Exophytic fibrous dysplasia of the rib

George Hermann MD and Roberto Garcia MD
Department of Radiology and Pathology Mount Sinai Medical Center New York NY. USA

Summary: We report a case of a 56-year-old male who presented with an exophytic variant of fibrous dysplasia of the rib. The lesion appeared as a calcified mass that involved the posterior aspect of the left ninth rib. Fibrous dysplasia is a common benign fibro-osseous lesion that arises in the medullary cavity of the bone. This rare presentation of fibrous dysplasia should be differentiated from cartilaginous and non-cartilaginous masses involving the surface of the bone. The differential diagnosis will be discussed.

EXOPHYTICUS FIBROSUS DYSPLASIA BORDÁN

A szerzők egy 56 éves férfibeteg esetét ismertetik, aki egyik bordáján kialakult, kifelé növekvő fibrosus dysplasiával jelentkezett. Az elváltozás a bal oldali IX. borda hátsó felszínére terjedő, elmeszesedett terimeként ábrázolódott. A fibrosus dysplasia gyakori, jóindulatú, a csont velőüregében kialakuló fibro-osseális elváltozás. Ehelyütt bemutatott, ritka formáját a csontfelszínen képződő porcos, illetve nem porcos szövetszaporulatoktól kell megkülönböztetni. A közlemény az elkülönítő kórismézést is tárgyalja.

Fibrous dysplasia (FD) is a common benign intramedullary lesion of the skeleton characterized by proliferation of fibro-osseous tissue. It is viewed as a developmental abnormality that is typically seen in adolescents and young adults. The lesion is usually asymptomatic and many times discovered as an incidental finding. Exophytic FD is an extremely rare variant that has to be differentiated from benign and malignant surface lesions of bone.

CASE PRESENTATION

A 56-year-old male patient presented to the psychiatric clinic with suicidal ideation.

He had complained of left back pain. His past medical history included hypertension, hyperlipidemia, asthma and posttraumatic stress disorder following World Trade Center exposure. Physical examination failed to demonstrate any abnormality. There was no tenderness over the spine and rib cage. The laboratory findings were unremarkable except for high cholesterol level.

He underwent a chest X-ray followed by a CT scan of the thorax, which revealed a large mass at the posterior aspect of the left ninth rib. The rib demonstrated extensive cortical erosion surrounded by amorphous calcifications. The calcified mass produced displacement of the left erector spinae muscle and involved the costovertebral junction. There were punctate calcifications in the mass (Fig 1). The findings could be compatible with a tumorous process, probably neoplastic.

The differential diagnosis includes a fibro-osseous lesion, fibrous dysplasia or less likely a metastasis. A biopsy was performed followed by resection of the lesion.

PATHOLOGY

Macroscopically, the resected specimen consists of a segment of rib with a well-circumscribed, hard mass exhibiting exophytic growth from within the medullary cavity. The cut surface of the mass is homogeneous, tan-white, hard and ivory-like (Fig 2). Microscopically, the mass is composed of cytologically bland, moderately cellular fibrous tissue with irregular, curvilinear and slender trabeculae of woven bone (Fig 3). Osteoblastic rimming of the bone trabeculae is minimal to absent. Mild focal nuclear atypia is present in the spindle cells of the fibrous tissue component but no mitotic figures are identified. No invasion of the adjacent soft tissue is seen. The histological features are diagnostic of FD.

Based on a Lecture at the International Skeletal Society, Rome, Italy, September 13, 2012. E-mail: george.hermann@mountsinai.org

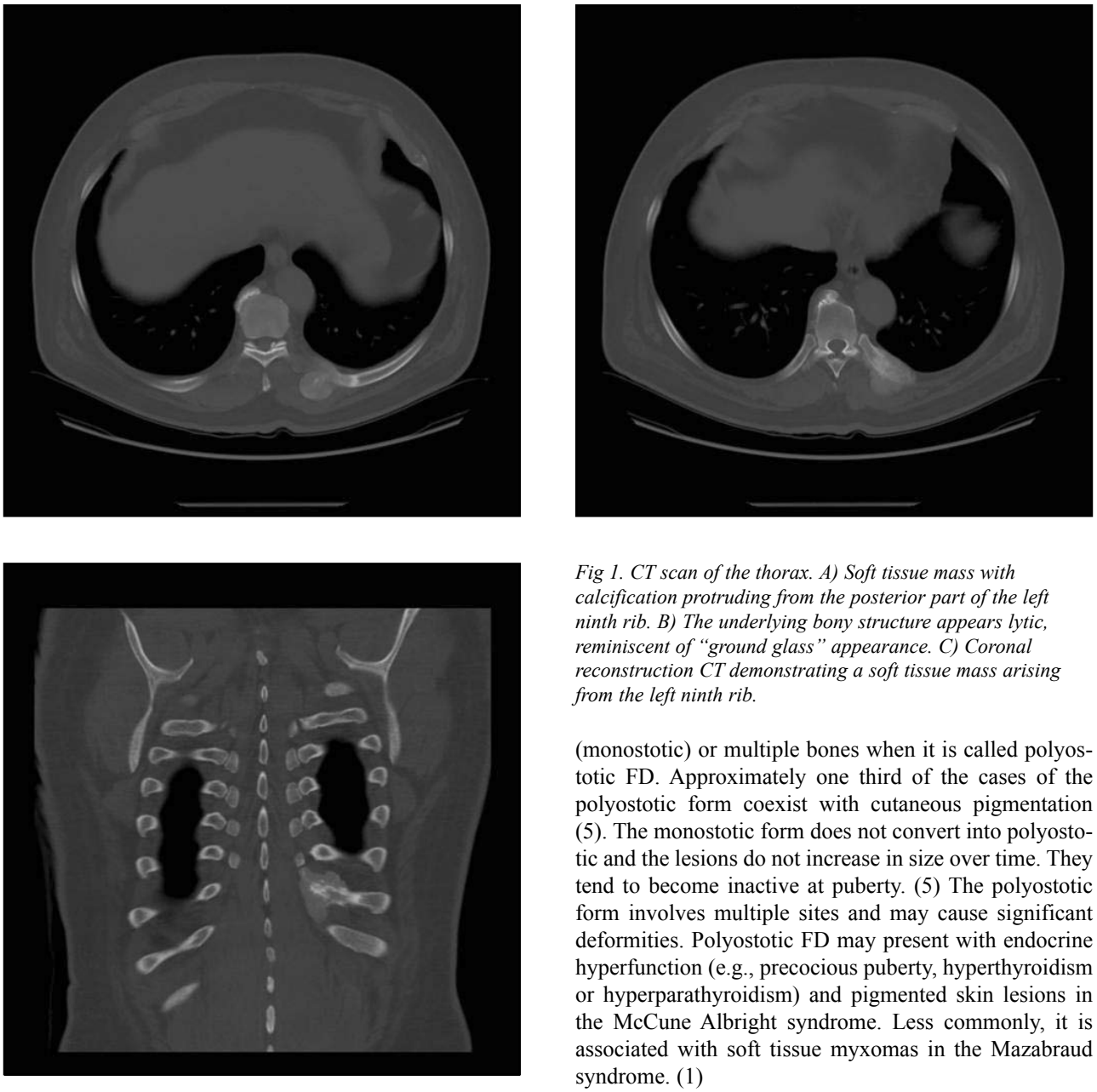

Fig 1. CT scan of the thorax. A) Soft tissue mass with calcification protruding from the posterior part of the left ninth rib. B) The underlying bony structure appears lytic, reminiscent of "ground glass" appearance. C) Coronal reconstruction CT demonstrating a soft tissue mass arising from the left ninth rib.

Discussion

FD is a relatively common, non-inherited disorder in which the normal bone marrow is replaced by fibro-osseous tissue. (1) It makes up approximately 7% of all primary benign bone tumors. (2) The craniofacial bones, femur, tibia, ribs and pelvis are most commonly affected. The lesion usually occurs in the metaphysis or diaphysis of long bones and does not involve the epiphysis. Most cases are diagnosed before the age of 30 years. There is no gender predilection. The lesion is usually asymptomatic and frequently detected incidentally. FD may be complicated with pathological fracture. Malignant degeneration is extremely rare (less than 1%). (3, 4) This benign tumorous process may affect a single bone (monostotic) or multiple bones when it is called polyostotic FD. Approximately one third of the cases of the polyostotic form coexist with cutaneous pigmentation (5). The monostotic form does not convert into polyostotic and the lesions do not increase in size over time. They tend to become inactive at puberty. (5) The polyostotic form involves multiple sites and may cause significant deformities. Polyostotic FD may present with endocrine hyperfunction (e.g., precocious puberty, hyperthyroidism or hyperparathyroidism) and pigmented skin lesions in the McCune Albright syndrome. Less commonly, it is associated with soft tissue myxomas in the Mazabraud syndrome. (1)

In 1938, Lichtenstein described fibro-osseous replacement of bones and called the phenomenon FD of bones. (6) In l942, Lichtenstein and Jaffe reviewed 75 previously reported cases in the literature and they added 15 cases of their own describing FD as a distinct clinical entity. (7).

For many years, FD has been considered a developmental disorder but recent genetic studies have demonstrated clonal chromosomal aberrations that suggest a probable neoplastic nature. (8) Activating mutations in the GNAS1 gene encoding for the alpha subunit of stimulatory G protein have been demonstrated in monostotic and polyostotic FD. (8) This mutation is responsible for the abnormal fibro-osseous proliferation and hyperfunction of endocrine cells and can be detected in peripheral blood samples by Polymerase Chain Reaction (PCR). (2)

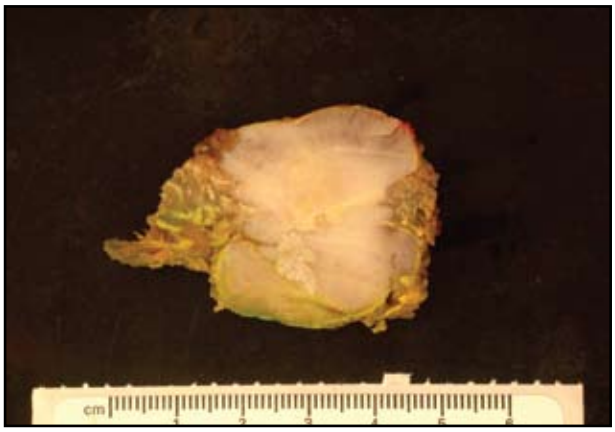

Fig 2. Cross section of resected well-circumscribed rib tumor with a homogeneous, ivory-like, tan-white cut surface.

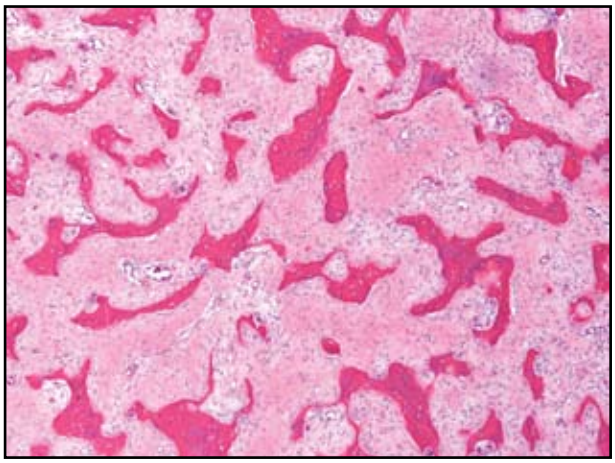

Fig 3A. Photomicrograph of the rib tumor with irregular curvilinear trabeculae of woven bone in cytologically bland fibrous tissue. (hematoxylin and eosin, 4x).

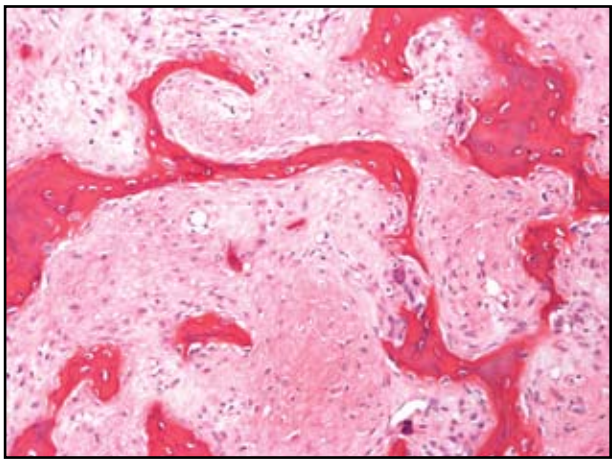

Fig 3B. On higher power, the woven bone trabeculae lack prominent osteoblastic rimming. The fibrous tissue has low cellularity and lacks cytological atypia. A few osteoclasts are present on the right side. (hematoxylin and eosin, 10x).

As we mentioned before, FD is a benign tumorous process that arises and remains confined to the medullary cavity. The involved bone may be expanded and severely deformed. (6) Pathologically, it is characterized by a benign fibrous tissue proliferation with cytologically bland spindle cells and a low mitotic rate, associated to irregular curvilinear trabeculae of woven bone without conspicuous osteoblastic rimming. (8) The presence of cartilaginous tissue has been described in FD and is called by some authors fibrocartilaginous dysplasia. (21)

The exophytic variant of FD is extremely rare. It may mimic an exostosis and should be differentiated from various types of lesions that arise on the surface of the bone.

In 1994 Dorfman et al were the first to describe two cases where the masses extended out of the surface of a rib and proximal tibia respectively. (9) They called it an exophytic variant of FD, or FD protuberans and considered it to be analogous to enchondroma protuberans, which was also called eccentric enchondroma. Hamadani et al. in 2006 reported a case where the lesion extended from the second rib in McCune Albright syndrome (10). Vigorita et al reported FD of the end of a short tubular bone. (11) The location of the lesion is similar to our case and may be considered exophytic FD.

Radiologically, the affected bone is distended due to replacement of the marrow by fibro-osseous tissue. The outer surface of the bone is smooth and the endocortex shows septation. (3).

The medullary cavity shows increased density compatible with ground glass appearance. While the tumorous tissue may enlarge the bone, the tumor is confined to the medullary cavity. (12)

CT can accurately delineate the extent of the lesion. Areas of ossification may appear as high attenuation. (8) On MRI, FD appears hypointense to muscle on T1 weighted images (WI). On T2 WI, it may be either hypo- or hyper-intense with locally increased signal in areas containing fluid. The degree of contrast enhancement depends on the amount of bone trabeculae, cellularity, collagen, cystic and hemorrhagic changes (13)

According to Utz, FD is well vascularized with numerous small vessels in the center and large peripheral sinusoids. (14) In Won's series, central contrast enhancement occurred in 73 % and peripheral rim enhancement in 27%. (13)

Protuberant fibro-osseous lesions of the temporal bone have been described in the pathology literature by Selesnick et al in l999 (15), and recently by Fung et al (16, 17).

Radiologically, these abnormalities appear as calcified masses that must be differentiated from other surface lesions such as osteochondroma, enchondroma protuberans, surface osteoma and parosteal osteosarcoma. (18, 19, 20)

CONCLUSION

We presented a rare feature of FD, "exophytic fibrous dysplasia" in a 56 years old male patient. His clinical symptoms were vague pain over the left upper thorax. The lesion should be differentiated from various types of surface lesions of the bone, both benign and malignant.

The authors declare that they have no conflict of interest.

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