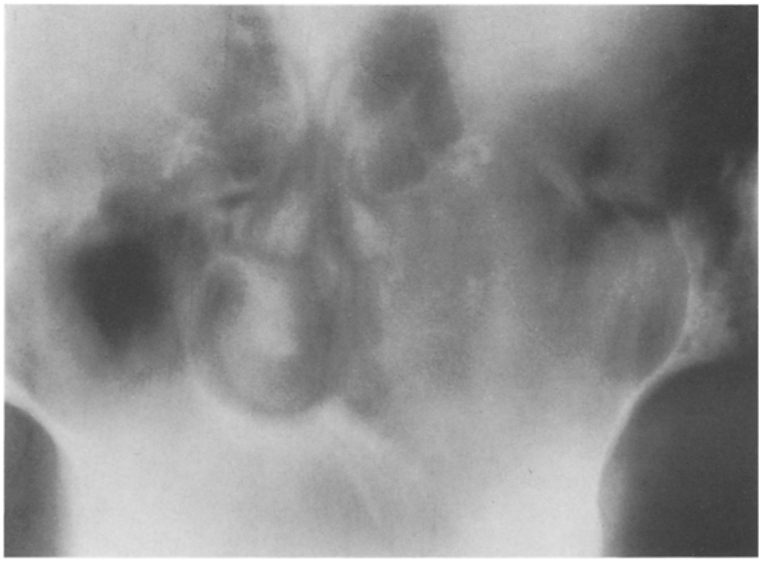

Fig. 3. *Case 3.* AP polytomogram of the maxilla reveals inhomogenous opacification of the left maxillary antrum with central speckled calcification. The inferior and medial walls of the antrum are poorly defined and involve the left nasal fossa

Case 3

A 21-year-old black woman noted progressively increasing swelling of the left nasolabial fold unassociated with pain or tenderness for six months. Physical examination revealed expansion of the left maxilla, extending into the left nasal cavity and depressing the left side of the hard palate. A 2–3 cm mass was palpated on the buccal surface at the nasolabial fold (Fig. 3).

Case 4

A 40-year-old black woman noted a localized swelling of the right mandible over a period of one year. A 5 mm tender mass was palpated in the right premolar region of the mandible on the lingual side (Fig. 4).

Case 5

A 12-year-old white boy complained of tenderness of the left anterior jaw over an eight month period. The pain was intermittent, elicited only with direct pressure, and did not interfere with mastication. There was slight facial asymmetry, the left side being slightly larger. A firm, tender, fixed, 1 cm nodule was palpated in the left anterior mandible. The overlying buccal mucosa was normal, without erythema (Fig. 5).

Discussion

Fibrous dysplasia is a disease of unknown pathogenesis in which the medullary cavity is replaced by fibrous tissue with the potential for new bone formation by osseous metaplasia. The proportion and distribution of fibrous and osseous tissue within the lesion determines its roentgen features. Its classic "ground glass" appearance results from homogenous, diffuse calcification of the fibrous elements.

Fries [2] divided fibrous dysplasia of the skull and facial bones into three radiographic patterns: sclerotic, cystic, and pagetoid. In the sclerotic form, there is a diffuse, uniform, homogenously dense area, often with a wide zone of transition between the normal and the abnormal bone. The lesion tends to follow the contour of the bone and expand it. This type is frequently found in the maxilla, as in Cases 1 and 2. The differences in radiographic appearance in these

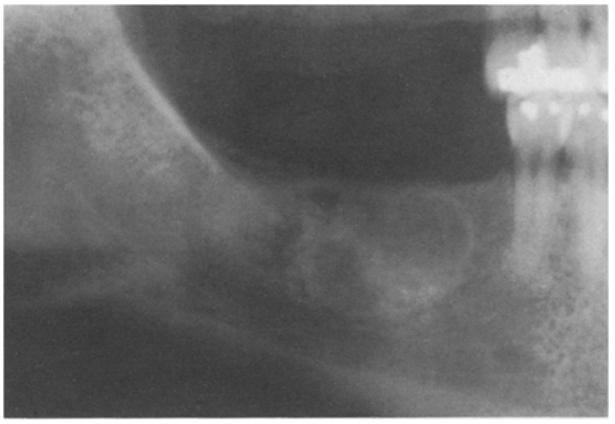

Fig. 4. *Case 4.* Panorex examination shows a 2×3 cm lucent, oval lesion in the body of the right mandible with sharp, sclerotic margins. Note the irregular central calcifications

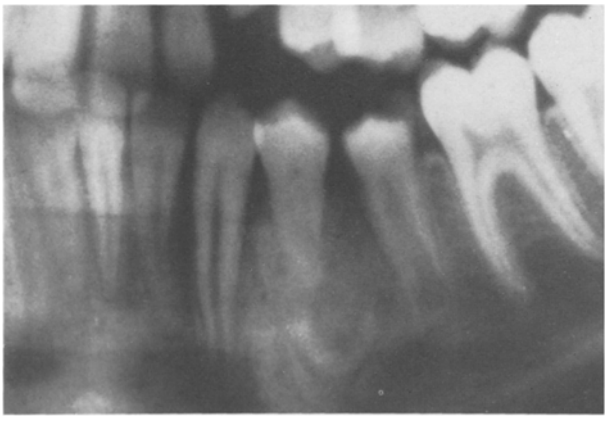

Fig. 5. *Case 5.* Panorex examination shows a "ground glass" lesion in the body of the left mandible with ill-defined margins. Note destruction of the premolar teeth roots and loss of the lamina dura adjacent to the lesion

two examples result from the much greater degree of osseous metaplasia in Case 1.

The cystic form of fibrous dysplasia appears as a radiolucent area with sharp and often sclerotic borders. It is frequently an oval, sometimes septated lesion causing cortical thinning and expansion. Small flecks of irregular calcification may be seen within it. The cystic form is common in the mandible, as in Cases 4 and 5.

In the pagetoid involvement, there is marked expansion of bone with alternating areas of radiolucency and radiodensity. Expansion tends to be outward. As its name implies, this form resembles Paget's disease radiographically. It is usually seen in older patients and in those with longstanding symptoms [2].

Periosteal reaction and soft tissue masses are not usually associated with fibrous dysplasia. Osseous erosions are uncommon and, when present, often cause a diagnostic problem because they resemble more aggressive lesions. The sutures in the facial bones usually do not restrict growth or expansion of the lesion [2]. Separation and malposition of the dental structures may result when lesions of fibrous dysplasia are in contact with the teeth. The lamina dura are destroyed in 5% of such cases. In children, teeth in the affected area may not erupt [1, 4].

Previously published studies of cranio-facial fibrous dysplasia differ in their assessment of the frequency of distribution of bony involvement. In Fries series of 39 cases, the lacrimal bone was the most frequently involved facial bone, followed by the maxillary and nasal bones [2]. However, the maxilla was the most commonly involved facial bone in a series reported by Zimmerman et al., while other authors report the mandible to be more frequently involved [7].

Clinically, these patients may present with painless swelling or asymmetry of the face or jaw, or with nasal obstruction. Other symptomatology includes loosening or malrotation of the teeth, proptosis, visual disturbances, and epistaxis. The disease is more common in females by a 3 to 1 ratio. It usually occurs in late childhood and early adolescence, but first appearance in adulthood is not uncommon. Serum calcium and phosphorous levels are normal, but occasionally serum alkaline phosphatase levels are slightly elevated [1].

Surgical excision is the treatment of choice. In children, delay in surgery is recommended, if possible, until after puberty when growth of the lesion generally slows or ceases. Radiotherapy is contraindicated due to increased risk of malignant transformation [5, 6]. The polyostotic form of fibrous dysplasia is more likely to undergo malignant change than the monostotic form, with or without radiation treatment.

Acknowledgements. We would like to thank Dr. Stanton Braid, Department of Oral Surgery, for allowing us to publish the Panorex radiographs of two of his patients. We also appreciate the assistance of Ms. Ann Prather in typing this manuscript.

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