Case 1355

Eurorad

Polyostotic craniofacial fibrous dysplasia in infancy

Published on 14.01.2002

DOI: 10.1594/EURORAD/CASE.1355
ISSN: 1563-4086
Section: Musculoskeletal system
Imaging Technique: CT
Imaging Technique: CT
Case Type: Clinical Cases
Authors: A. Loshkajian
Patient: 9 years, female

Clinical History:

Progressive craniofacial deformity and visual disturbance.
Imaging Findings:

The patient was admitted with headaches, craniofacial deformity and visual disturbance.
A CT scan was performed demonstrating pathological craniofacial bones, and bilateral optical canal compression.

Discussion:

Fibrous dysplasia (FD) of bone is one of the most frequently encountered anomalies of skeletal development. It may involve one or more bones. This idiopathic benign condition is a slowly 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. The ribs, femur, tibia and maxilla are most commonly involved. Craniofacial involvement is less frequent.
Three clinical variants are usually described:

- the monostotic form accounts for 70-75% of cases. It presents as painless swelling. Craniofacial involvement occurs in approximately 30% of cases.
- the polyostotic form accounts for 30% of cases. These lesions occur at an earlier age and are of longer duration. Craniofacial involvement occurs in 50% of cases.
- McCune-Albright syndrome, associating polyostotic fibrous dysplasia, skin pigmentation and endocrine disturbances; this form has a female predominance.

Painless swelling and facial deformity are the most common presenting symptoms in craniofacial FD. Orbital involvement is common with proptosis, diplopia and loss of visual acuity resulting from compression of the optic canal or the chiasma.
The radiological features of craniofacial FD are variable but occasionally characteristic. They can be divided into three patterns:

- the first is pagetoid with bone expansion and alternate areas of radiodensity and raduiolucency. It occurs in more than half the patients, most of whom are older than 30 years of age and have had symptoms for an

average of 15 years.
- the second pattern is sclerotic, with bone expansion and homogenous radiodensity (ground glass appearance).
- the third type is cyst-like, usually a round or oval lesion with a sclerotic border.

The sclerotic and cyst-like patterns seem to occur in younger patients with a history of symptoms of less than 3 years. The groung-glass appearance with an expanded cortex is the most common radiological form.
Computed tomography (CT) scanning is a helpful adjunct for diagnosis and surgical planning purposes and is still the most commonly performed radiological procedure in FD. CT is more useful than plain films in defining the nature and extent of the lesion. It shows a characteristic uniform amorphous texture of higher density than soft tissues. It is also useful for measuring growth rate in relatively asymptomatic patients.

MRI is also an accurate technique for the follow-up of these patients. It has become the procedure of choice for routine follow-up of patients treated conservatively.

The natural history of the disease has two phases; an active phase until puberty and a later quiescent phase.

The differential diagnosis in monostotic craniofacial fibrous dysplasia includes osteoma, ossifying fibroma, meningioma, osteoblastoma, osteosarcoma and chondrosarcoma. Polyostotic fibrous dysplasia must be differentiated from hyperparathyroidism, Paget's disease, neurofibromatosis and tuberous sclerosis.

The diagnosis of FD is made on a combination of clinical, radiological and pathological criteria.

Differential Diagnosis List: Polyostotic craniofacial fibrous dysplasia

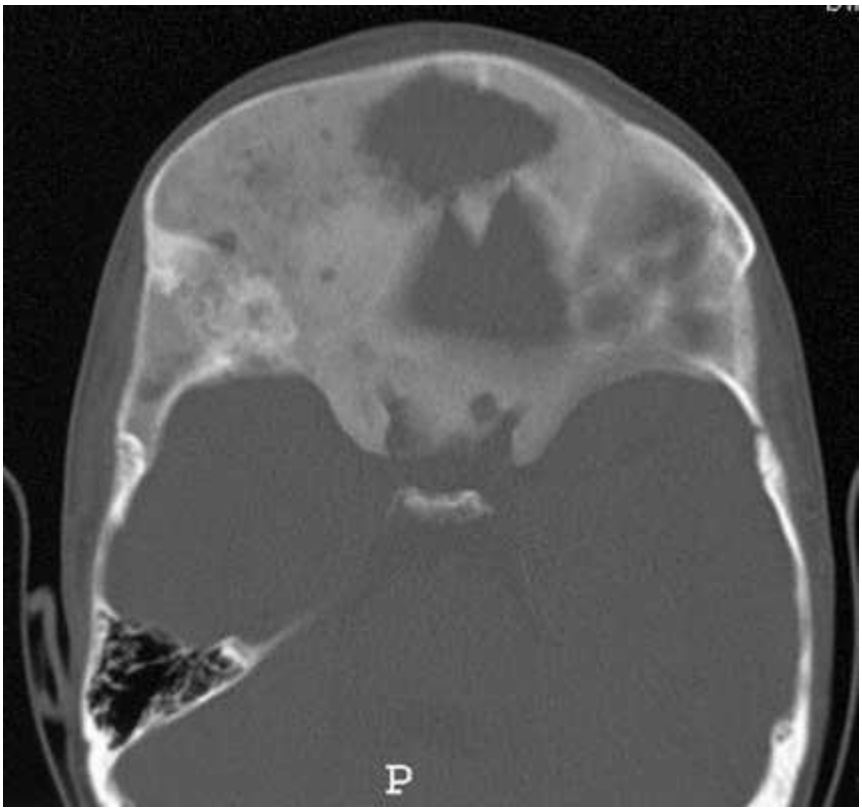
Final Diagnosis: Polyostotic craniofacial fibrous dysplasia

References:

Ricalde P, Horswell BB.
Craniofacial fibrous dysplasia of the fronto-orbital region: a case series and literature review.
J Oral Maxillofac Surg. 2001 Feb;59(2):157-67. (PMID: 11213984)
Papadopoulos MC, Casey AT, Powell M.
Craniofacial fibrous dysplasia complicated by acute, reversible visual loss: report of two cases.
Br J Neurosurg. 1998 Apr;12(2):159-61. (PMID: 11013671)
Camilleri AE.
Craniofacial fibrous dysplasia.
J Laryngol Otol. 1991 Aug;105(8):662-6. (PMID: 1919325)
Ricciardelli EJ, Borrow JA, Makielski KH.
Three-dimensional computed tomography in a case of craniofacial fibrous dysplasia.
Ann Otol Rhinol Laryngol. 1992 Mar;101(3):275-9. (PMID: 1543339)

Figure 1

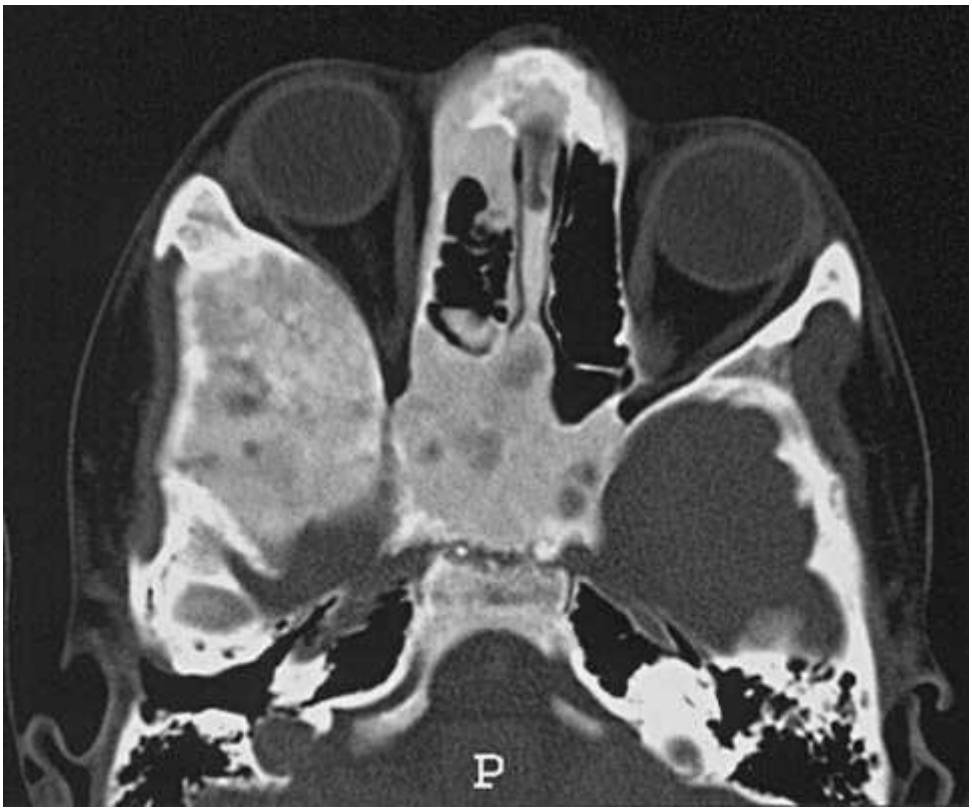
a



Description: Important extensive frontal bone involvement. **Origin:**

Figure 2

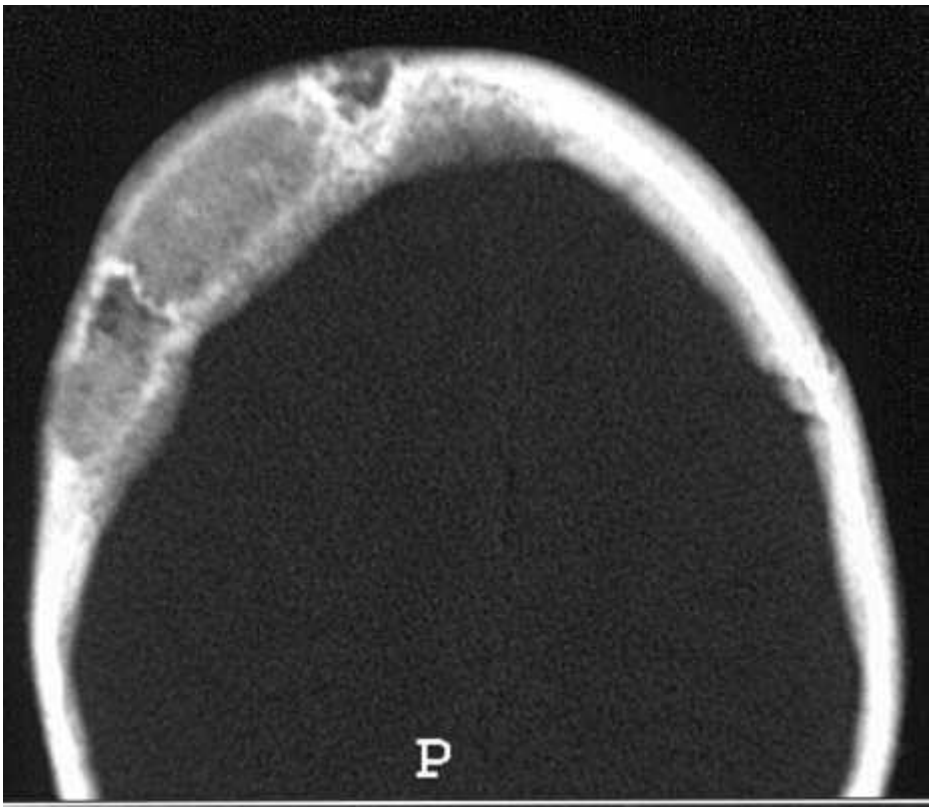
a



Description: Almost all the skull base bones are involved. The sphenoid bone is pathological. The optic canals are narrowed, explaining the optic nerve compression and the visual loss. **Origin:**

Figure 3

a



Description: This view, focused on the frontal bone demonstrates the characteristic "ground glass" appearance and cortical erosion. **Origin:**