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LETTER TO THE EDITOR

Shepherd's crook deformity: Do you recognize this sign?

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Fibrous dysplasia (FD) is a genetic skeletal disorder caused by a sporadic mutation in the α subunit of the G stimulatory protein, leading to the replacement of normal bone and marrow by poorly organized fibro-osseous tissue.[1] The disease may be localized to a single bone (monostotic FD) or involve multiple bones (polyostotic FD).[2] McCune-Albright and Mazabraud syndromes are two rare forms of the disease, respectively associated with endocrine disorders and intramuscular myxomas.[3] Polyostotic FD most commonly affects the skull, mandible, pelvic bones, and femur.[3] Sometimes severe skeletal deformities can be seen, the most characteristic being the shepherd's crook deformity.[3]

A 36-year-old woman was evaluated for a one-year history of diffuse bone pain. The patient had a history of fractures of the left femur and humerus at pediatric age, after relatively minor traumas, like falls from standing height. On clinical examination, she presented with a left-sided coxa vara and leg length discrepancy. Skin lesions were not evident, and she had no history of precocious puberty. Laboratory findings showed a mild elevation of alkaline phosphatase (155 U/L, normal value <120 U/L) and 25-hydroxy-vitamin D deficiency (9 ng/mL, normal value >30 ng/mL). Other bone-related laboratory tests, including albumin-adjusted serum calcium, phosphate, and parathyroid hormone, and renal profile were normal. Thyroid stimulating hormone and thyroxine had normal values. Osteoarticular X-rays showed multiple expansive radiolucent lesions with a ground-glass appearance and thin bone cortex in both femurs, iliac bones, left fibula, humerus, and right ulna (Figure 1a). Pelvis X-ray revealed a heterogeneous, mixed lytic and sclerotic, expansile lesion involving the neck and proximal diaphysis of the left femur, left acetabulum, and ischiopubic ramus. A shepherd's crook deformity with bowing and varus angulation of the proximal femur was also apparent (Figure 1b). In cranial X-ray, occipital expansive sclerotic lesions were identified. The technetium-99 scintigraphy scan showed increased tracer uptake in multiple locations (Figure 1c). All these findings were compatible with polyostotic FD. The patient was treated with vitamin D supplementation and three cycles of intravenous pamidronate (180 mg, spread over three consecutive days, every six months), which partially relieved symptoms and normalized alkaline phosphatase levels.

Fibrous dysplasia is a rare disorder of striking complexity that can affect all age groups but is usually diagnosed in children or young adults. Currently, the search for medical care only at

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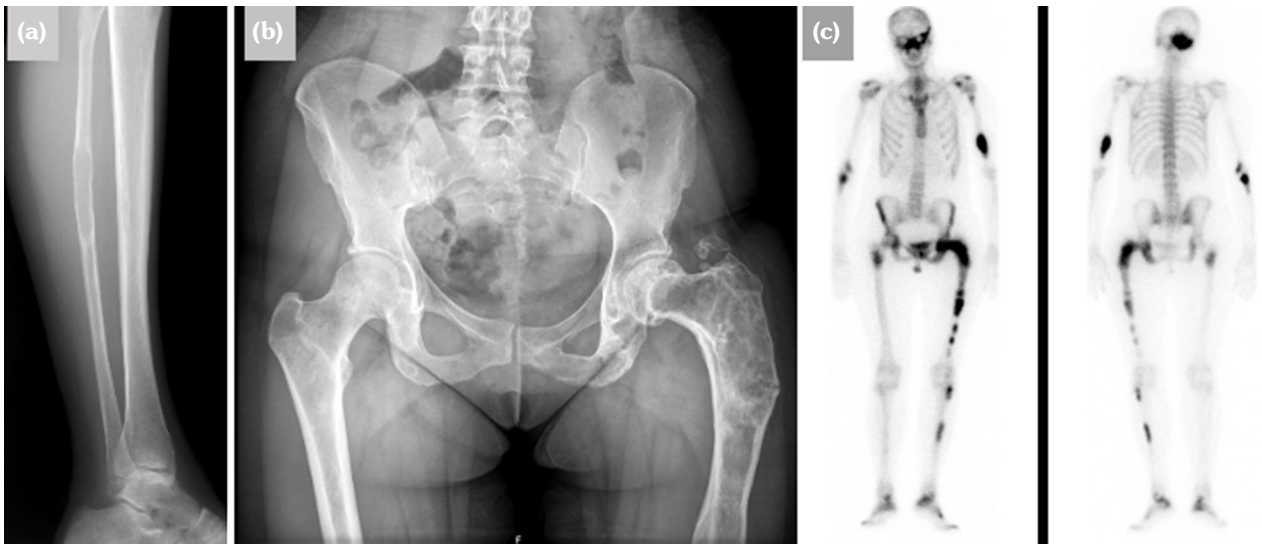

Figure 1. (a) Lateral X-ray of the left leg showing expansive radiolucent lesions and cortical bone thinning of the fibula. **(b)** Anteroposterior X-ray of pelvis with a shepherd's crook deformity of the proximal left femur. **(c)** Technetium-99 scintigraphy scan revealing increased tracer uptake in areas of fibrous dysplasia including skull, humerus, right ulna, both femurs, iliac bones and left fibula.

the age of 36 (with important alterations in the physical examination) is rare, almost questioning the diagnosis. The diagnosis of polyostotic FD can be made based on clinical and imaging findings after a complete staging evaluation for skeletal, endocrine, soft tissue, and dermatologic features, as it was performed in this case.[1,4] The presence of the shepherd´s crook deformity makes the diagnosis straightforward. This term is used to describe a pronounced coxa vara angulation and lateral bowing of the proximal femur.[5] This deformity results in pain, restricted range of motion, limb shortening, leg length discrepancy, gait alteration, and a dramatically increased risk of pathologic proximal femur fractures.[5] Although classically linked with FD, there are some reports of this finding associated with osteogenesis imperfecta and Paget disease.[6] In this case, the clinical history and the radiographic findings exclude these diagnoses. Surgical correction of this deformity remains technically challenging.[7] To this date, there are no known medical therapies capable of altering the course of FD, although antiresorptive therapy may have a role in reducing pain.[1]

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