Polyostotic fibrous dysplasia: a rare cause of pathological fractures in young patients

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DESCRIPTION

Polyostotic fibrous dysplasia (PFD) is a sporadic benign skeletal condition that typically presents with bone pain, deformities and pathological fractures. It preferentially affects long bones. Sarcomatous transformation, most commonly osteosarcoma, occurs in 7% of patients with PFD.¹ Diagnostic challenges exist as radiographic appearances range from cystic foci to pagetoid or sclerotic lesions. Differential diagnoses include juvenile bone cysts, aneurysmal bone cysts and giant cell tumours.²

We describe a case of a 27-year-old woman who had been diagnosed with PFD at the age of 12 years, and who had an extensive history of pathological fractures. On this admission, she presented with inability to weight-bear and pain in her right thigh following a fall from standing height. Plain radiographs showed advanced disease throughout her skeleton. **Figure 1** depicts typical changes including bone expansion, multiple well-circumscribed cystic lesions and thinning of the cortex without periosteal reaction in the pelvis (**figure 1A**), and in the left tibia and fibula (**figure 1B**). **Figure 2** shows the same changes along with

bowing deformity and a pathological displaced transverse fracture of the distal right femur, worsening the pre-existing deformity, immediately after the initial injury (**figure 2A**) and 10 weeks later with callus formation (**figure 2B**).

Surgery in PFD is usually reserved for major deformities or fractures. When making decisions regarding surgical repair, poor remodelling, bone quality and residual angulation in the skeletally immature patient should be considered. Owing to a lack of normal cortical bone, intramedullary internal fixation devices are preferred to plate and screws, as they achieve better fracture stabilisation and may correct existing deformities in this patient group.³

Displaced femoral fractures are difficult to reduce non-operatively. Nevertheless, surgical intervention was deemed inappropriate in our patient because of concerns surrounding high risk of complications such as periprosthetic fractures associated with our patient's extensive loss of cortical thickness. She was instead managed conservatively with skin traction for 8 weeks followed by intensive physiotherapy and has recovered well, returning to baseline at 3 months postinjury.

Figure 1 Plain radiographs illustrating extensive bone expansion, cyst formation and cortical loss in (A) the pelvis, (B) the left tibia/fibular.

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Figure 2 Plain radiographs of the right femur with associated pathological distal transverse fracture (A) immediately after the injury and (B) 10 weeks later with callus formation.

Learning points

- ▶ Polyostotic fibrous dysplasia is a rare complex condition causing bone pain, pathological fractures and progressive deformity in early years.
- ▶ Radiological findings vary and include bowing deformity of long bones, bone expansion, cystic or sclerotic foci and thinning of the cortex.
- ▶ Primary management for major deformity/fracture is surgical, with preference for intramedullary fixation devices, but conservative management may be appropriate in certain cases.

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