Paediatric osteofibrous dysplasia-like adamantinoma with classical radiological findings

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DESCRIPTION

A 16-year-old girl presented with a 1-month history of a painful lump on the right tibia. There was no history of trauma; she had no medical conditions and was physically active. On examination, she had a diffuse bony lump in the middle third of the right tibia with some localised tenderness. There was no neurovascular deficit and no bony masses were present elsewhere.

Plain radiograph of the right leg (figure 1) showed a well-circumscribed, slightly expansile, mixed sclerotic and lytic lesion of the anterior diaphysis of the right tibia. The sclerosis and lysis were seen as a multilocular 'soap bubble' appearance. No aggressive features including periosteal reaction or soft-tissue abnormality were present. A subsequent MRI scan confirmed the presence of an extensive lesion involving the anterior cortex of the midshaft of the right tibia, extending a vertical distance of 20 cm. The lesion appeared confined to the cortical bone, with no evidence of soft-tissue extension or medullary invasion. Within the area of thickened cortex, there were multiple, confluent areas of signal abnormality, showing high signal on T2-weighted and intermediate signal on T1-weighted images (figure 2). Following injection of gadolinium, MRI on T1-weighted images revealed significant enhancement of the confluent areas of signal abnormality within the lesion (figure 3).

Histopathology showed a benign fibro-osseous lesion consisting of bland spindle cells with irregular trabeculae of woven bone (figure 4). Immunohistochemical staining for keratin highlighted small clusters of epithelial cells (figure 5). Given the radiological appearance and histological findings, a diagnosis of osteofibrous dysplasia-like

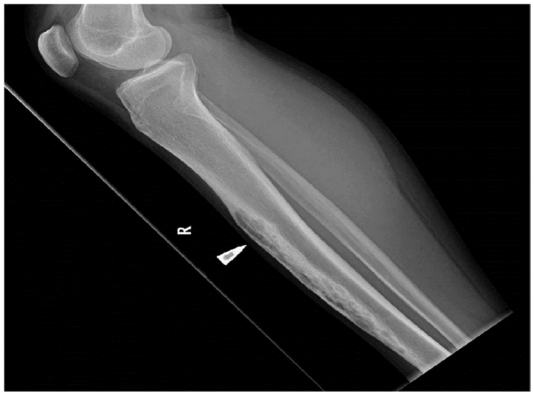

Figure 1 X-ray of the right leg shows an intracortical lesion of the diaphysis of the right tibia with a 'soap bubble' appearance.

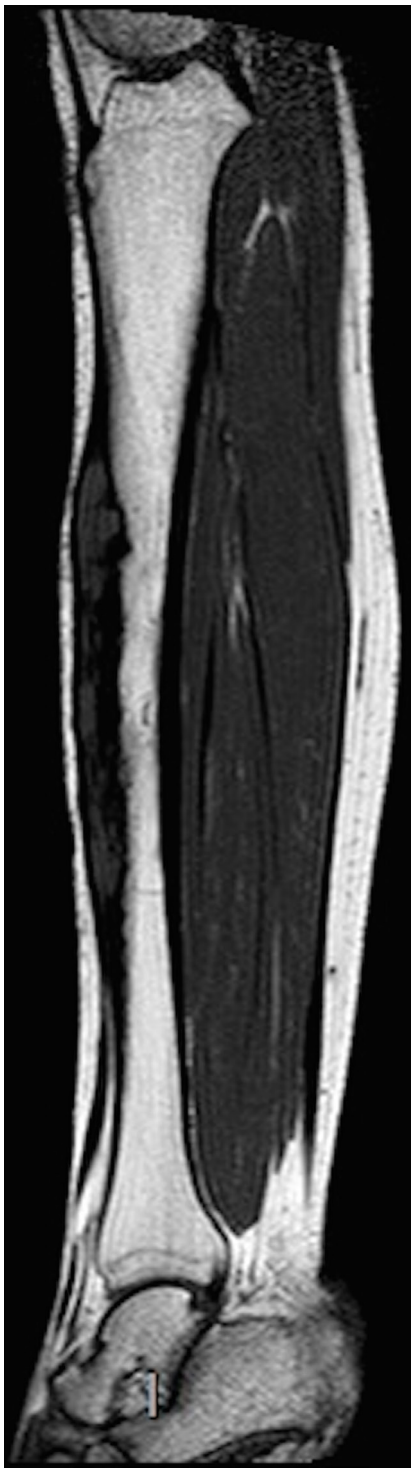

Figure 2 MRI on T1-weighted sequences in sagittal section demonstrating an extensive intracortical lesion involving the anterior cortex of the midshaft of the right tibia.

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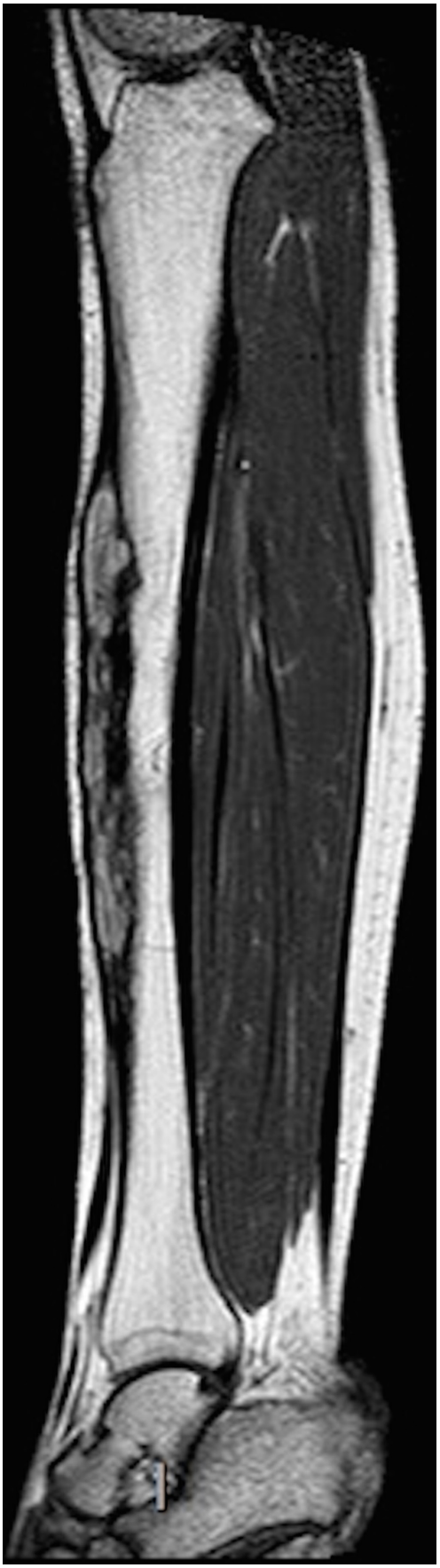

Figure 3 MRI on T1-weighted sequences in sagittal section following gadolinium administration showing significant enhancement of the lesion.

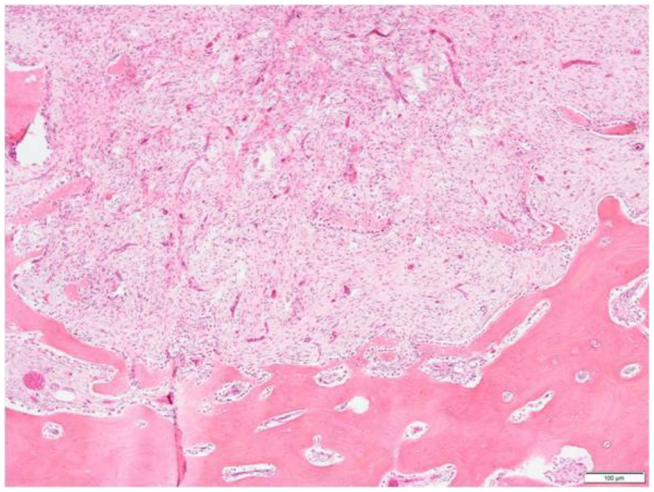

Figure 4 H&E stain showing a fibro-osseous lesion consisting of spindle cells with irregular trabeculae of woven bone.

adamantinoma was established. The patient underwent resection of the border of the tibia and replacement with an allograft. She made a full recovery.

Osteofibrous dysplasia (OFD), OFD-like adamantinoma and classic adamantinoma are postulated to represent a spectrum of morphologically similar disease and cannot be reliably distinguished by imaging studies. On plain radiography, the typical appearance of these lesions is an intracortical, expansive, mixed osteolytic and sclerotic lesion of the diaphysis of the anterior tibia.[1] In addition, circumscribed radiolucent lesions create a 'soap bubble' appearance.[2] This classic radiographic appearance was seen in our patient. MRI studies are crucial for assessing medullary and soft-tissue extension and for surgical planning.[1 2] These lesions demonstrate low-signal intensity on T1-weighted images and high-signal intensity on T2-weighted MRI. However, this appearance is typical of most tumours and is of limited diagnostic value.[3]

OFD is a non-malignant fibro-osseous lesion.[3] Classic adamantinoma is a malignant tumour that represents 0.1%–0.5% of primary bone tumours.[1 2] OFD-like adamantinoma is a histological subtype of adamantinoma, characterised by predominantly benign osteofibrous tissue with inconspicuous nests of malignant cells that

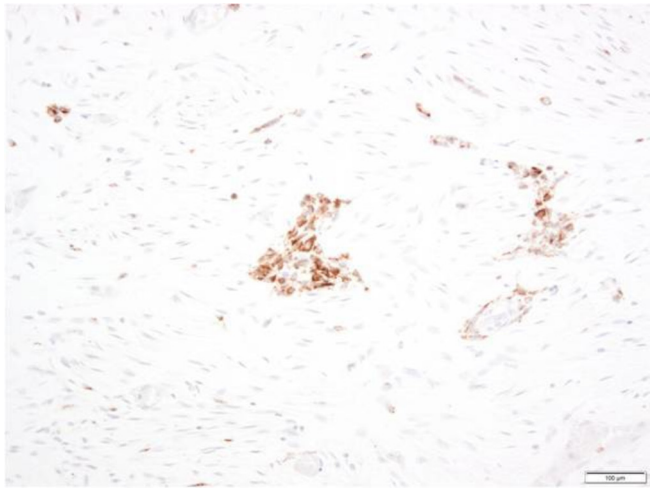

Figure 5 Immunohistochemical stain for keratin showing positivity within the tumour cells.

may only be detected by immunohistochemical analysis.[1 2] There is significant variation in the clinical course of classic and OFD-like adamantinoma, and establishing the correct diagnosis by histological evaluation is vital.[1–3] Classic adamantinoma is an aggressive tumour with metastatic potential, and surgical intervention is essential. OFD-like adamantinoma follows a relatively indolent course and surgery is indicated for extensive tumours, correction of deforming lesions and stabilisation of fractures.[1 2]

Learning points

- Adamantinoma is a rare primary bone tumour that must be considered in a child presenting with a bony mass. The radiological appearances are characteristic and should help narrow the differential diagnosis.
- Histological evaluation is necessary to distinguish between osteofibrous dysplasia OFD (OFD), OFD-like adamantinoma and classic adamantinoma as these lesions are indistinguishable radiologically.
- OFD-like adamantinoma is a variant of adamantinoma. Extensive, deforming and unstable lesions require en bloc tumour resection and limb reconstruction.

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