Fibrous Dysplasia

53

Liliana G. Olvi and Eduardo Santini-Araujo

Abstract

Fibrous dysplasia is a benign intramedullary fibro-osseous "dysplastic" noninherited bone lesion. The disorder may be monostotic or polyostotic. There is slight female predominance. Monostotic lesions (75 %) are more frequent in second and third decades. Polyostotic lesions (25 %) are more frequent in the first decade. Approximately 3 % patients with polyostotic lesions develop McCune–Albright syndrome associated with café-au-lait pigmentation of the skin and endocrine anomalies. Monostotic and polyostotic forms arise more frequently in long bones (proximal femur, tibia, humerus), craniofacial bones—especially in the jaws (the maxilla is more involved than the mandible)—and ribs. Any bone may be affected. Roentgenologic appearances in monostotic or polyostotic form are similar. Centered, intramedullary, elongated lesions arise in metaphysis or diaphysis of long bones. Epiphyseal location is uncommon while the growing plates are open. The lesion has a ground glass–like or lytic appearance. Histologically, the lesion is composed of a bland proliferation of plump fibroblasts in a dense collagenous, well-vascularized matrix. Within the fibrous tissue, in typical fibrous dysplasia, a variable presence of discontinuous immature woven osteoid or bone trabeculae is seen with flat or spindle-shaped osteoblasts rimming. Treatment in monostotic asymptomatic lesions is observation, surgical treatment, curettage, and grafting (auto- or allografts) or resection in cases of pathological fracture or risk of fracture or to correct deformities.

Definition

- Benign intramedullary fibro-osseous "dysplastic" noninherited bone lesion.
- The disorder may be monostotic or polyostotic.

Etiology

- At present, it is postulated that the immature bone production is determined by a post zygotic point mutation of the *GNAS1* gene, which encodes the α-subunit of the Gs α-stimulatory protein.

Clinical Features

Epidemiology

- It is a common lesion.
- Slight female predominance.
- Monostotic lesions (75 %): more frequent in the second and third decades.

L.G. Olvi, MD
Laboratory of Orthopaedic Pathology, Buenos Aires, Argentina

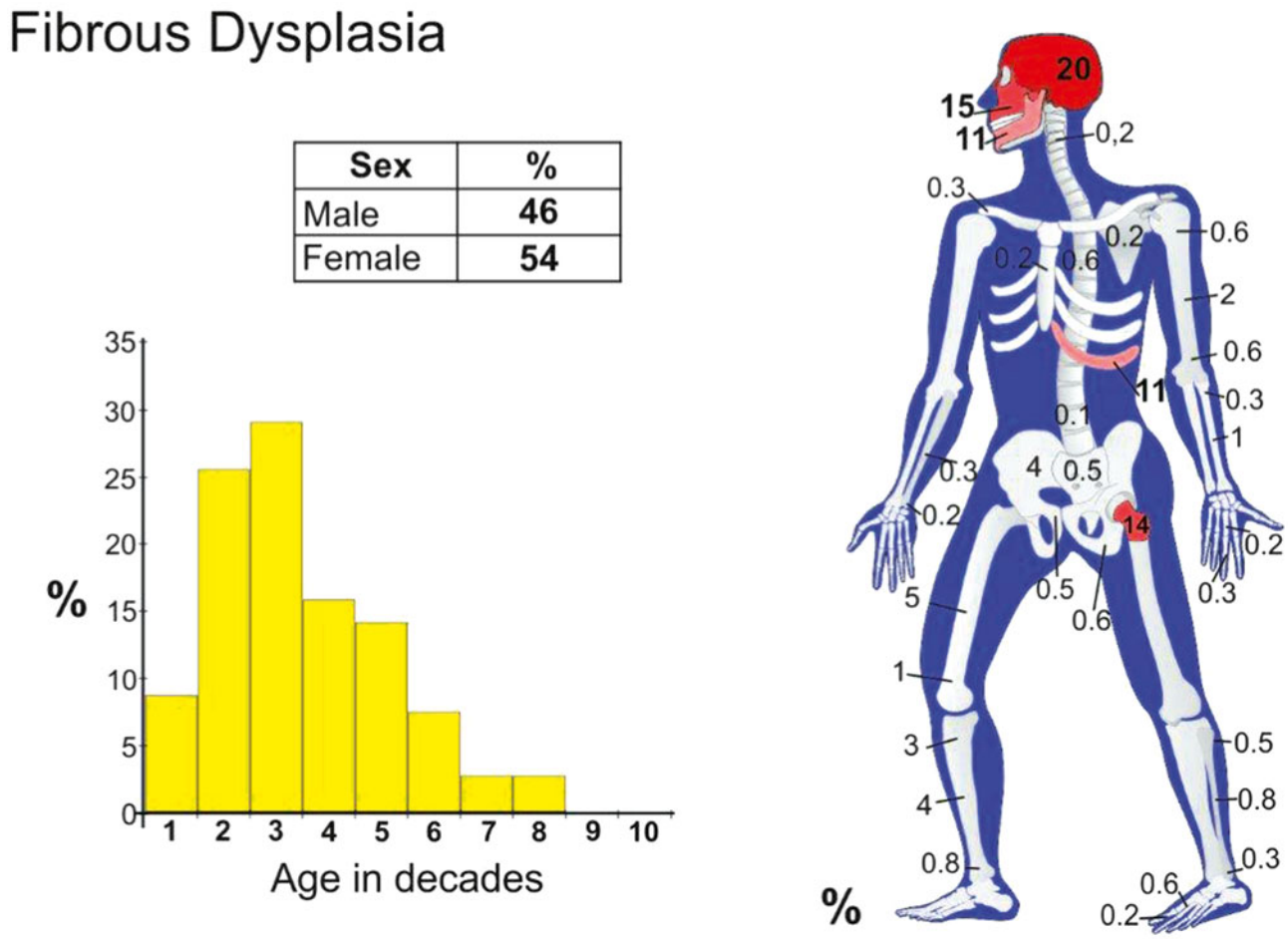
E. Santini-Araujo, MD, PhD (✉)
Laboratory of Orthopaedic Pathology, Buenos Aires, Argentina

Department of Pathology, School of Medicine and School of Dentistry, University of Buenos Aires, Buenos Aires, Argentina

Central Army Hospital, Buenos Aires, Argentina
e-mail: santiniaraujo@laborpat.com.ar

E. Santini-Araujo et al. (eds.), *Tumors and Tumor-Like Lesions of Bone: For Surgical Pathologists, Orthopedic Surgeons and Radiologists*, DOI 10.1007/978-1-4471-6578-1_53, © Springer-Verlag London 2015

- Polyostotic lesions (25 %): more frequent in first the decade.
- Approximately 3 % patients with polyostotic lesions develop McCune-Albright syndrome, associated with café au lait pigmentation of the skin and endocrine anomalies.
- Rarely, fibrous dysplasia is associated with soft tissues myxomas, usually in intramuscular location (Mazabraud syndrome).



Sites of Involvement

- Monostotic and polyostotic forms arise more frequently in long bones (proximal femur, tibia, humerus) and craniofacial bones – especially in the jaws (the maxilla is more involved than the mandible) – and ribs. Any bone may be affected.
- Monostotic disease affects a single bone usually with one focus. Rarely, more foci may be seen in the same bone.
- Polyostotic fibrous dysplasia is more frequently monomelic, polyostotic monomelic fibrous dysplasia – involving homolateral lower limbs and ilium. The more severe cases are polymelic.
- Spine is an infrequent location, more frequently associated with the polyostotic form.
- Rib location is more frequent in older patients, probably due to older patients having more commonly chest radiography, and the lesions are incidentally discovered.

Clinical Symptoms and Signs

- Usually asymptomatic.
- Incidental radiographic finding after a roentgenological examination for other reasons, most frequently in ribs.
- Pain may appear depending on site and extent of the lesion.
- Occasionally, localized swelling may appear, more frequently in the jaws and the skull.
- Monostotic lesions may progress and enlarge, most commonly during skeletal growth. They tend to stabilize after puberty, but lesions may not be quiescent in adulthood.
- Some lesions may grow during pregnancy.

- Some lesions may produce deformity.
- Pathologic fracture – frequently in the proximal femur – and deformity of weight-bearing bones are commonly seen.
- Craniofacial lesions may produce asymmetry, exophthalmos, impingement of cranial nerves – like the optic nerve – affection of middle ear structures, and deformity. Chronic headache may be present.
- Polyostotic form may involve a few bones of a single extremity or compromise more than 50 % of the skeleton. This presentation begins early in life – before 10 years of age – and the disease has a rapid progress.
- Limb length discrepancies.
- Polyostotic forms associated to McCune-Albright syndrome – which is more common in females – present cutaneous pigmentation in the form of yellow or brown patches frequently located on neck, back, shoulder, chest, and pelvis. Albright macules of fibrous dysplasia have irregular and dented borders like the coast of Maine, whereas in neurofibromatosis, the macules are irregular but with smooth borders resembling the coast of California, endocrine hyperactivity; precocious sexual development – sometimes starting as early as 3 years – vaginal bleeding; development of breasts; hyperthyroidism; frequently secondary hyperparathyroidism; Cushing's syndrome; acromegalia; hyperprolactinemia; adrenal hyperplasia; early bone maturation with premature closure of the growth plates and consequent short stature; and renal phosphate wasting – in 50 % of the patients. Infrequently, excess FGF-23 is produced, determining osteomalacia as in the phosphaturic mesenchymal tumor. If not detected in time, these patients may present deformities and fractures, leading to a crippling disease.

Image Diagnosis

Radiographic Features

- Roentgenological appearances in monostotic or polyostotic form are similar.
- Centered, intramedullary, elongated lesions arising in metaphysis or diaphysis of long bones. Epiphyseal location is uncommon, while the growing plates are open.
- Well-circumscribed areas of rarefaction, commonly bordered by a sclerotic rim, frequently so thick that appear as a rind.
- The upper or lower limit frequently shows a triangular shape.
- The lesion has a *ground-glass*-like or lytic appearance. The roentgenological density varies in relation with the intralesion amounts of fibrous or osseous tissue.
- The lesions may expand the affected bone – but have sharp circumscription – especially in rib, pelvis, fibula, and small tubular bones. Lesion scallops and erodes the endosteal surface, thinning the cortex, sometimes expanding bone and determining a newly formed cortex.
- Periosteal reaction and soft-tissue extension are extremely infrequent, without a pathologic fracture occurring.
- Rarely some lesions produce a large expansile mass that bulges into soft tissues. More frequent in craniofacial skeleton. Some authors classified this case as *fibrous dysplasia protuberans*.
- Craniofacial lesions may be extremely dense, especially the maxilla and skull base.
- Bone deformity – bowing – usually affects weight-bearing long bones such as the femur – "shepherd's crook deformity" – and the tibia.
- In rare lesions with cartilaginous component, a radiographic pattern of ring and spotty calcifications is frequently present. This pattern is especially frequent in the upper end of the femur and in polyostotic disease.
- Pathologic fracture may occur. The fracture line is often transverse similar to what occurs in Paget's disease fractures. This pattern of fracture reveals that the microstructure of bone is disturbed, like in Paget's disease or osteopetrosis.
- Rarely secondary ABC may give an aggressive appearance to the lesion that simulates a sarcoma.

CT Features

- Expansile well-circumscribed lesion.
- Soft mineralization in relation with ground grass roentgenographic pattern.
- The density is related with the mineralization of the lesion.
- Helpful in delimiting the involvement especially in maxillary and craniofacial bones.

MRI Features

- Well-circumscribed T2-weighted lesion.
- The ground-glass pattern is seen as a soft mineralized lesion.
- Cysts appear with fluid signal characteristics, with a homogeneous hyperintense signal on T2-weighted images.
- Shows the scalloping on the endosteal surface of the cortex.

Bone Scan

- High signal

PET Scan

- High uptake

Image Differential Diagnosis

Periosteal Desmoid
- Radiologically similar to cortical fibrous defect
- Different histological pattern

Osteofibrous Dysplasia
- Cortical lesion arising in the tibia and fibula of children
- Frequently produce an anterior bowing of the affected bone
- Not a medullary lesion

Benign Fibrous Histiocytoma
- Radiolucent with sclerotic margins
- Epiphyseal and diaphyseal location
- Intramedullary location

Giant Cell Tumor
- Epiphyseal lesion involving metaphysis in its evolution.
- Highest incidence in the third decade of life.
- More frequent in females.
- Inner border of the lesion is lytic. Extremely rare to be sclerotic.

Solitary Bone Cyst
- Well-outlined centrally and symmetrically located metaphyseal-diaphyseal lucency, expanding, thinning, and scalloping the cortices.
- The cyst has its greatest diameter near the epiphyseal plate and its conic end in the shaft.
- A multilocular or trabeculated appearance may be seen due to prominent endosteal bony ridges in the inner cortical wall.
- MRI usually confirms the cystic nature and its fluid content without solid components that can be bloody in fractured lesions.

Chondroma, CHS
- Usually present spotty dense calcifications

Chondromyxoid Fibroma
- Is usually a more radiolucent lesion in comparison with FD

Eosinophilic Granuloma
- Radiolucent, eccentrically elongated lesion in the metaphysis of long bones, juxtaposed to the endosteal surface of one cortex

Metaphyseal Fibrous Defect
- In small bones or long bones of small diameter, the lesion may occupy the whole diameter of the medullary cavity and pose the differential with MFD.
- Radiologically, MFD is a radiolucent, eccentric lesion juxtaposed to the endosteal surface of one cortex in contrast to FD, which is more centrally located in the medulla.
- As long bone grows, it becomes separated from the epiphyseal plate, moving toward the diaphysis.
- Lesions tend to thin and slightly expand one or both cortical surfaces.
- The inner border is well defined and slightly sclerotic, suggestive of a nonaggressive lesion, producing a scalloped margin.
- Infrequently, large lesions may involve the entire width of the bone.

Desmoplastic Fibroma
- Usually associated to cortical disruption

ABC
- Area of lucency situated eccentrically in the medullary cavity in the metaphysis of a long bone.
- Most ABCs are completely lytic, but a few contain traces of mineral.
- Frequently presents a multiloculated appearance.
- Less commonly, the lesion may be situated centrally.
- Later on, "ballooned" or "aneurysmal" cystic expansion of the affected bone –"blowout" – is evident. Usually forms a thin sclerotic rim of ossification due to periosteal new bone formation.
- CT and MRI show a cystic and radiolucent lesion, with internal septations and multiple fluid-fluid levels. Some FD, however, may show features of secondary ABC. In any case, both CT and MRI help to highlight the eventual presence of an underlying lesion.

Well-Differentiated Intramedullary Osteosarcoma
- Lacks a sclerotic border
- Permeative margins

Pathology

Gross Features

- Frequently, the material is obtained by core needle biopsy.
- In surgical biopsies, tissue is tan-gray, dense, and fibrous.
- The number and size of immature osteoid or bone trabeculae present within the fibrous tissue give it a more or less gritty quality to the cutting procedure.
- When the lesion is more mineralized, the appearance is white to yellow.
- Sometimes the lesion may present a dense ossification, most common in craniofacial and jaws locations.
- In surgical specimens, the lesions are intramedullar centrally located and well defined, the bone contour is expanded, and the cortex is thinned. In older lesions, an edge of reactive sclerotic bone may be seen.
- Cyst formation is commonly present, and the cyst cavity is filled with yellowish clear fluid. Cyst formation may be slight or extensive.
- Secondary aneurysmal bone cyst may sometimes appear with blood-filled multicameral cyst.

- Yellowish areas are related to lipid-laden histiocytes – xanthoma cells.
- Chondroid foci, when present, are seen as bluish translucent nodules. Rarely, the cartilaginous component may predominate.
- Hemorrhages and hemosiderin pigment are found after pathologic fractures.

Histological Features

- Histologically, the lesion is composed by a bland proliferation of plump fibroblasts in a dense collagenous, well-vascularized matrix by thin capillary vessels.
- The spindle cells present a storiform or whorled pattern without cytological atypia, hyperchromatism, or nuclear pleomorphism.
- Mitoses are extremely infrequent. Mitoses are seen in the picture of pathologic fractures. Atypical mitoses are not seen.
- Within the fibrous tissue, in typical fibrous dysplasia, a variable presence of discontinuous immature woven osteoid or bone trabeculae is seen, with flat or spindle-shaped osteoblast rimming.
- Bony trabeculae of different sizes are arranged in a haphazard, nonfunctional fashion. They are peculiar with so-called "Chinese characters" shapes. They are curvilinear, in C, U, and sometimes circumferential shapes.
- In more mature lesions, trabeculae may present reversal cement lines simulating the histological appearance of Paget's disease.
- The presence of a lamellar pattern in some trabeculae is extremely infrequent.
- With silver stain for reticulin fibers or polarization microscopy, it is possible to see the continuity between newly formed reticulin fibrils of the dysplastic stroma and those that form the matrix of woven trabeculae. This corresponds to a preexisting compressed network of newly formed reticulin fibers, which later mineralize with calcium granules.
- Typical fibrous dysplasia may show large areas without bone formation. These areas have a particular storiform pattern of the spindle cells.
- The presence of rounded ossicle-like or cementicle-like structures resembling psammoma bodies or *cementoid bodies* is frequently found in fibrous dysplasia. This feature is most commonly seen in lesions arising in the base of the skull and sometimes is erroneously diagnosed as meningiomas. This pattern is also seen in other sites as long bones but rarely are a prominent component of the lesions.
- Osteoclasts can be sometimes observed resorbing immature bony trabeculae.
- Hemorrhagic areas and hemosiderin pigment deposits are found after pathologic fractures as well as areas of necrosis.
- Other cytological tissue patterns that can be observed are:
  - Collections of foam cells.
  - Frequent clusters of multinucleated giant cells.
  - An occasional fibrous dysplasia will present extensive areas of hypocellular myxoid matrix and may be called myxoid fibrous dysplasia.
  - Islands of cartilage may predominate in some cases of fibrous dysplasia as round nodules that in some lesions may resemble the structure of epiphyseal plate cartilage. Endochondral ossification may be observed. Some authors have called these cases *fibrocartilaginous dysplasia*.
  - Areas of cystic degeneration with serous fluid content may be found, sometimes surrounded by lipophages and multinucleated giant cells.
  - Secondary aneurysmal bone cyst formation may be present in fibrous dysplasia.
- Rarely, sarcomas may be associated to fibrous dysplasia – most commonly in polyostotic disease. Post-radiation sarcoma in FD may be originated from radiation therapy. Most commonly, these sarcomas are: osteosarcomas, fibrosarcomas, and chondrosarcomas. Facial bones and the femur are the more frequently involved bones.

Pathologic Differential Diagnosis

Osteofibrous Dysplasia

- Both entities share histological feature's resemblance.
- OFD presents immature curvilinear bone trabeculae rimmed by prominent plump osteoblasts.
- Spindle cells in a dense collagenous stroma.
- Spindle cells express keratin and EMA.
- OFD is a cortical lesion, localized predominantly in the tibia of children.

Ossifying/Cementifying Fibroma

- Fibro-osseous lesion of the jaws.
- FD contacts with the cortex and merges into it. Frequently erodes and thins the cortex.
- OCF is more clearly delimitated and separated by fibrous tissue from the endosteal surface of the cortex.
- Images are extremely useful to make a sharp differential diagnosis.

Metaphyseal Fibrous Defect

- Histologically, some FD present areas of foamy cells that determine that, in small biopsy specimens, the differential with MFD may be considered.
- In MFD, fibrous tissue presents a characteristic whorled – storiform or cartwheel – pattern more evident than in FD, with the presence of histiocytes and areas of lipid-bearing histiocytes and hemosiderin pigment-loaded histiocytes. Variable number of multinucleated

giant cells are scattered throughout the lesion, usually in clusters.
- Inflammatory elements – lymphocytes and plasmacytes – are another characteristic finding.
- Lack of new bone formation except in the margins of the lesion. The presence of new bone formation without pathologic fracture is extremely rare.
- The association with pathologic fracture presents periosteal and endosteal new bone formation and areas of necrosis.

Desmoplastic Fibroma
- Lack of woven bone formation
- No storiform pattern
- Presence of intersecting fascicles of spindle cells

Chondrosarcoma
- Small microscopic foci of chondroid tissue may be seen in FD, but sometimes the chondroid differentiation is massive and the differential between chondroid tumors must be considered.
- The chondroid areas in FD do not present cytological atypia.
- The cartilage lobules in FD present a characteristic encasement with endochondral ossification.
- Infiltration of bone marrow areas is never seen in FD.

Parosteal Osteosarcoma
- May be very difficult to diagnose on pure histological grounds. Knowledge of clinical and x-ray data by the pathologist is indispensable.
- Parosteal osteosarcoma arises in the surface of bone and not in the medullary cavity.

Low-Grade Central Osteosarcoma
- Stromal spindle cells are more elongated and present nuclear hyperchromasia and cellular atypia.
- More frequent mitoses.
- Lack of storiform pattern.
- Permeation of bone marrow areas encasing native lamellar trabeculae.
- Tumor cells express MDM2 and CDK4 by immunohistochemistry or molecular techniques.

Meningioma in the Base of Skull
- The presence of rounded ossicle-like or cementicle-like structures resembling psammoma bodies *or cementoid bodies* in FD, pattern that is especially prominent in skull base lesions, may mimic meningiomas.

Aneurysmal Bone Cyst
- FD may suffer secondary ABC changes. This differential is difficult to perform in small biopsies.
- The image correlation is of extreme value for the differential diagnosis.

Solid Aneurysmal Bone Cyst
- Almost all ABCs have some solid area that may be prominent in some of them and where cystic spaces and septae may not be identified. The term "solid ABC" has been adopted for these cases.
- More intralesional reactive bone.
- Less storiform pattern of the stromal spindle cells.

Fibromyxoma of the Bone
- Many of the cases reported are examples of myxoid fibrous dysplasia.

Paget's Disease
- In more mature lesions, trabeculae may present reversal lines simulating the appearance of Paget's disease trabeculae.
- Paget's disease has a peculiar mosaic pattern of lamellar bone, never seen in FD.
- The pagetic bony trabeculae show numerous cement lines.
- The pagetic trabeculae are rimmed by prominent osteoblasts alternating with areas of osteoclastic resorption.
- The bone marrow spaces are richly vascularized and with a peculiar loose and edematous fibrous tissue.
- The disease has a similar skeletal distribution to FD but affects older people.

Ancillary Techniques

- Immunohistochemistry has no value for diagnosis.
- Spindle cells express vimentin and do not express EMA, MDM2, or CDK4.

Genetics

Severity of manifestations of McCune-Albright syndrome depends on the number and types of cells that harbor mutation in GNAS protein. The GNAS gene provides instructions for making one part of a protein complex called a guanine nucleotide-binding protein, or a G protein.

Prognosis

- Monostotic lesions: excellent prognosis.
- Polyostotic have more risk of deformities and fractures.
- In large rib lesions, the patient may suffer respiratory problems.

- Spontaneous malignant transformation is rare. The risk is greater with the polyostotic variant. More frequent around the fourth decade of life, associated to lesions treated with radiotherapy.

Treatment

- In monostotic asymptomatic lesion: observation.
- Surgical treatment, curettage, and grafting (auto- or allografts) or resection in cases of pathologic fracture or risk of fracture or to correct deformities. Some patients require the use of nail/plate fixation.
- Polyostotic disease: the use of bisphosphonates has been suggested.
- Radiotherapy must be avoided due to risk of development of post-radiation sarcoma.

Images

See Figs. 53.1, 53.2, 53.3, 53.4, 53.5, 53.6, 53.7, 53.8, 53.9, 53.10, 53.11, 53.12, 53.13, 53.14, 53.15, 53.16, 53.17, 53.18, 53.19, 53.20, 53.21, 53.22, 53.23, 53.24, 53.25, 53.26, 53.27, 53.28, 53.29, 53.30, 53.31, 53.32, 53.33, 53.34, 53.35, 53.36, 53.37, 53.38, 53.39, and 53.40 for illustrations of fibrous dysplasia.

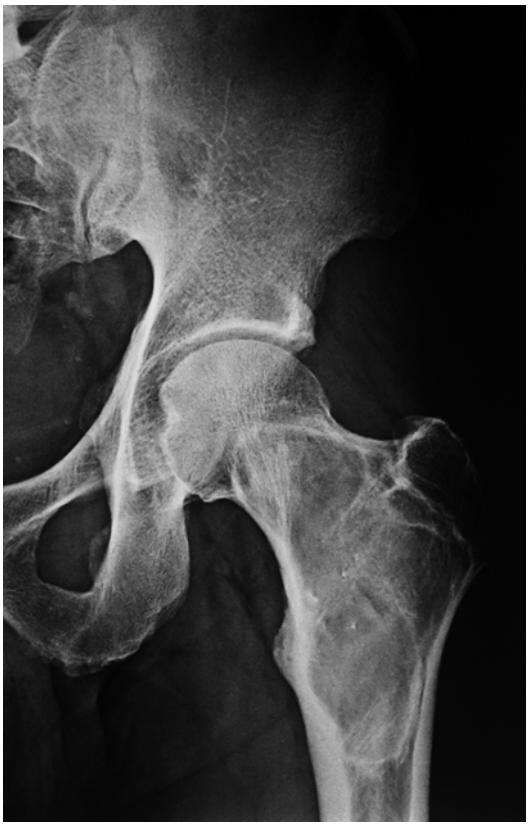


Fig. 53.1 FD of the upper third of the left femur. X-ray shows a radiolucent lesion with a "ground-glass" appearance

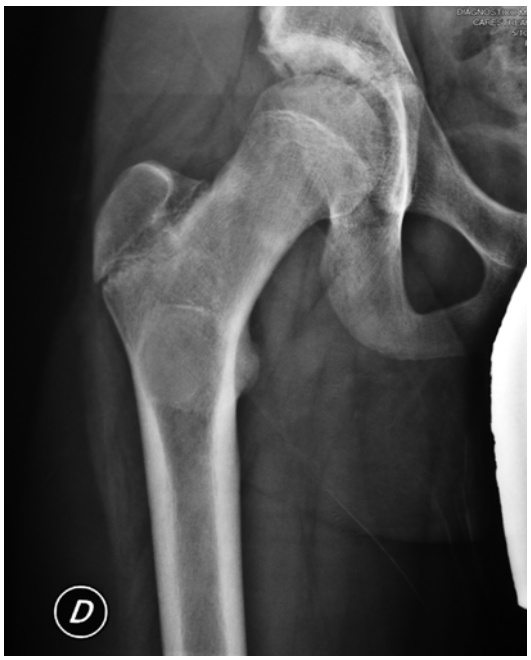


Fig. 53.2 FD. Radiographic features. Anteroposterior view of the right hip shows isolated lesion in metaphyseal-diaphyseal region with a typical "ground-glass" feature

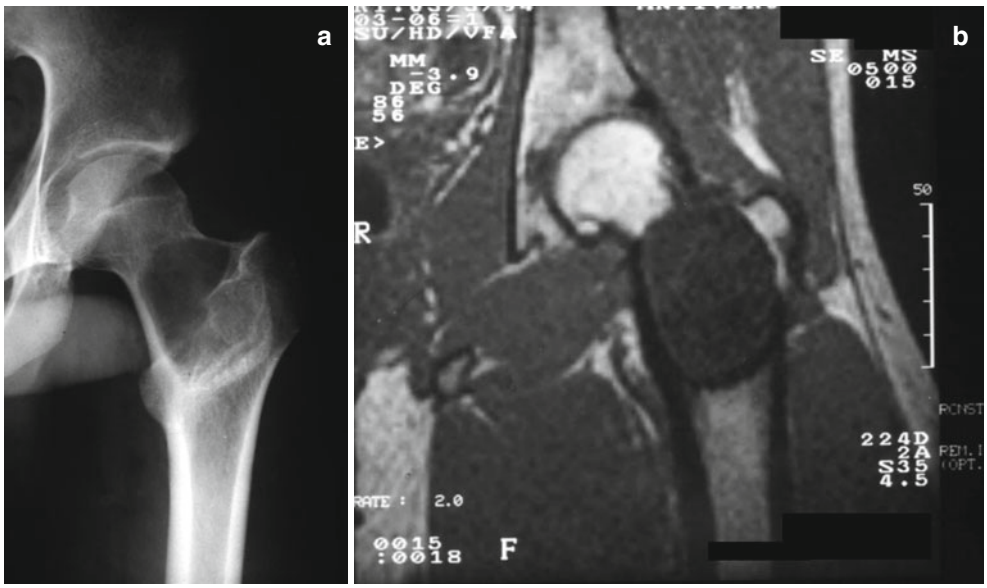


Fig. 53.3
(**a**) FD. Anteroposterior x-ray showing ringlike sclerosis of the bone at the periphery which is characteristic of FD. (**b**) T1-weighted MRI of the same lesion

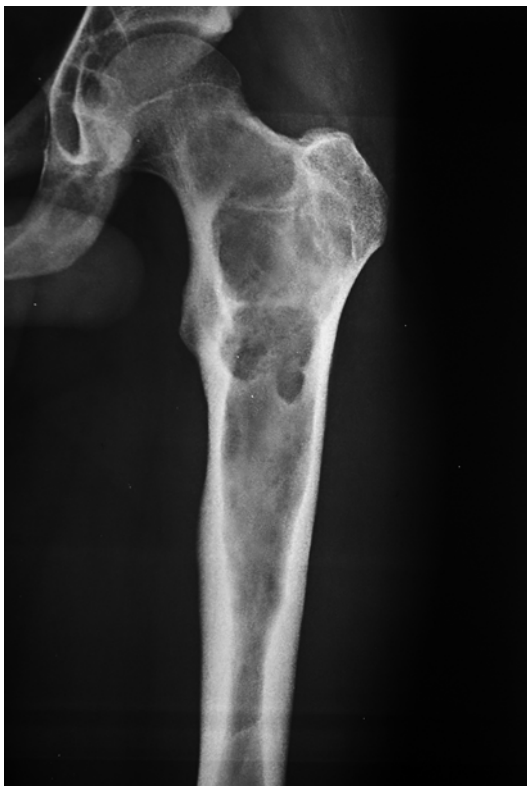


Fig. 53.4 FD. Anteroposterior radiograph of the left hip showing an involvement of the acetabular region and an expansile lesion in the proximal femur. Note the "ground-glass" density of the lesion

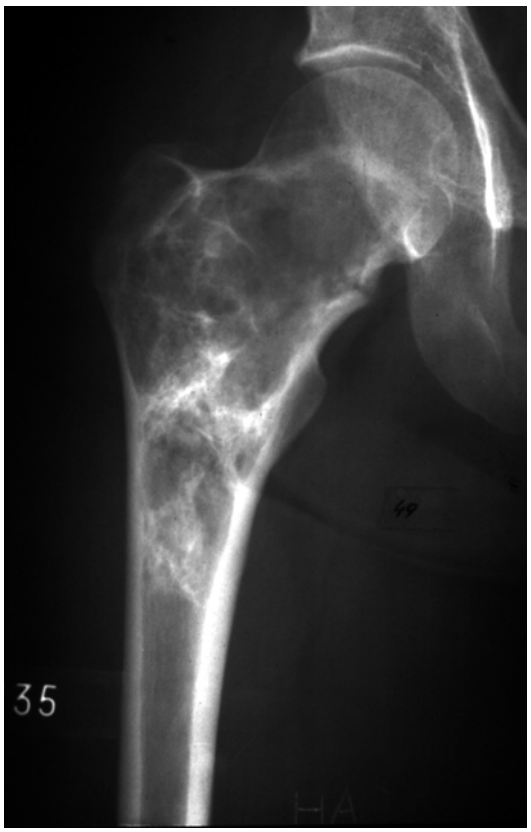


Fig. 53.5 FD. Anteroposterior radiograph of the right femur shows intramedullary lesion with central opacities and a fracture in the femoral neck

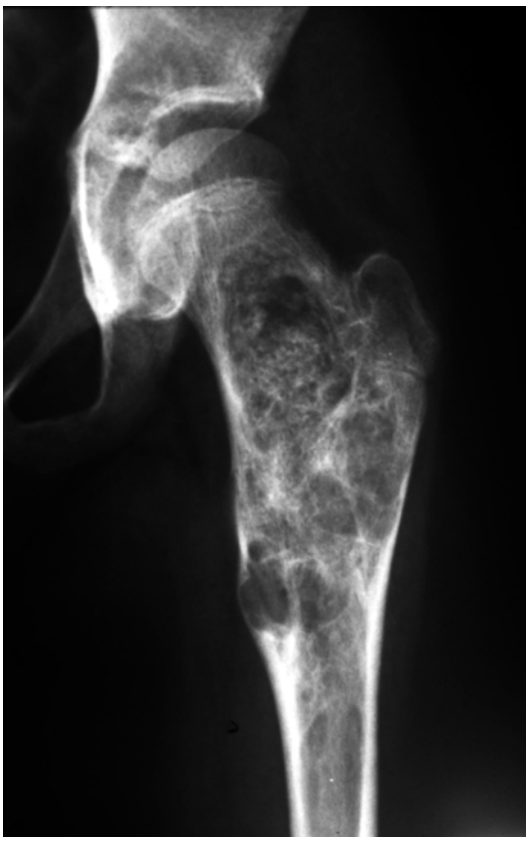


Fig. 53.6 FD. Typical fibrous dysplasia affecting the neck and shaft of the left femur showing a bowing deformity of the internal cortex due to secondary ABC

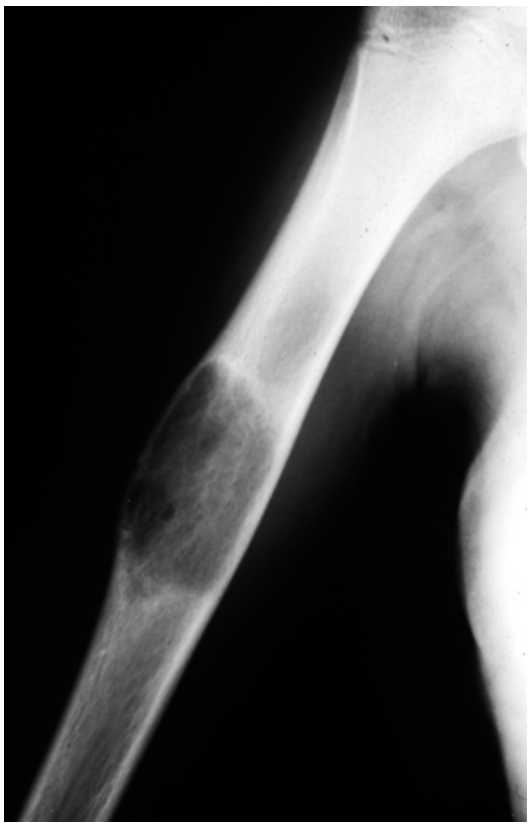


Fig. 53.8 FD. Anteroposterior x-ray of right humerus demonstrating expansion of bone contour with cortical thinning

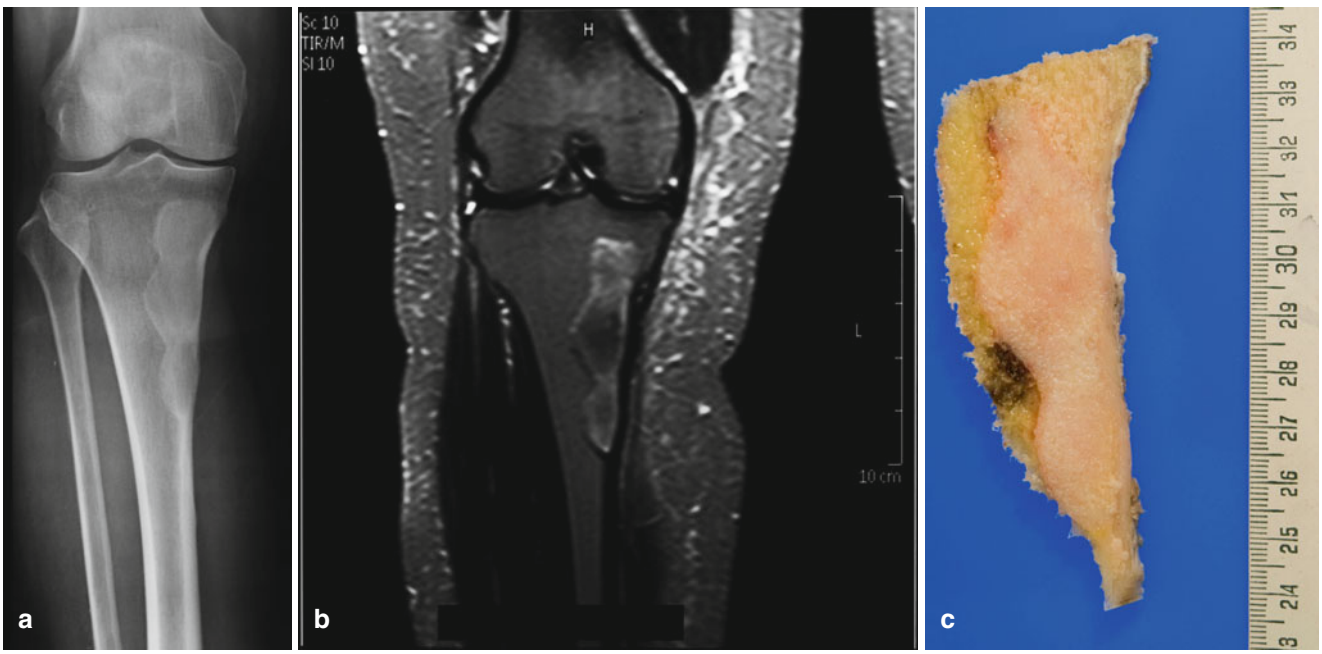


Fig. 53.7 FD. (**a**) Anteroposterior x-ray of the right tibia illustrating the "ground-glass" feature. (**b**) MRI of the same lesion. (**c**) Gross specimen

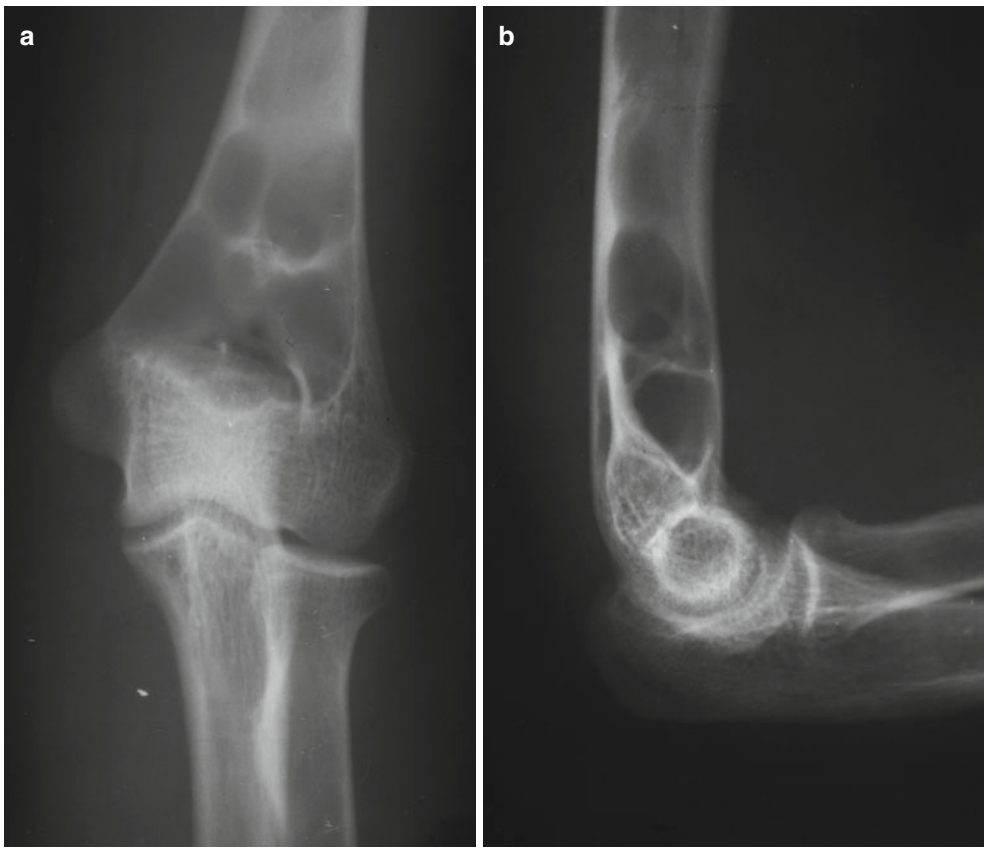


Fig. 53.9 FD. (**a**, **b**) Anteroposterior and lateral x-ray or distal humerus. "Ground-glass" feature

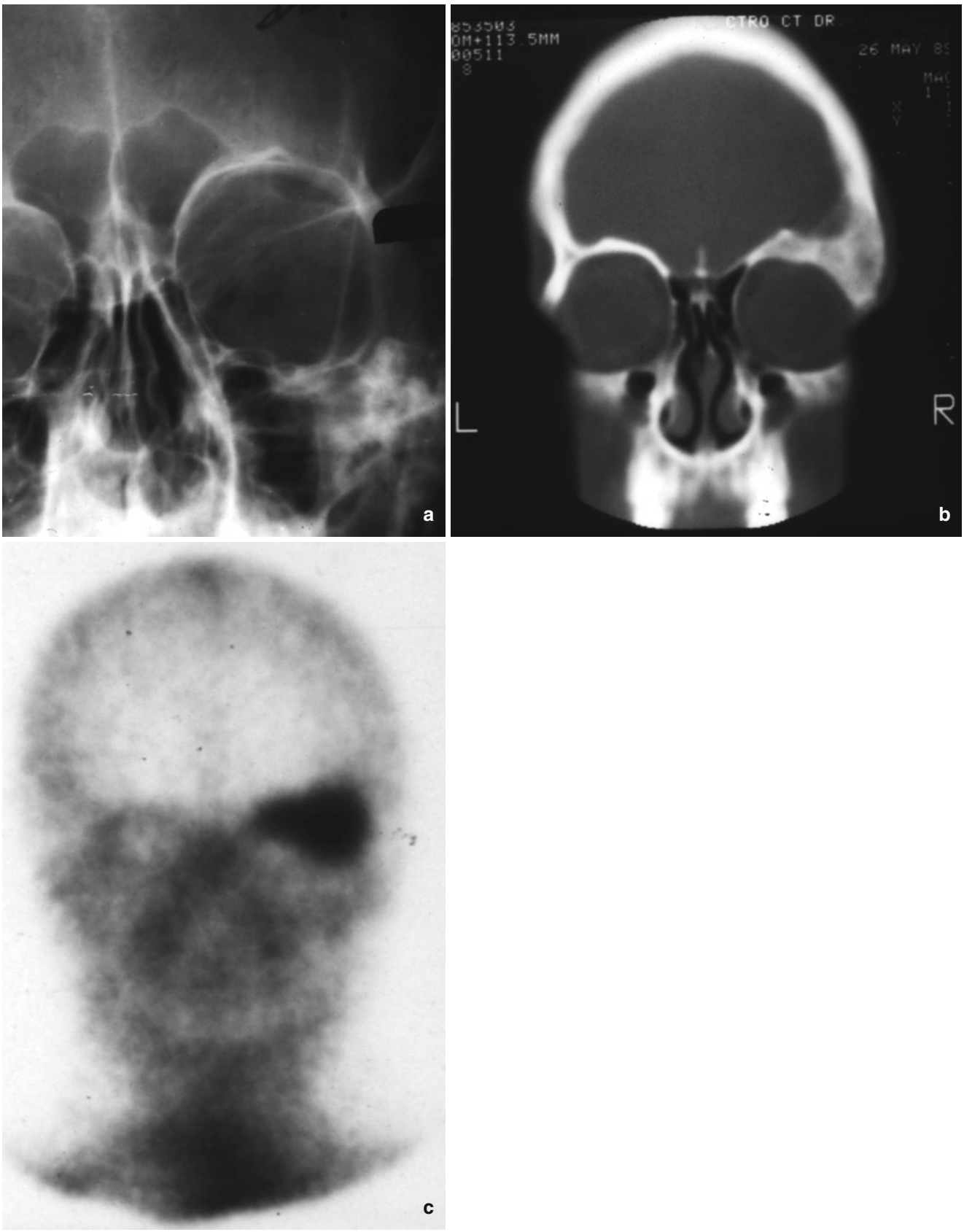


Fig. 53.10 FD. (**a**) Anteroposterior x-ray does not evidence lesion. (**b**) CT shows a "ground-glass" appearance of the supraorbital region. (**c**) Tc99m scan shows FD with dense uptake of the radioisotope

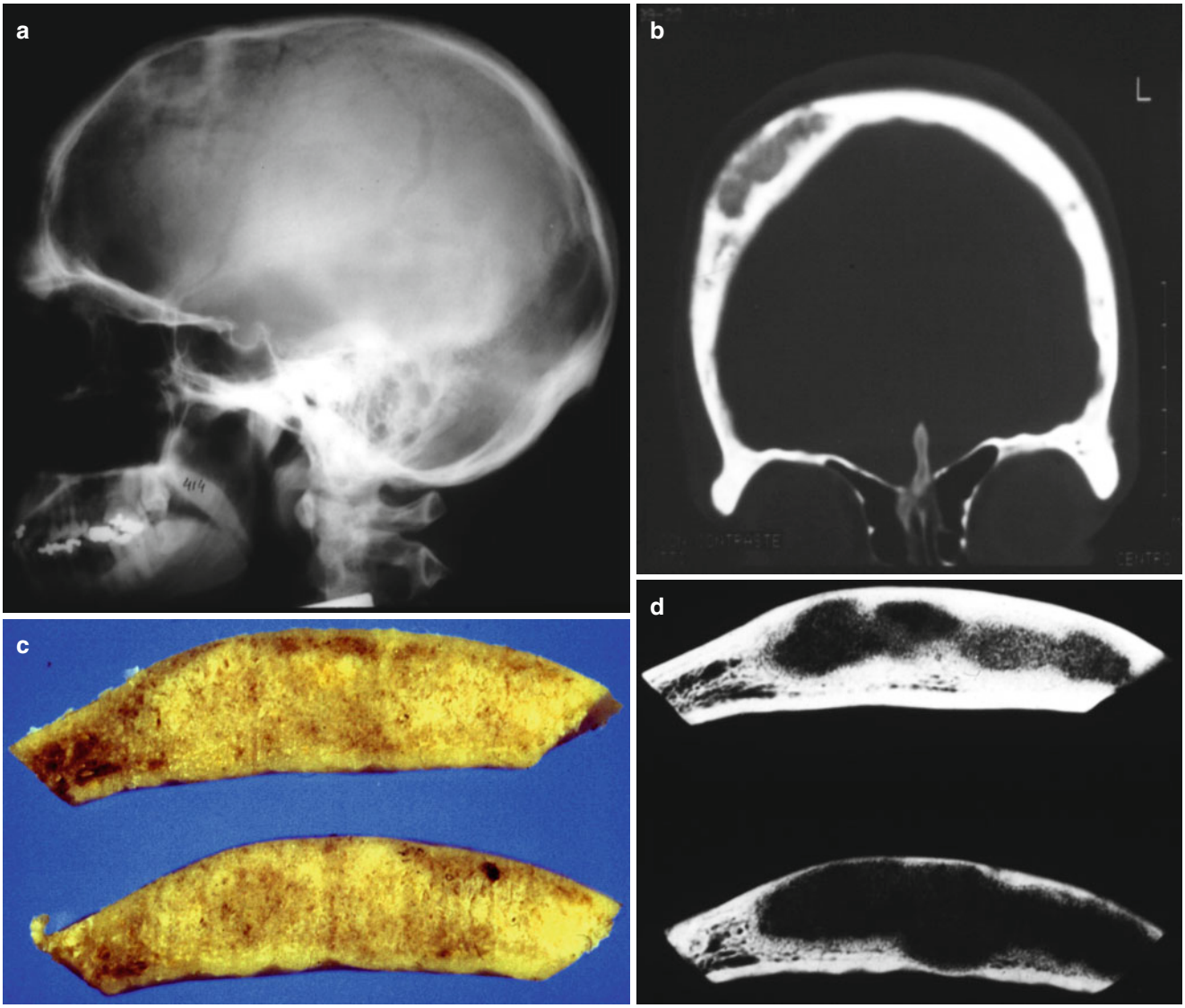


Fig. 53.11 FD. (**a**) Lateral x-ray of the skull showing a lytic lesion. (**b**) CT illustrating the same lesion. (**c**) Gross specimen. (**d**) X-ray of gross specimen

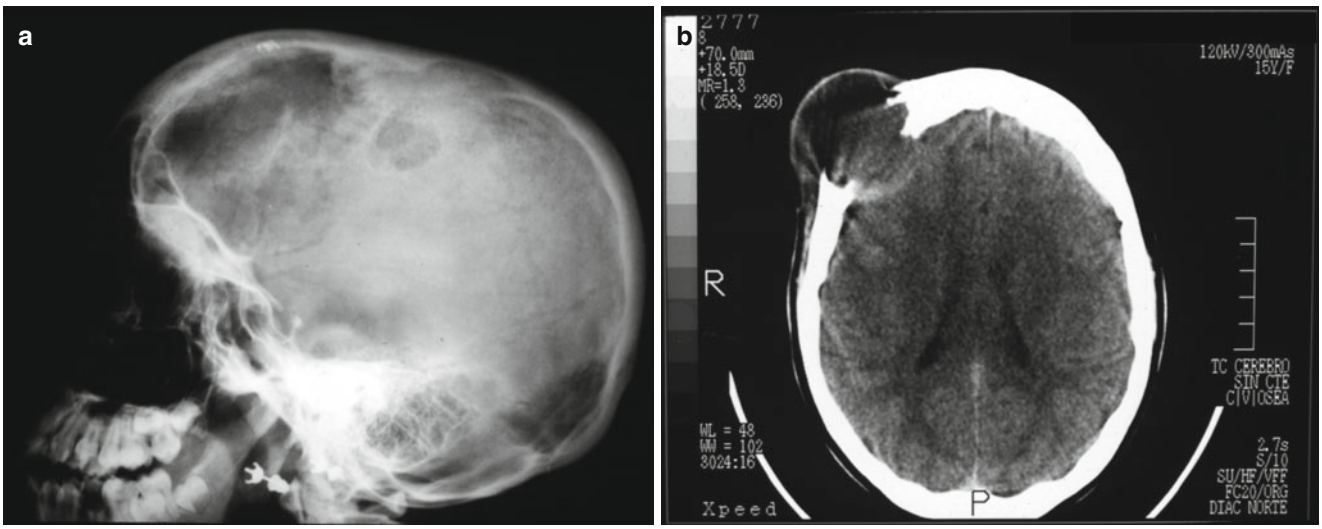


Fig. 53.12 FD. (**a**) Lateral x-ray of the skull showing a lytic lesion. (**b**) CT of the same lesion. FD in flat bones is often expansile

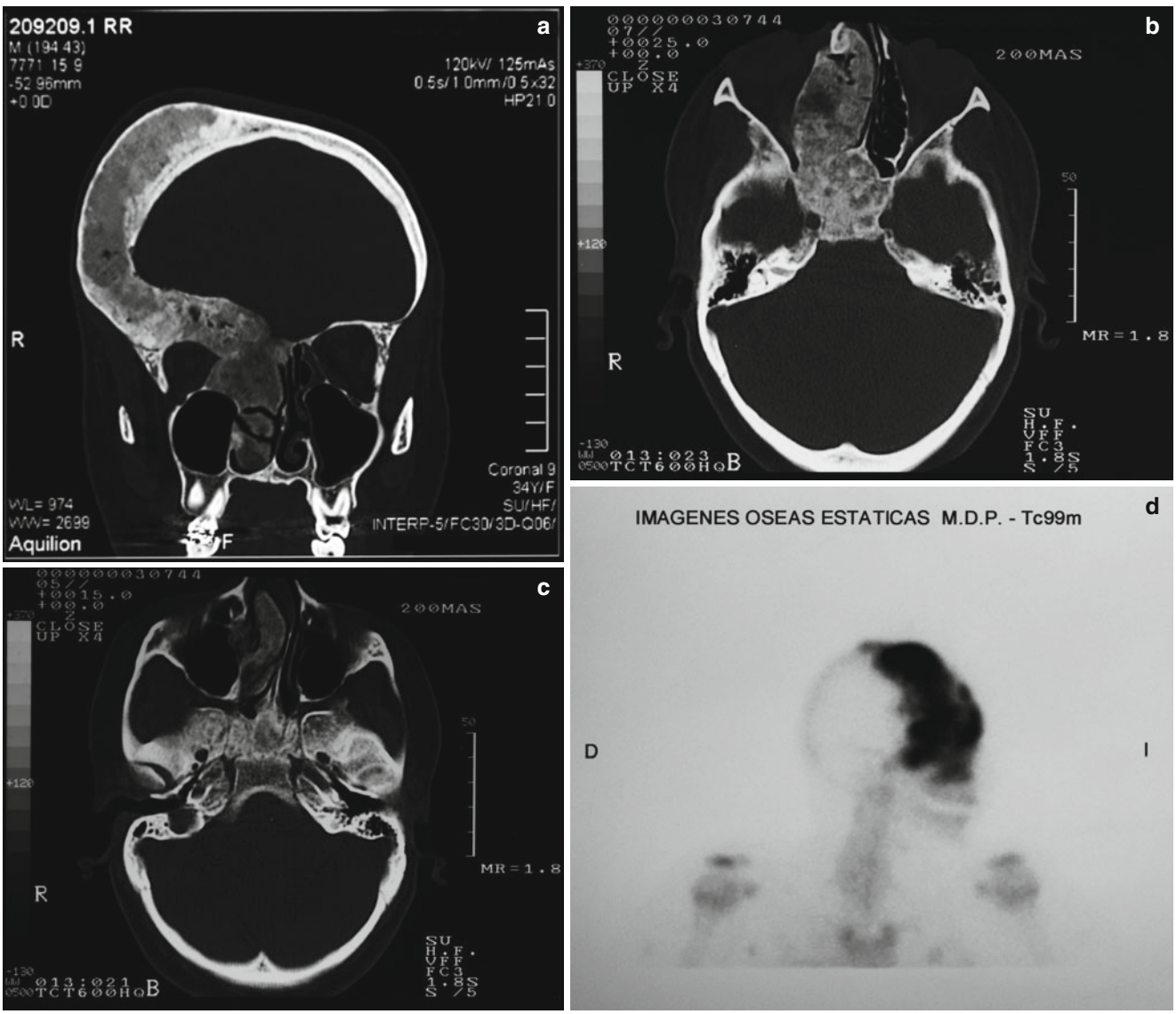


Fig. 53.13 FD. Coronal (**a**) and axial (**b**, **c**) CT view. Polyostotic involvement of skull bones with a "ground-glass" appearance. The lesion causes diffuse expansion of the diploic space and obliteration of the right sphenoid sinus and nasal cavity. Hot spot with Tc99m (**d**)

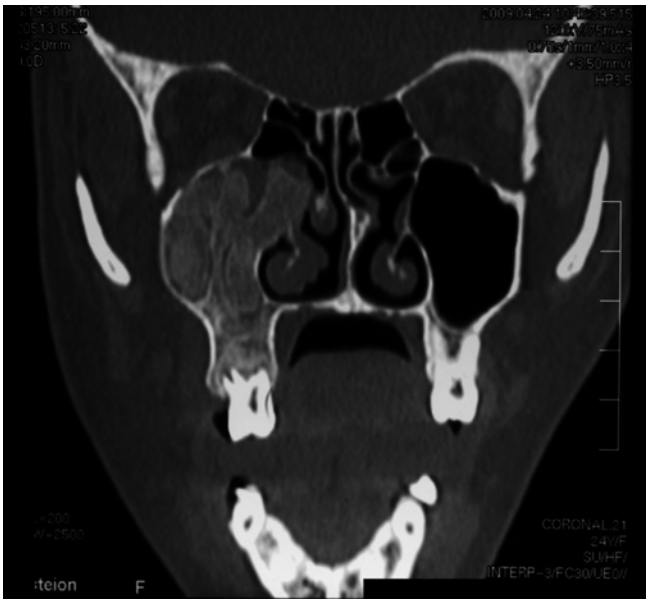


Fig. 53.14 FD. "Ground-glass" feature involving the right maxilla

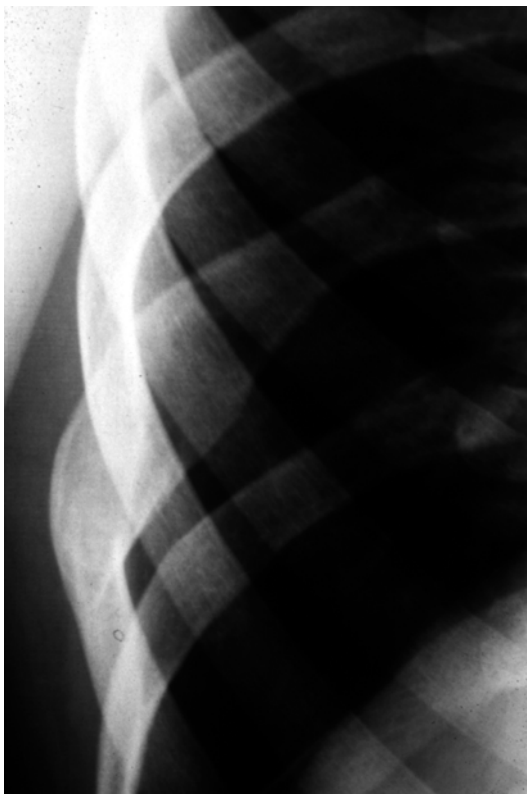


Fig. 53.15 FD of the rib. Rib is a common location for this lesion

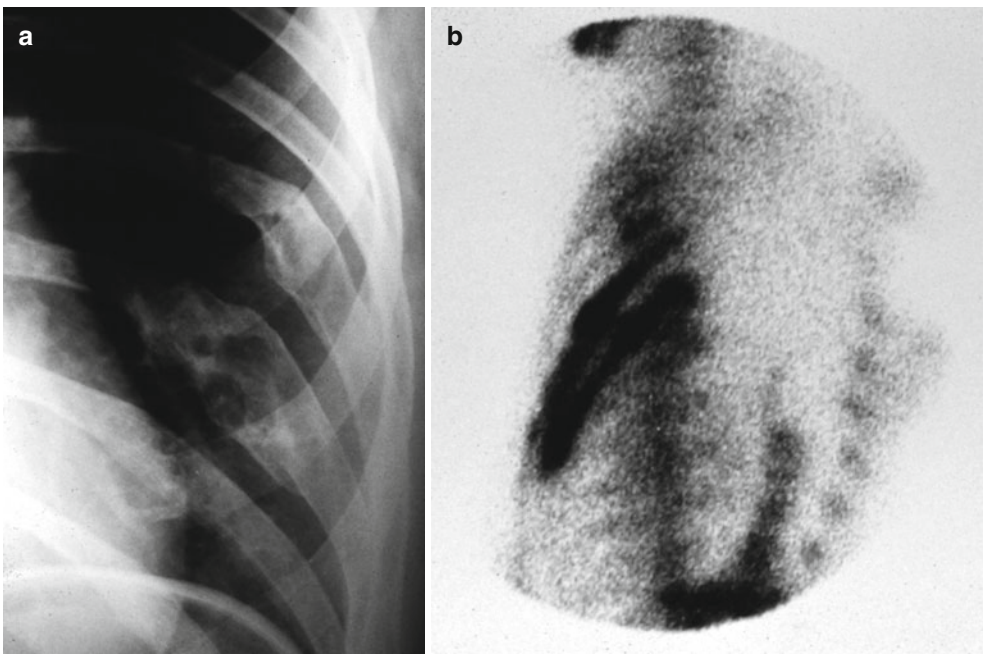


Fig. 53.16 FD. (**a**) Involvement of several ribs, large bubbly and expanded masses. (**b**) High uptake of radioisotope

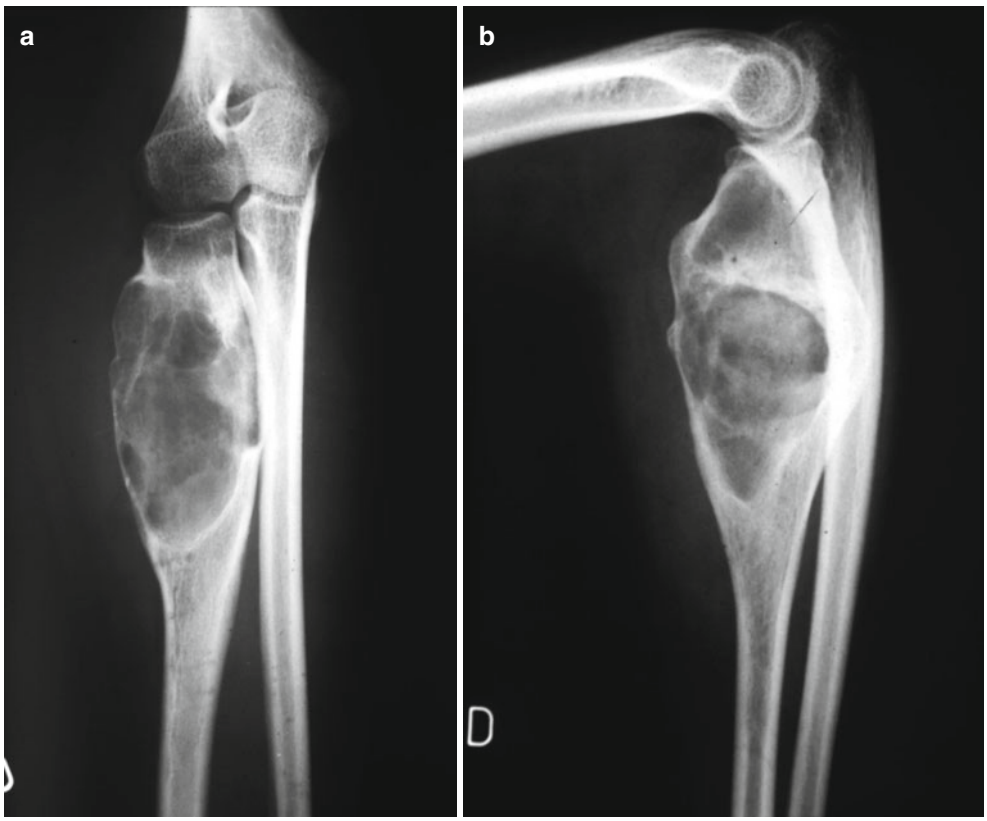


Fig. 53.17 FD. (**a**, **b**) Anteroposterior and lateral x-ray of the proximal radius showing a prominent "ground-glass" expanding lesion

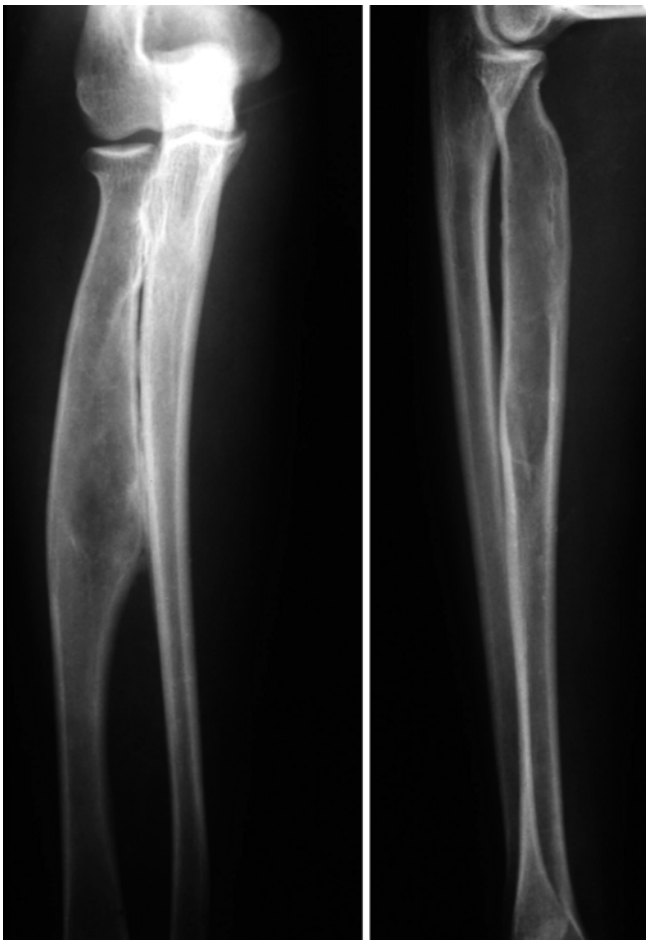


Fig. 53.18 FD. Anteroposterior and lateral x-ray of the radius. Intramedullary lesion with a "ground-glass" appearance

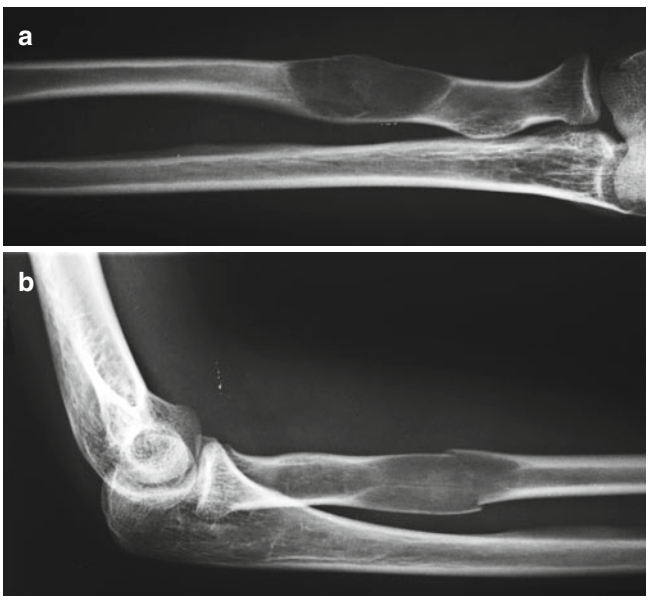


Fig. 53.19 FD. (**a**, **b**) Anteroposterior and lateral x-ray of the radius showing a pathologic fracture

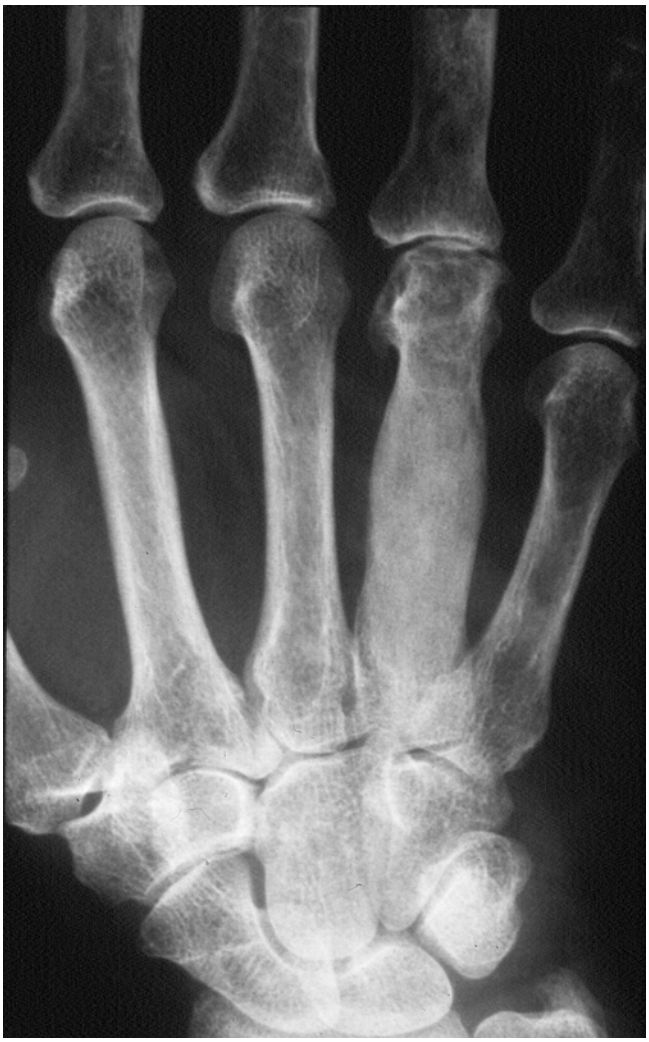


Fig. 53.20 FD. IV metatarsal bone. Unusual location for monostotic FD

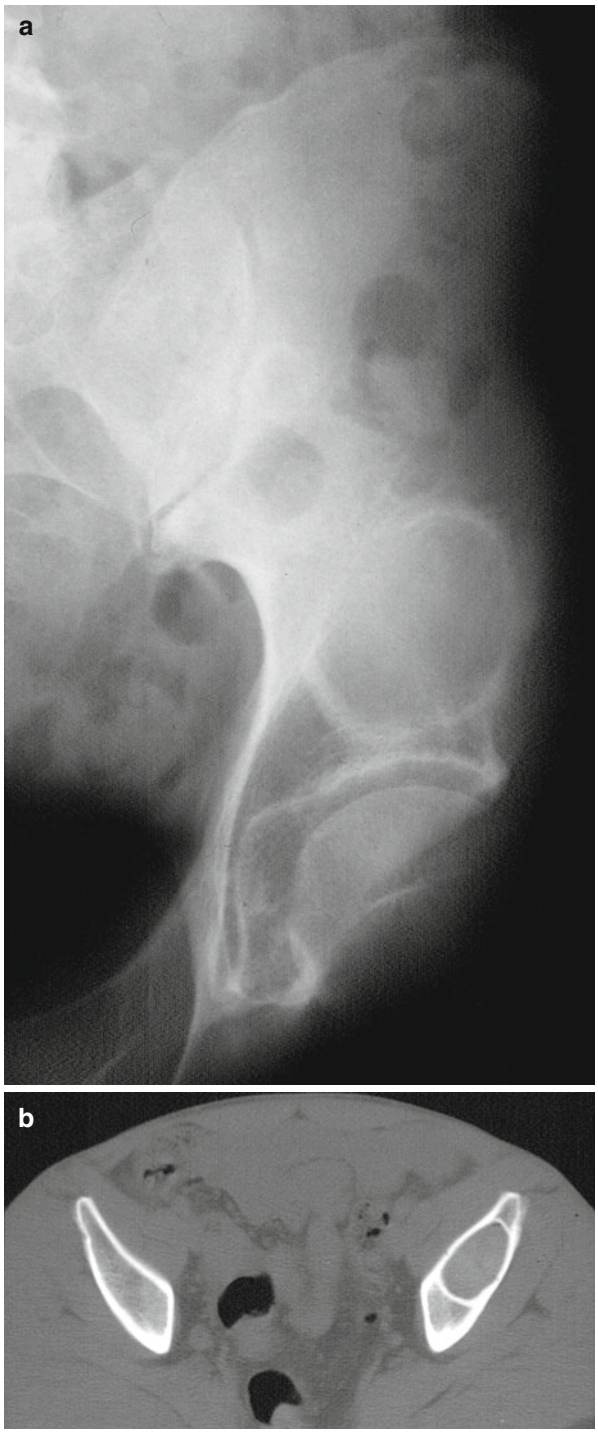


Fig. 53.21 FD. (**a**) Anteroposterior x-ray of the left hip showing a well-defined lesion in the pelvis. (**b**) CT scan illustrates a sharply marginated lesion of FD expanding the left iliac bone

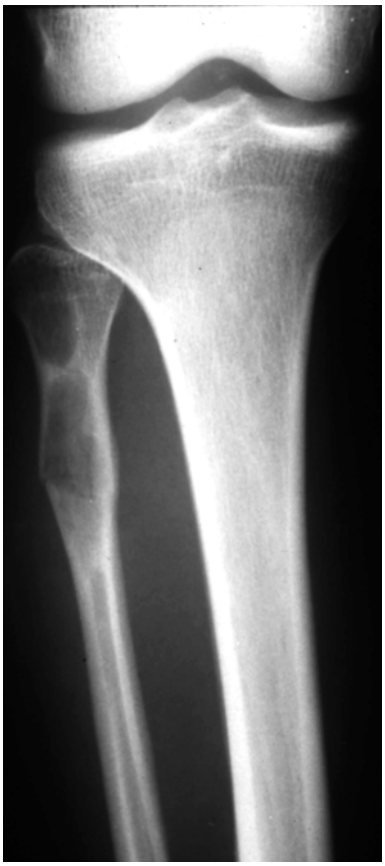


Fig. 53.22 FD. Radiographic features of multifocal FD of the fibula

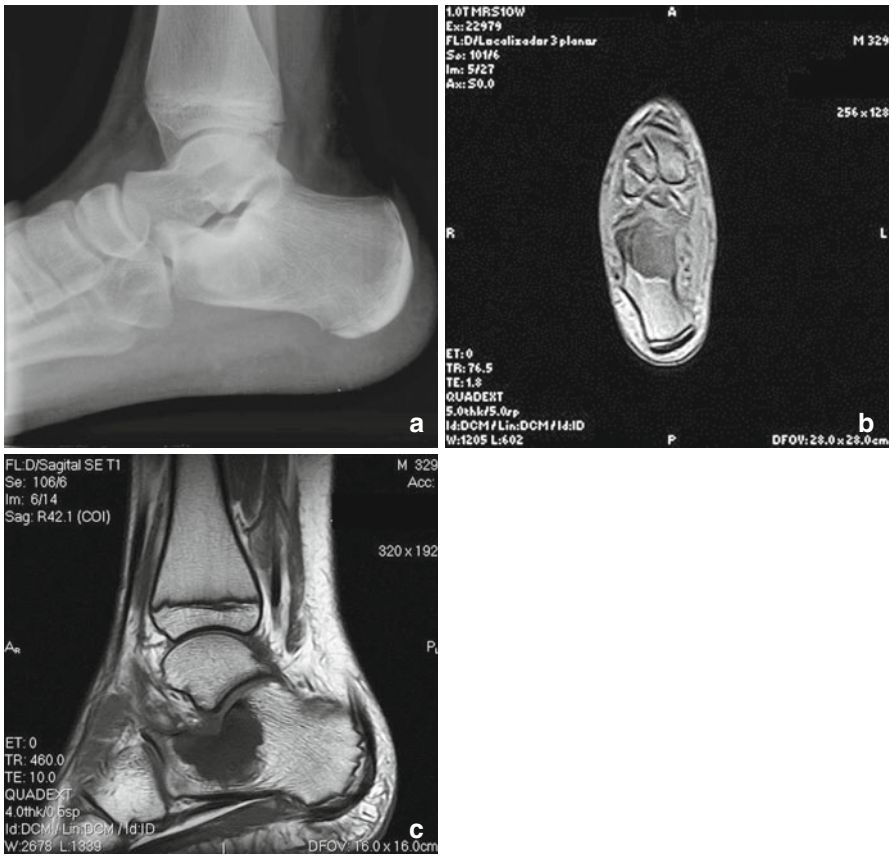


Fig. 53.23 FD. (**a**) Lateral x-ray of the calcaneus. (**b**, **c**) Axial and sagittal MRI of the same lesion

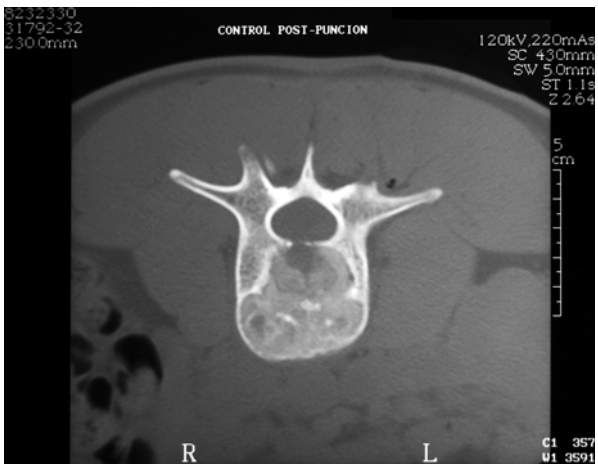


Fig. 53.24 FD. Axial CT shows an expansile lesion of lumbar vertebral body. Unusual location of monostotic FD

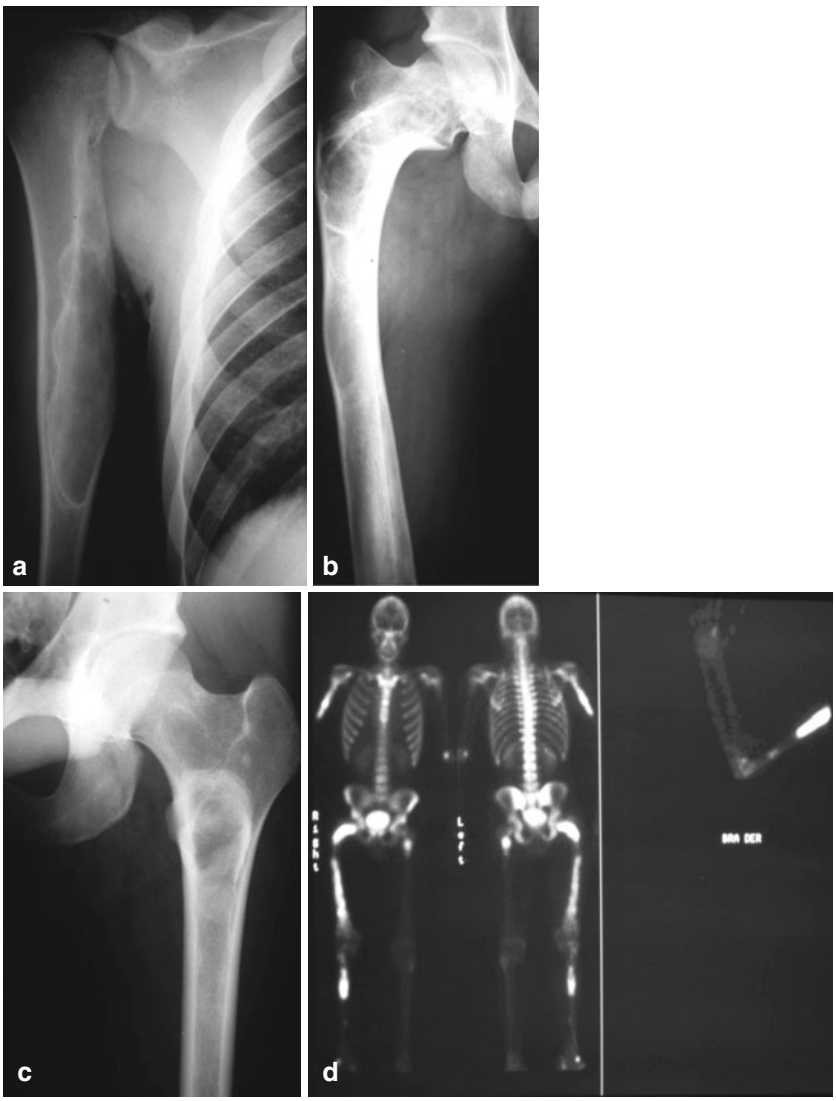


Fig. 53.25 Mazabraud syndrome. (**a**) Humerus. Well-delimited and "ground-glass" lesion typical of FD. (**b**) Diffuse involvement of the right femur. (**c**) Left femur showing the "ground-glass" density of FD. Note the thick ring of surrounding sclerosis. (**d**) Tc99 scan shows sites of polyostotic FD with dense uptake. (**e**, **f** and **g**) T1- and T2-weighted MRI show well-circumscribed soft-tissue mass and gross specimen of soft-tissue myxoma

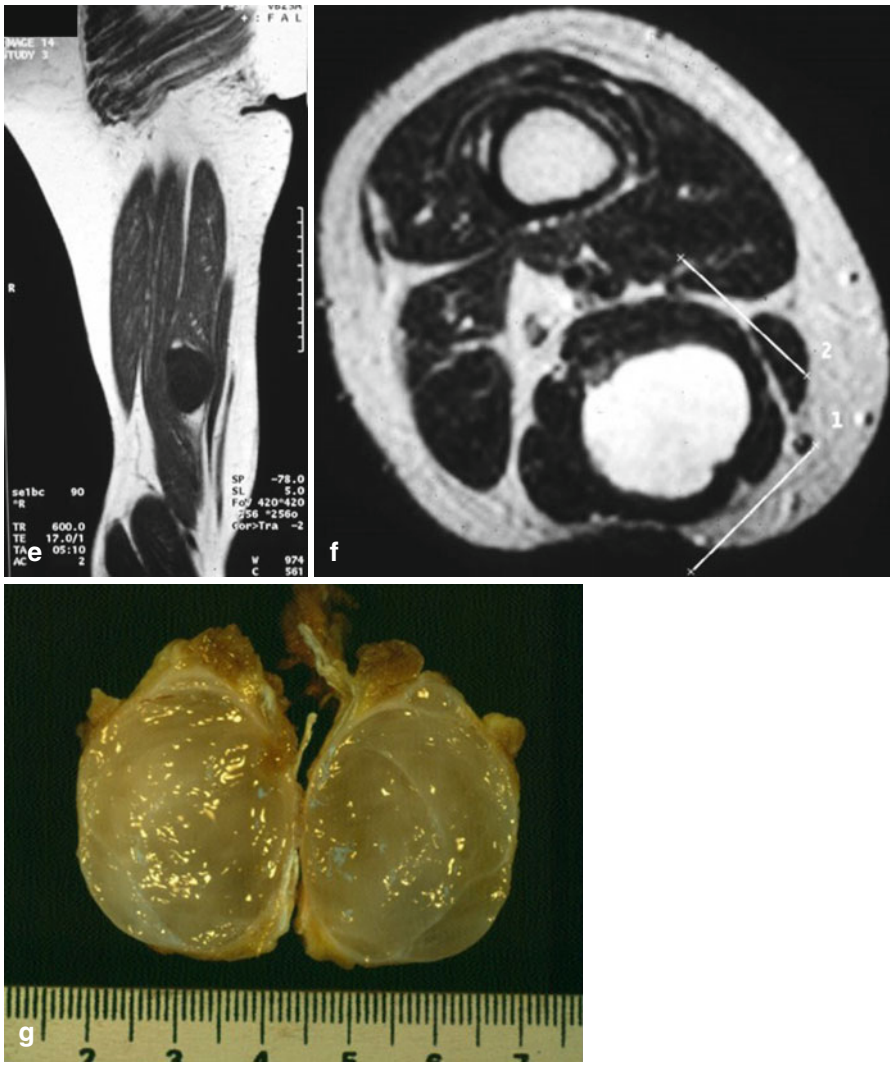


Fig. 53.25 (continued)

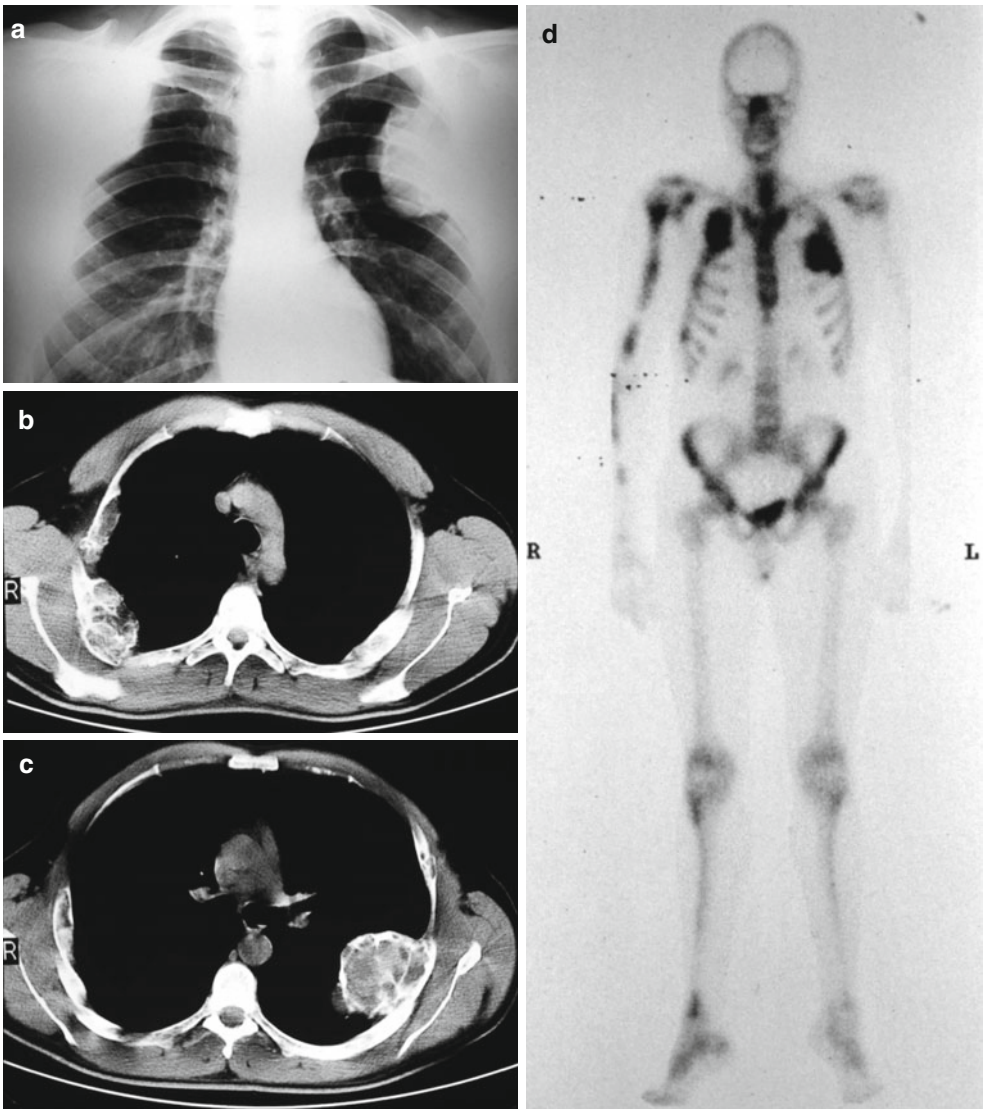


Fig. 53.26 Polyostotic FD. Involvement of ribs in both sides of the chest wall. (**a**) X-ray of chest wall. (**b**, **c**) Axial CT scan of FD protuberans of the ribs showing a large, heterogeneous mass arising from the posterior and lateral aspect of both sides (**b** right and **c** left), bulging into the pleural space. (**d**) Hot spot with the multiple uptake of the radioisotope

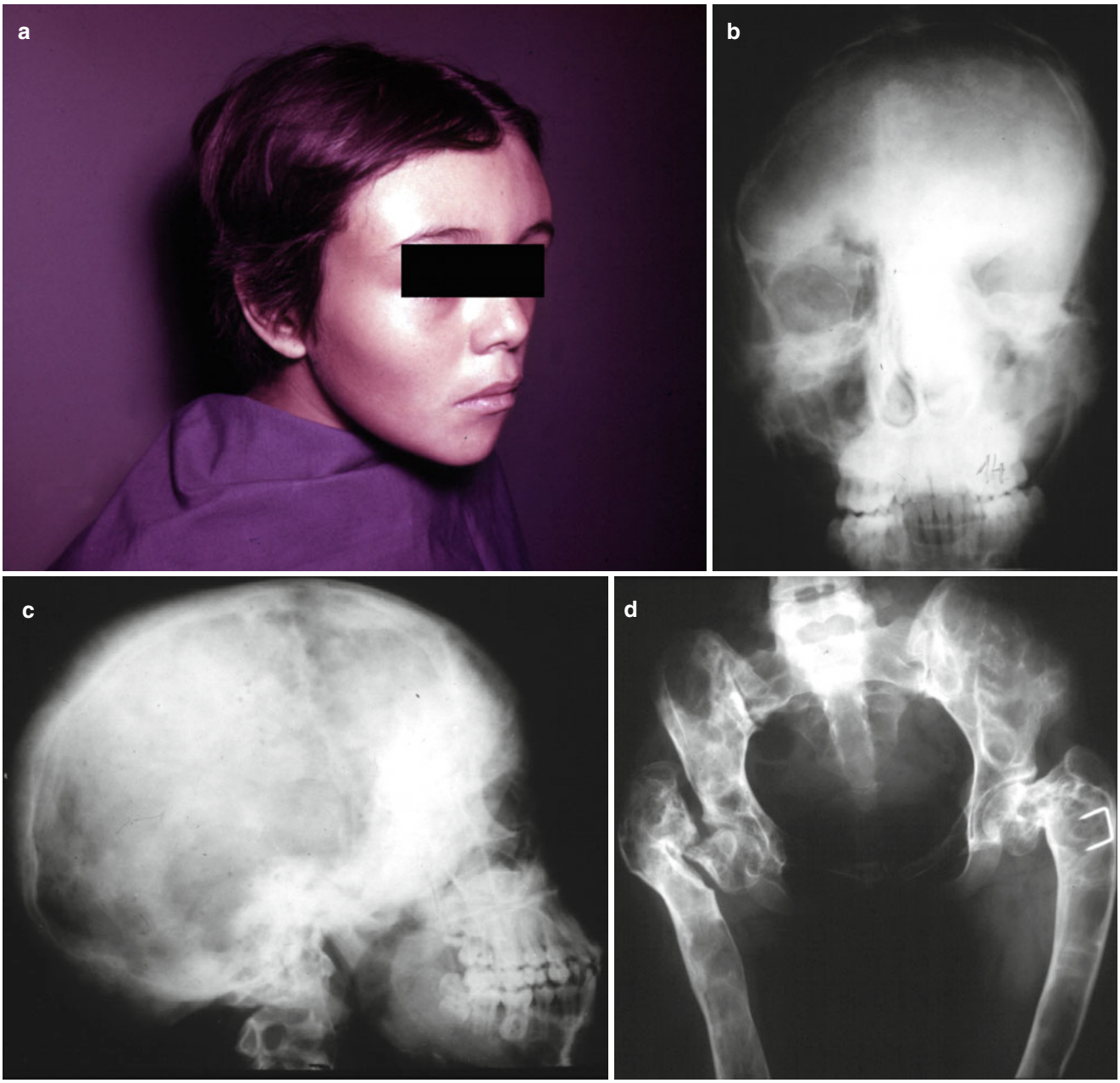


Fig. 53.27 (**a**) Facial deformity in polyostotic FD. (**b**, **c**) Anteroposterior and lateral x-ray. Extensive involvement of skull in polyostotic FD with McCune-Albright syndrome. (**d**) Anteroposterior x-ray of the pelvis and the proximal femur showing medullary lesions of both femurs with "shepherd's crook" deformity. (**e**) X-ray of the lower leg of the same patient showing extensive intramedullary involvement of the tibia and fibula, with thinning and scalloping of the cortex

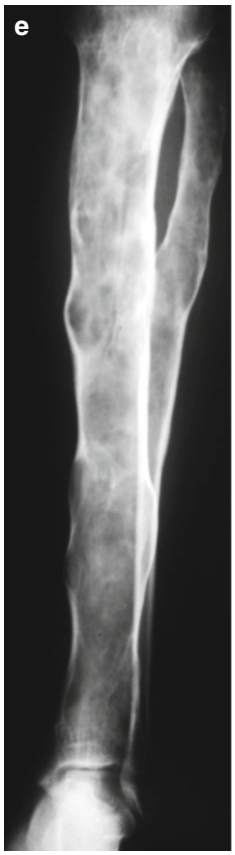


Fig. 53.27 (continued)

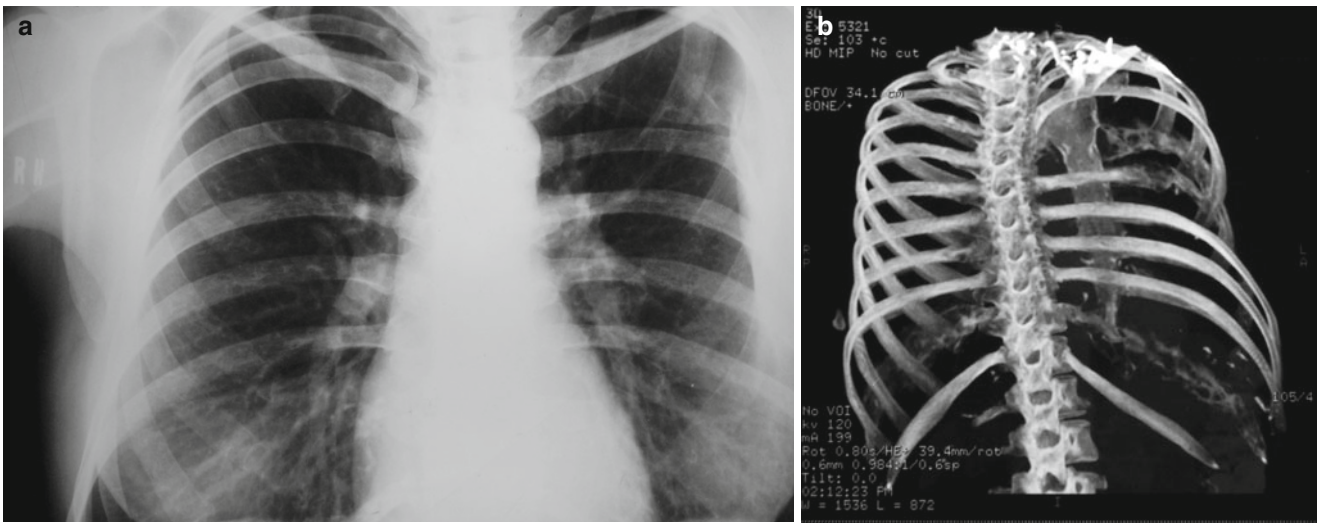


Fig. 53.28 Polyostotic FD. (**a**) X-ray of the chest wall. (**b**) 3D-reconstructed CT scan of the chest wall showing deformity of the ribs

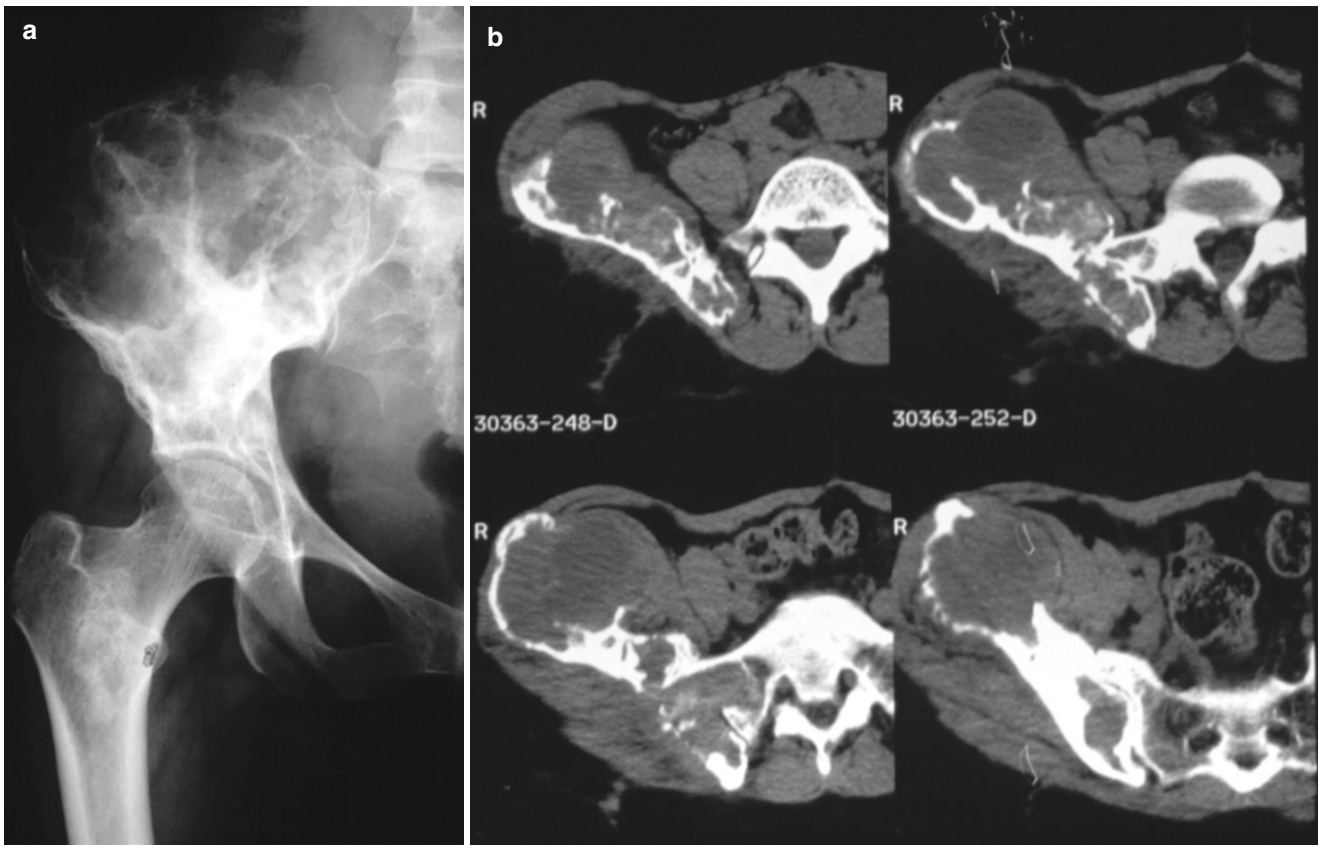


Fig. 53.29 (**a**) X-ray and (**b**) CT scan show secondary aneurysmal bone cyst in association with FD, producing an aggressive appearance in the image, with cortical destruction and soft-tissue extension

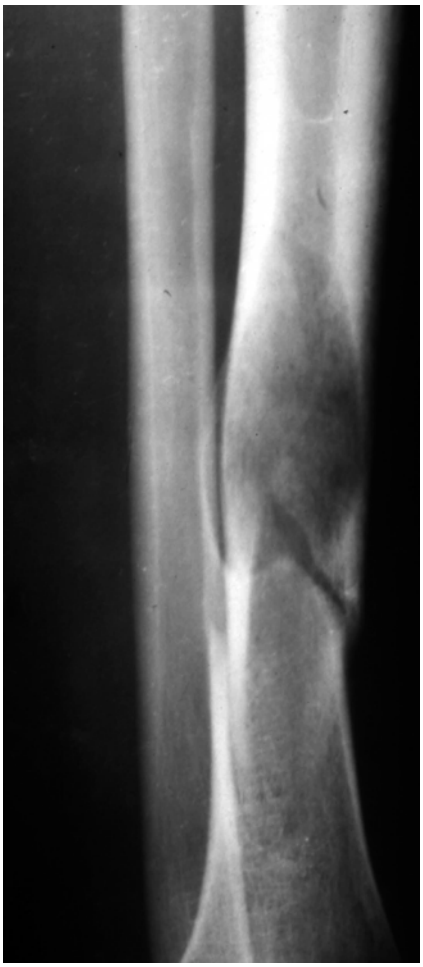


Fig. 53.30 Pathologic fracture in a patient with FD in diaphysis of the tibia

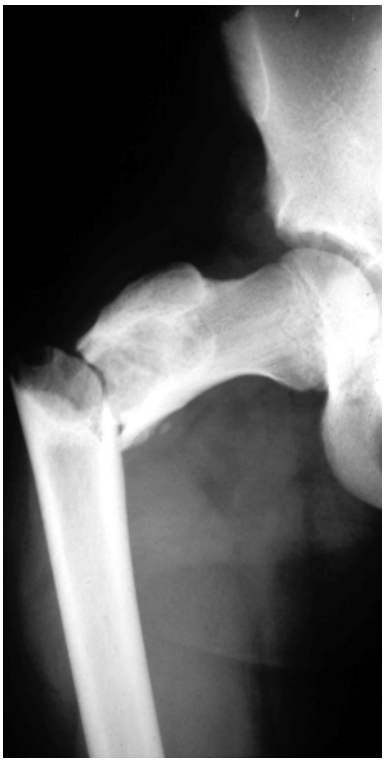


Fig. 53.31 Pathologic fracture of FD in the proximal femur

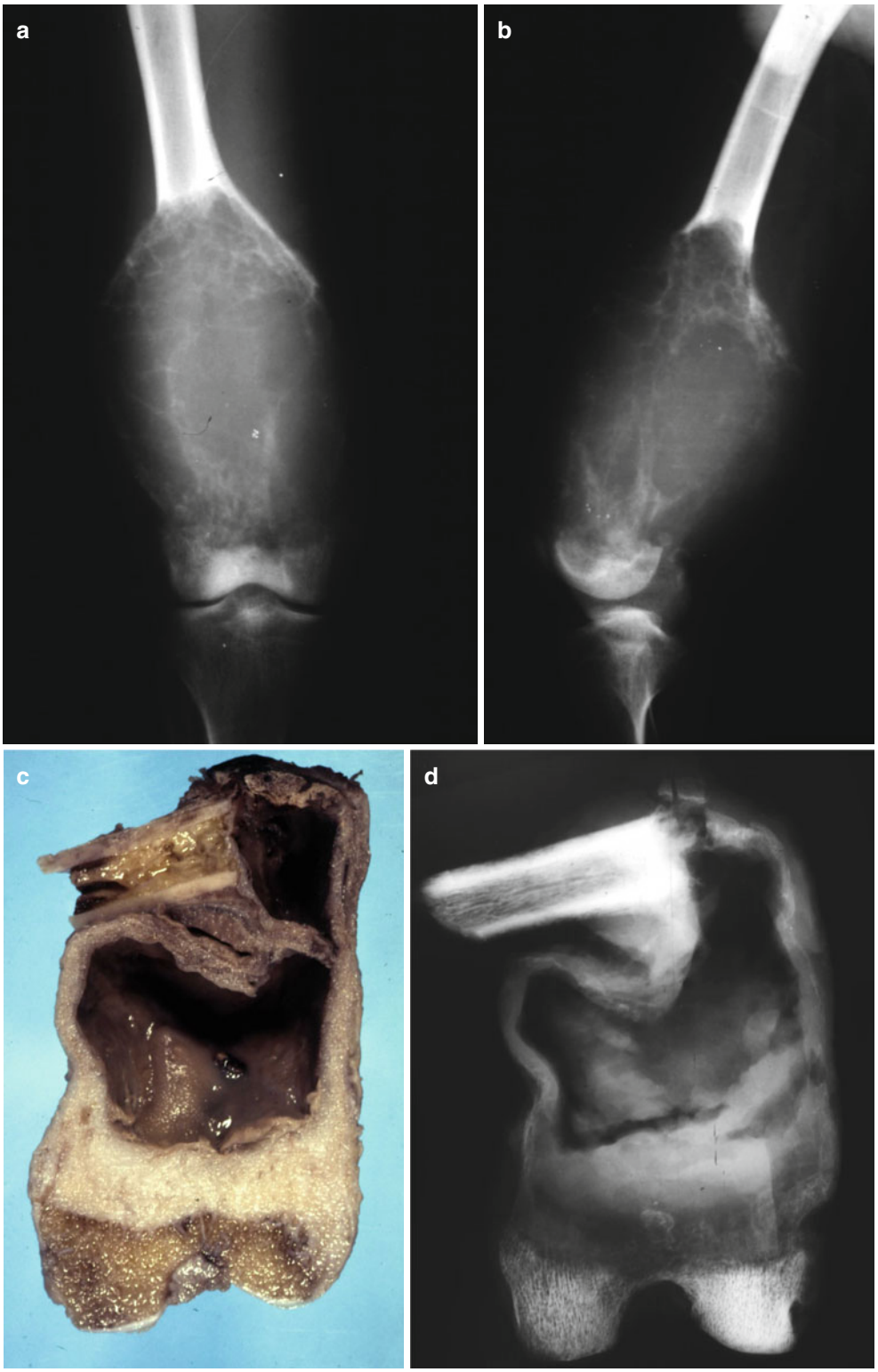


Fig. 53.32 Sarcoma in FD. Distal femur. (**a**, **b**) Anteroposterior and lateral radiographic image, showing a large, expansive, and lytic lesion. (**c**) Gross specimen. (**d**) X-ray of gross specimen

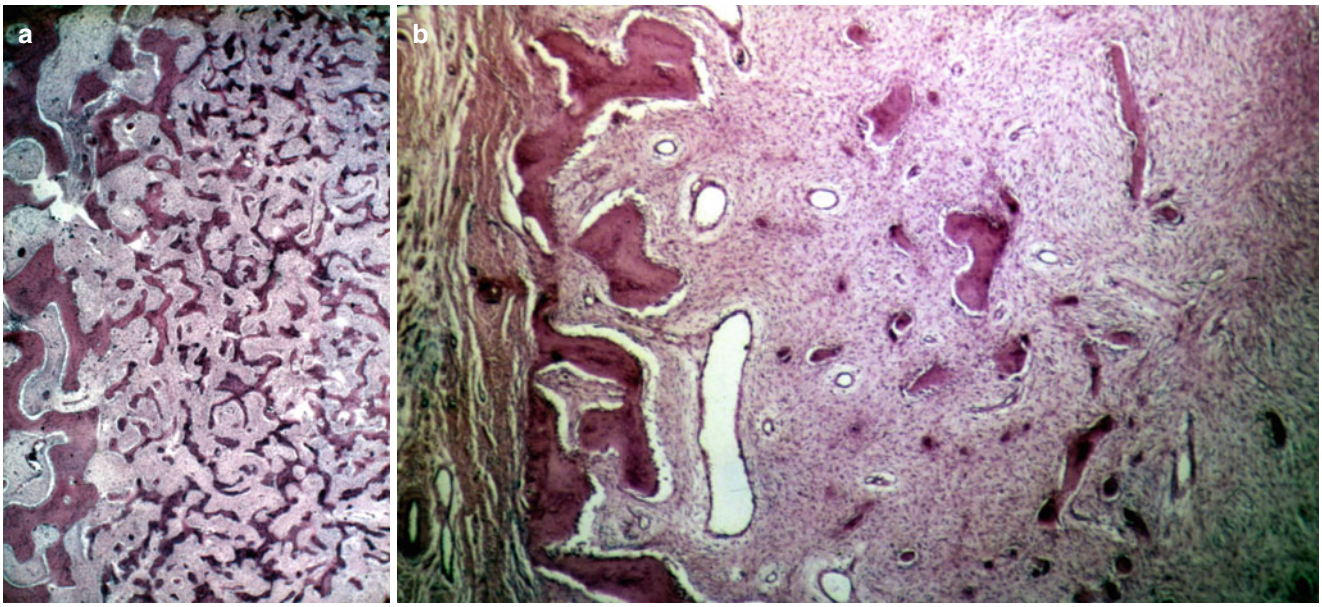


Fig. 53.33 (**a**) Low-power magnification of FD showing its well-circumscribed margin of growth composed of a spindle cell stroma with numerous irregular trabeculae. (**b**) Periphery of the lesion

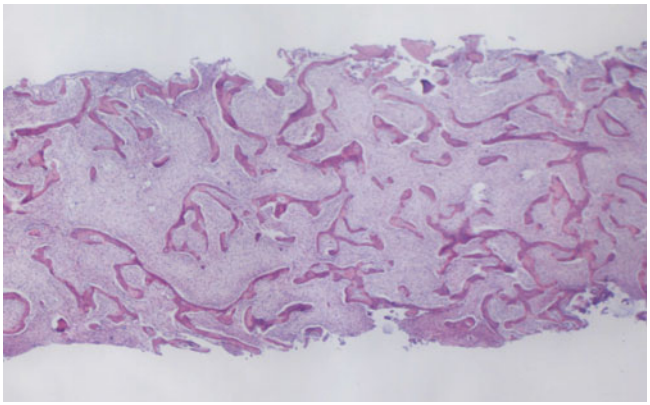


Fig. 53.34 Microphotograph of core needle biopsy

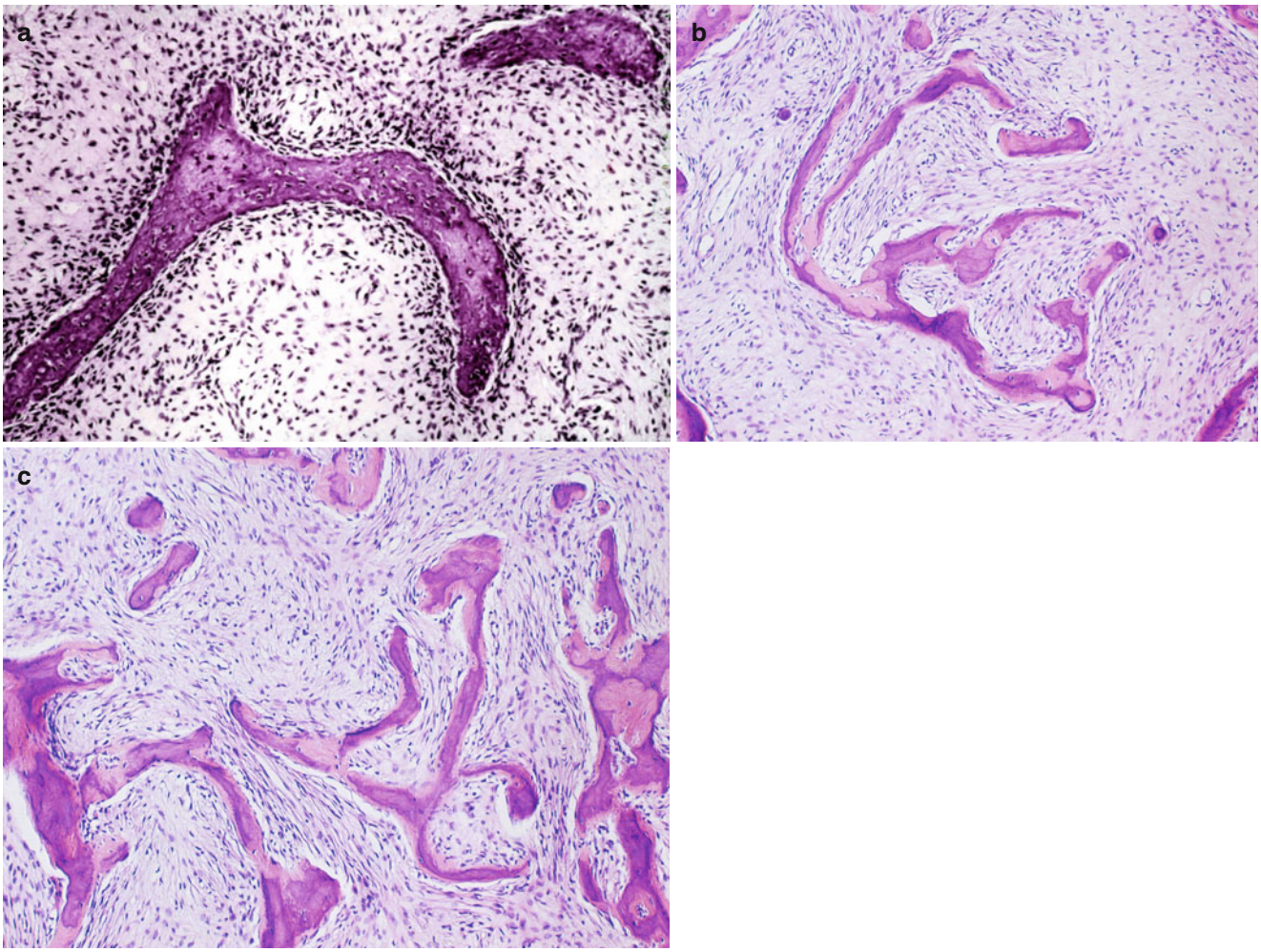


Fig. 53.35 (**a**–**c**) High magnification of dysplasia lesion composed of bony trabeculae of different size arranged in a haphazard, nonfunctional fashion, so-called "Chinese characters" shapes

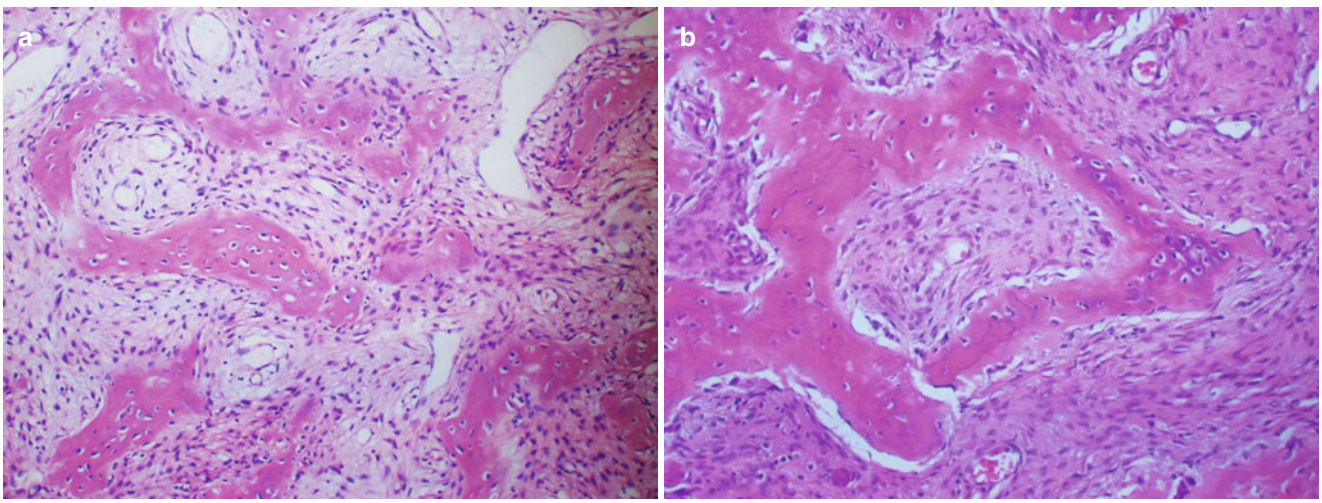


Fig. 53.36 (**a**) Microphotograph of the woven bone with the typical curvilinear, in C, U, and sometimes circumferential shape. Note the absence of active osteoblastic rim. (**b**) High-power magnification showing the fibroblastic stroma arranged in a whorling pattern

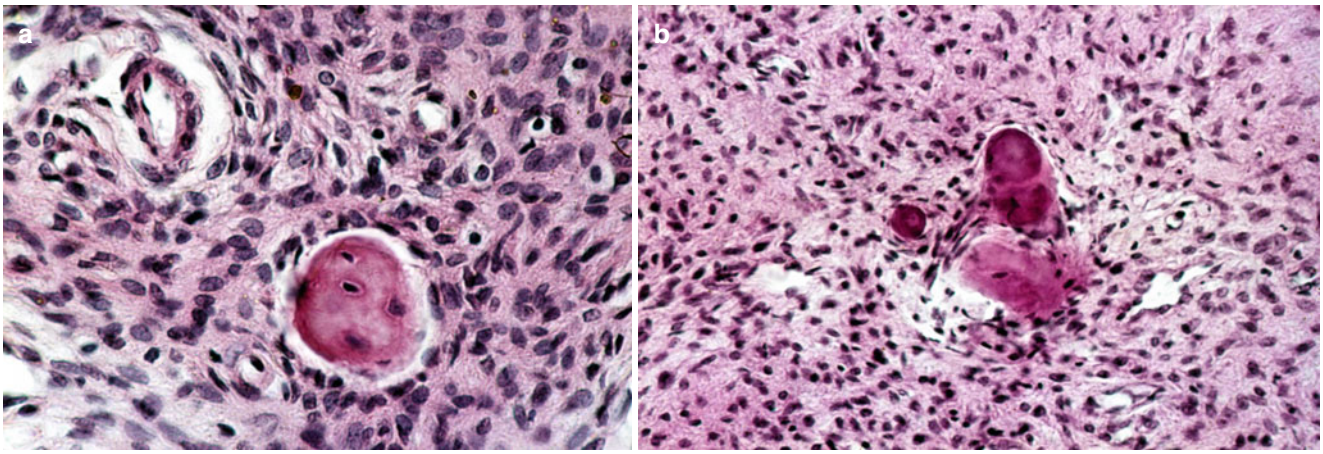


Fig. 53.37 (**a**) Axial section of a dysplastic trabecula. (**b**) Presence of cementoid bodies

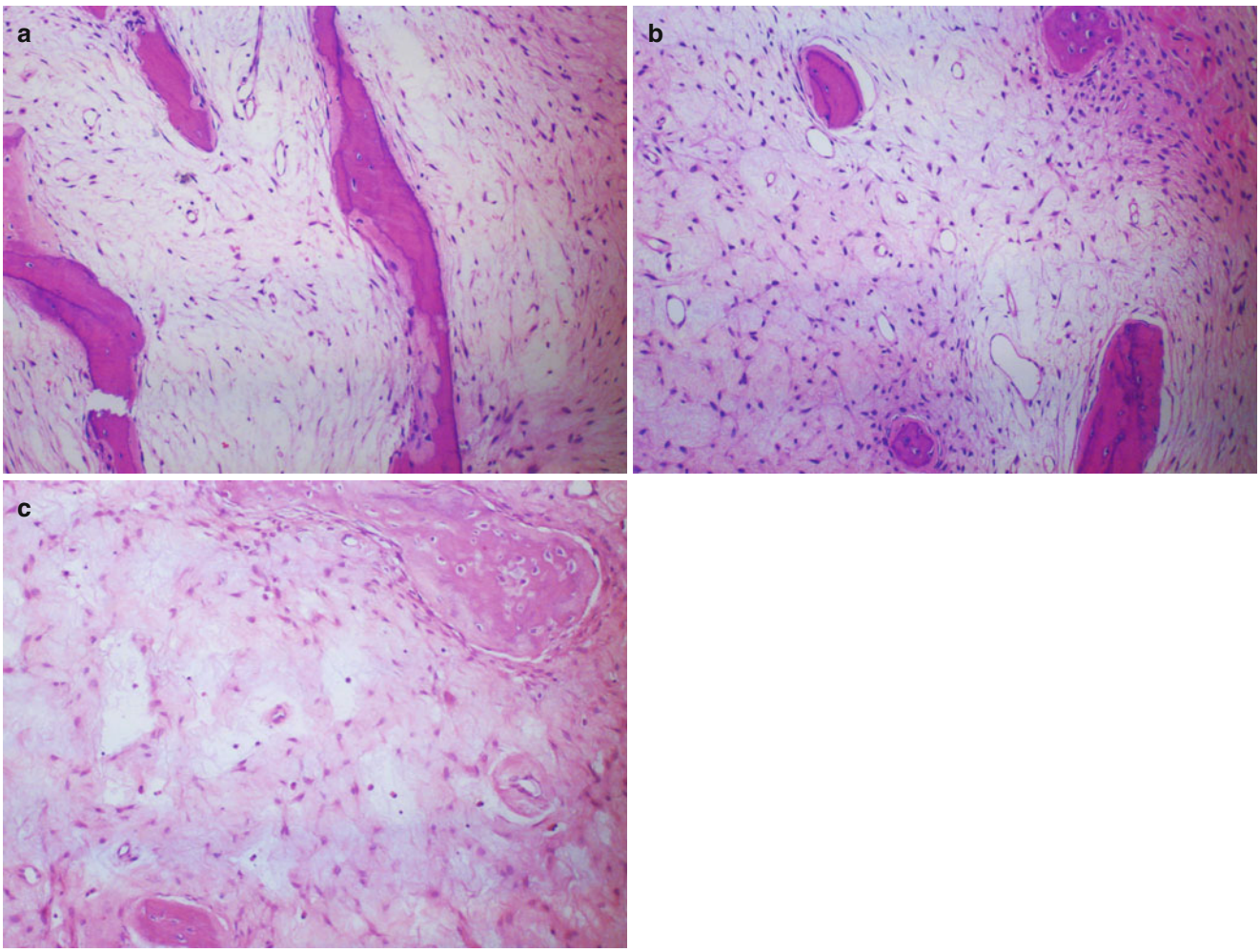


Fig. 53.38 (**a**–**c**) Sometimes the fibroblastic stroma may present myxomatous changes

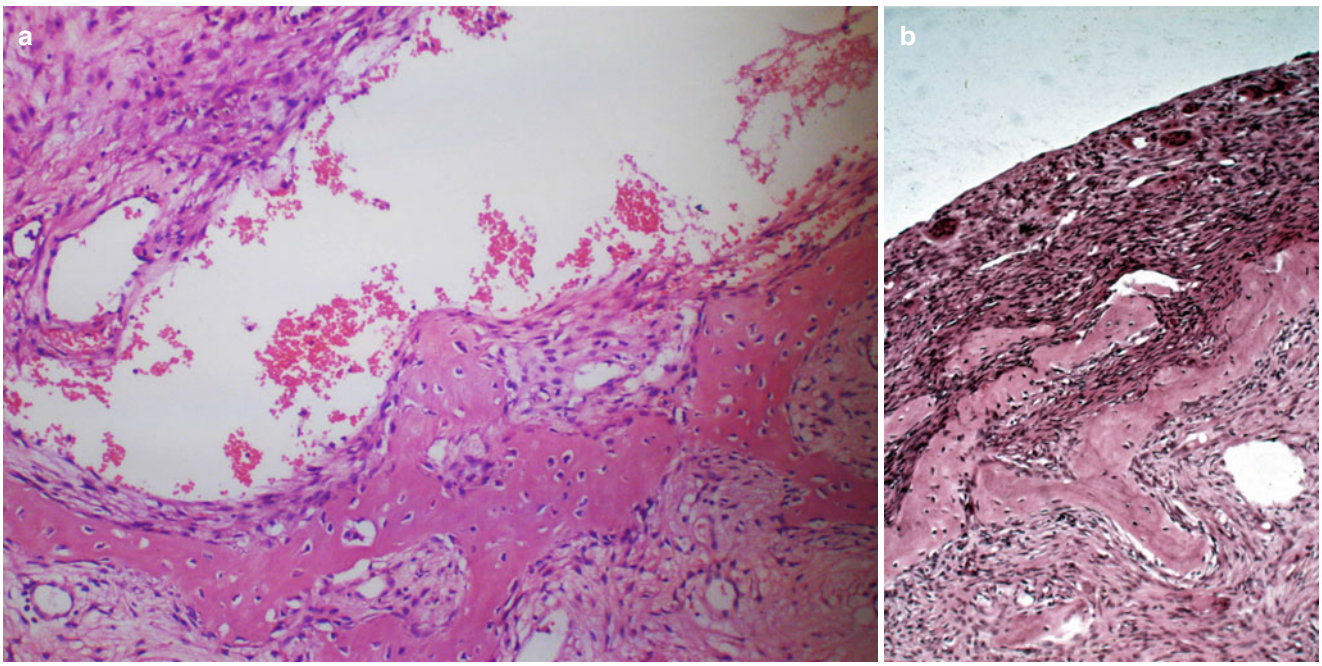


Fig. 53.39 (**a**) Secondary ABC changes in FD. (**b**) The wall of the cyst may be composed of a fibrous component with giant cell

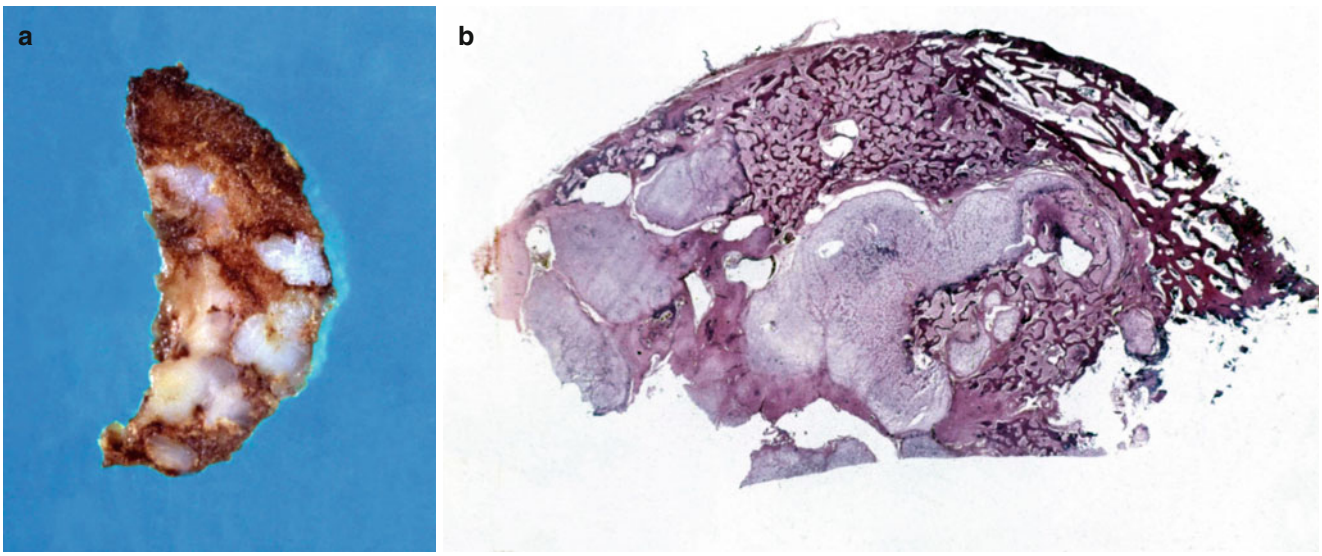


Fig. 53.40 (**a**, **b**) Gross specimen and low-power magnification with cartilaginous component

Recommended Reading

Candelier GA, Glorieux FH, Prud'homme J, St.-Arnaud R. Increased expression of the c-fos proto-oncogene in bone from patients with fibrous dysplasia. N Engl J Med. 1995;332:1546–51.

Harris WH, Dudley HR, Barry RJ. The natural history of fibrous dysplasia. J Bone Joint Surg Am. 1962;44:207–33.

Jee WH, Choi KH, Choe BY, Park JM, Shinn KS. Fibrous dysplasia: MR imaging characteristics with radiopathologic correlation. AJR Am J Roentgenol. 1996;167:1523–7.

Ruggeri P, Sim FH, Bond JR, Unni KK. Malignancies in fibrous dysplasia. Cancer. 1994;73:1411–24.

Sissons HA, Steiner GC, Dorfman HD. Calcified spherules in fibro-osseous lesions of bone. Arch Pathol Lab Med. 1993;117:284–90.

Unni KK. Conditions that commonly simulate primary neoplasms of bone. In: Dahlin's bone tumors: general aspects and data on 11,087 cases. 5th ed. Philadelphia: Lippincott-Raven; 1996. p. 369.

Weinstein LS, Shenker A, Gejman PV, Merino MJ, Friedman E, Spiegel AM. Activating mutations of the stimulatory G protein in the McCune-Albright syndrome. N Engl J Med. 1991;325:1688–95.