Fibrous Dysplasia

20

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Abstract

Fibrous dysplasia is a benign deforming lesion involving one or several bones in which the structure of bone is replaced with altered osteofibrous tissue. It appears in a mono or polyostotic form. Treatment is based on biphosphonates and also by surgery when deformed bones occur.

Keywords

Fibrous dysplasia • Tumor-like bone lesions • Fibrous bone lesions • McCune–Albright syndrome

Fibrous dysplasia produced bone lesions with altered structure of the bone as a tissue and organ in which the bone is replaced with fibrocystic zones. The bone is transformed becoming wider and with thin corticals. The bone becomes deformed and fragile [1–3]. Fractures in the proximal femur and the Shepherd's crook deformity are characteristics (Fig. 20.1).

The etiology is unknown, but associated chromosomal abnormalities have been found, probably a mutation in the gen GS alpha in the human chromosome 20q13. It is not a inherited disease. The high production of FGF-23 can lead to hypophosphatemia.

It can compromise one bone, as in the monostotic form, or many bones, as in the poliostotic form. The skull, jaw, ribs, pelvis, femur and tibia are mainly involved. In the poliostotic form there is a trend of the lesions to be placed towards one half of the body. The monostotic form makes up about 80% of the cases.

Early puberty (mostly in women), skin spots, coffee and milk markings as in the coast of Maine pattern, and poliostotic fibrous dysplasia constitutes the Albright syndrome or McCune–Albright syndrome (20% of cases of fibrous dysplasia).

The combination of poliostotic fibrous dysplasia and soft-tissue intramuscular myxomas is named the Mazabraud syndrome.

The age of diagnosis is between the first and third decades of life. Although the genetic lesion is present at birth, most times it remains asymptomatic until adolescence.

Clinical Symptoms

For many years patients can be asymtomatic or have a casual finding. The progression of the lesions can frequently produce swelling and deformation of the segment, without any pain or few pain except when a pathological fracture is produced.

Deformations of the lower extremities are very notorious with bowing of the thigh and or leg produced by the plastic deformity of the femur and tibia which curve the bones induced for the body weight.

Craniofacial bone affections produce deformation of the skull cap or facial asymetry and neurological symptoms of the cranial nerves produced by obstruction of the cranial hollows in the base of the skull.

The progression of the lesions sometimes halts with the end of skeletal growth. That is an important consideration for the treatment.

Malignant transformation can occasionally occurs [4].

Rx Imaging

The radiological aspect is characteristic, with zones of cysts with different sizes rounded with areas of fibrous dense bone. Sometimes the image resembles "frosted glass". The bones frequently compromised are femur, tibia, ribs, maxilla and craniofacial bones (Figs. 20.2 and 20.3).

The cranial bones show very calcified zones, which explains the closing of the hollows of the cranial nerves in the skull base.

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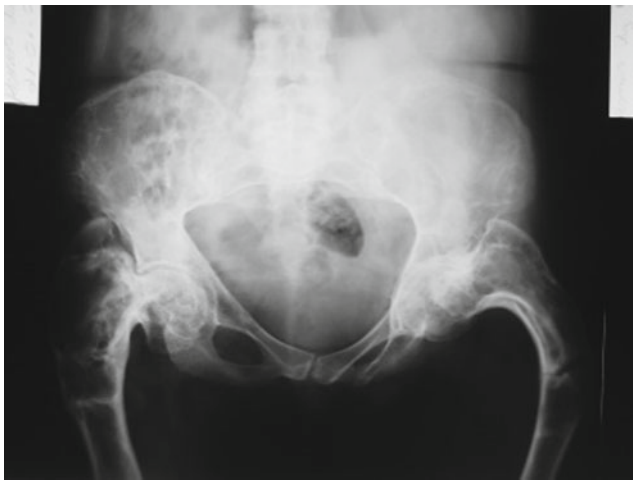

Fig. 20.1 Pelvis X-ray bilateral Shepherd's crook deformity in fibrous dysplasia

Scintigraphy (with Tc99)

It is very useful to detect the distribution of the affected bones showing an hypercaptation of the radioisotope. Also it can be useful to measure the diminution of the activity of the lesion.

Magnetic Resonance

The signal is low in T1 but high in T2 in the affected bones.

Histology

The normal bone is changed into new bone with deformed bone trabeculaes, fibrous tissue with fibroblastos, osteoid tissue and giant cells replacing the normal bone marrow.

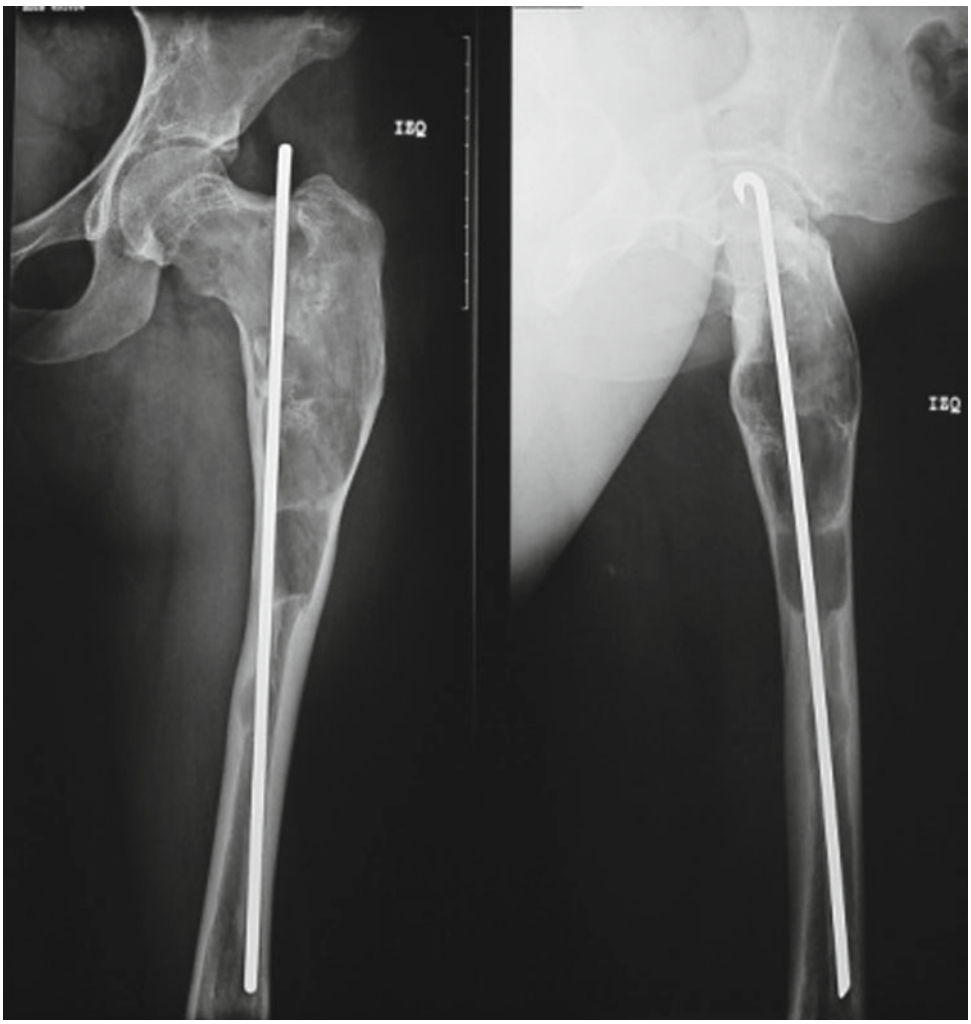

Fig. 20.2 Rx of a patient 23 years old, previous surgery at 15 years old; cystic images, frosted glass aspect, varus deformity

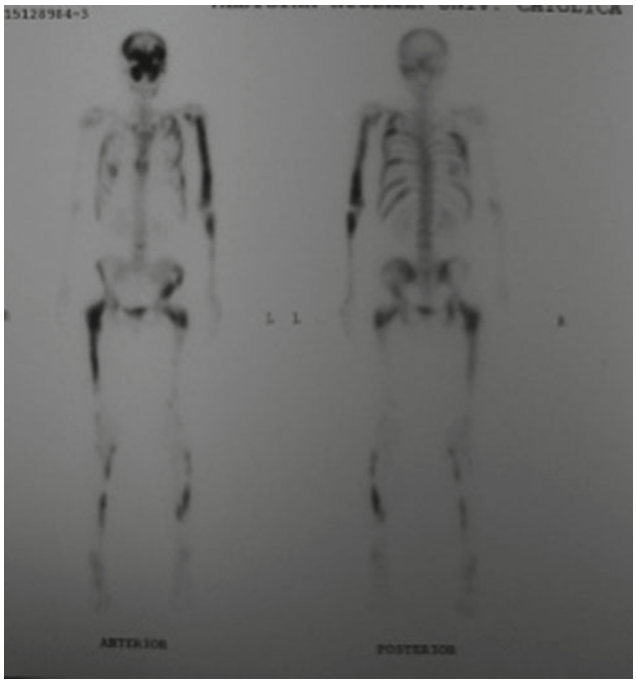

Fig. 20.3 Bone scan of patient with polyostotic fibrous dysplasia

There is fibroblast proliferation surrounding islands of woven bone. The truncated dysplastic trabeculaes are named as the "soup of Chinese alphabet." Another typical finding is the lack of osteoblasts around the bone trabeculaes.

Diferential Diagnosis

When there is an image of cartilage enchondroma can be proposed and if it is poliostotic, Ollier disease can be proposed. Poliostotic lesions can be proposed with neurofibromatosis but in these cases the typical intramedular bone findings of fibrous dysplasia are not present. A solitary cyst can be also be proposed.

If there is a lonely tibia lesion the diferential diagnosis will be with osteofibrous dysplasia or adamantinoma [12].

Treatment

A biopsy is useful to define the diagnosis.

Observation is indicated in asymptomatic pre-adolescent patients. In painful, big monostotic lesions intramedular fixation is indicated [8] Curretage alone has a high incidence of relapse, so must be combined with a structural cortical bone graft (fibula) or cancellous allograft and endomedular synthes according to the case. Autogenous bone graft is not useful because it quickly turns into fibrous dysplastic bone.

Coxa vara deformity is an indication for intertrocanteric osteotomy [6].

In adults with deformity and extensive proximal femur lesions a hip endoprosthesis can be indicated (Figs. 20.4 and 20.5).

The medical treatment with pamidronate disodium [5, 7, 9–11] can be useful in painful cases. Doses: 0,5 to 1,5 by

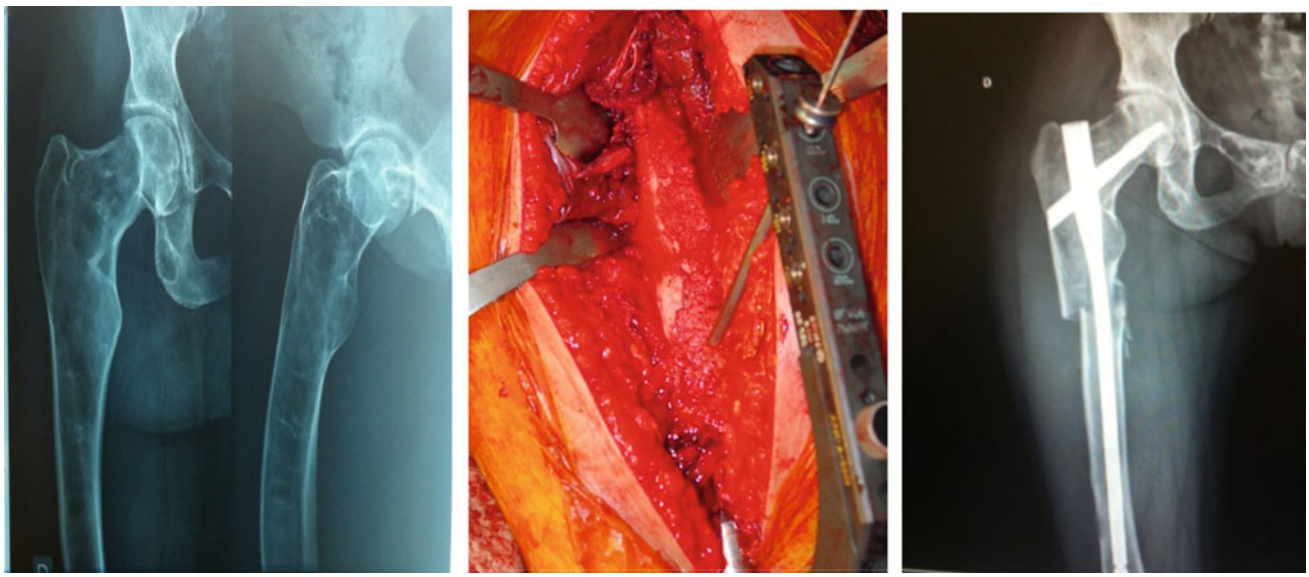

Fig. 20.4 Left: Rx showing fibrous dysplasia in the right femur; Center: intraoperative cancellous allograft; Right: OTS with endomedulary system (TFN)

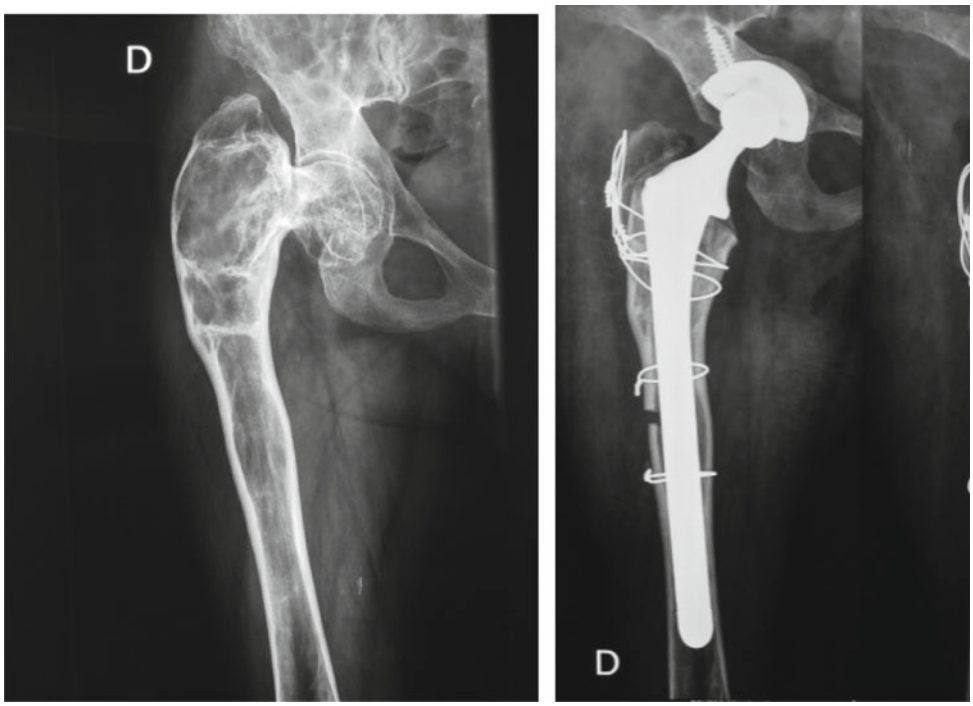

Fig. 20.5 Rx proximal femur: reconstruction with a non-cemented endoprosthesis stem and cup full-porous coated, trocanter osteotomy and proximal lateral osteotomy

each kg of weight of the patient Iiv in three days repeated every three or four months.

There is no indication for radiotherapy. There is a risk of long term secondary malignant lesions.

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