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Case study

Juvenile juxtacortical chondromyxoid fibroma of bone: a case report

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Summary Conventional intramedullary chondromyxoid fibroma (CMF) is a rare benign tumor of bone; juxtacortical lesions are rarer still, and juxtacortical lesions occurring in children are heretofore essentially unreported. We present a case of such a lesion in a 12-year-old boy. This patient, who was previously healthy, presented with a 1-week history of poorly defined pain and mild swelling in the region of the left proximal tibia. Magnetic resonance imaging and bone scan showed changes most consistent with an aggressive biological process. However, the permanent histologic sections showed a (pseudo) lobular pattern of spindle cells with minimal pleomorphism and other features consistent with CMF. A clonal abnormality was detected in 15% of tumor cells karyotyped, characterized by a break in the long arm of chromosome 6 and a balanced Robertsonian translocation involving chromosomes 14 and 21. The patient has remained well and free of recurrence for more than 4 years. In general, CMF needs to be distinguished from its mimicker low-grade chondrosarcoma, and it must be recognized as occurring on bone surfaces among a wide age range of individuals. Juxtacortical CMF has not proven to be unusually aggressive in adults nor in this child, and marginal (en-block) resection remains the treatment of choice.

1. Introduction

Chondromyxoid fibroma (CMF) of bone is a rare benign primary bone neoplasm accounting for less than 1% of bone tumors [1]. The neoplasm was first described by Jaffe and Lichtenstein [2] as an entity that could be separated from chondrosarcoma and chondroblastoma, having unique radiologic characteristics and clinical course, so that treatment and prognosis were different. The juxtacortical region is an exceedingly unusual location for this tumor, with only a few sporadic cases reported in the English language medical literature, and then in an older aged population, unlike the conventional form [3].

The World Health Organization defines CMF as a benign tumor that is characterized by lobulated areas of spindle-shaped or stellate cells with abundant myxoid or chondroid

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intracellular material. Zones of more cellular tissue, rich in spindle-shaped or rounded cells of different sizes, and separately larger pleomorphic cells are the standardized morphological picture. As noted above, the most important lesion to distinguish it from is a well-differentiated chondrosarcoma, which can have pleomorphic cells, hyperchromatic nuclei, and areas that are largely myxoid. Furthermore, chondrosarcomas may also have a lobular pattern and a relative lack of mitotic figures. In the juxtacortical location, the differential diagnosis would include principally parosteal (juxtacortical) chondroma, juxtacortical chondrosarcoma, and periosteal osteosarcoma. Although the age of this patient is appropriate for the first and third entities, the radiologic appearance is not that of periosteal chondromas, as they typically present as "cup-shaped erosive defects" with punctate calcifications not present in our case and histologically by a relatively pure population of hypercellular hyaline cartilage nodules. In regard to juxtacortical chondrosarcomas, these lesions typically occur in slightly older individuals, are usually larger at presentation, radiologically lack a shell of periosteal new bone, and invade the surrounding tissues while demonstrating histologically malignant features. Periosteal osteosarcomas are typically metadiaphyseal in location, display characteristic radiologic features, and histologically contain tumor osteoid (although in select fields may have overlapping morphological features of cartilage admixed with fibroblastic/mesenchymal cells). We report here a rare case of a juxtacortical CMF in the tibia in a child and discuss the karyotypic analysis, including comparative genomic hybridization of this particular case, and place this in the context of the cytogenetics previously reported in the literature for conventional cases [4].

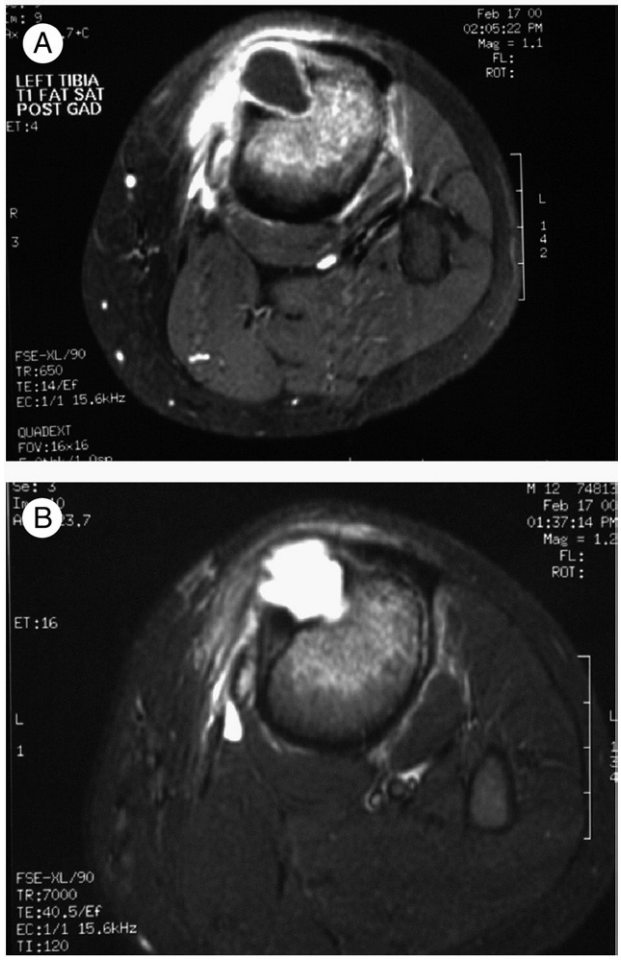

Fig. 1 A, T1 fat saturated postgadolinium MRI in the axial plane. The mass can be seen compressing the underlying bony cortex with a thickened "rind." B, T2-weighted MRI in the axial plane showing the lesional contents having a more intimate less well-defined interaction with the underlying cortex while pushing, rather than invading, the surrounding soft tissues.

2. Case report

The patient was a previously healthy 12-year-old boy with a 1-week history of poorly defined pain in the region of his left proximal tibia with recurrent mild swelling of his "upper shin" present for 3 months. On physical examination, a deformity of the proximal tibial surface was noted on the lateral side. The area was warm, mildly erythematous, and markedly swollen. There was no swelling in the anterior compartment nor evidence of a knee effusion, but the patient had point tenderness over the area just superior to the pes anserinus insertion. His neurovascular examination was normal.

2.1. Radiology

No conventional radiographs were obtained before surgery. The magnetic resonance imaging (MRI) and bone scan were interpreted as those of an aggressive process of the proximal tibia most consistent with a malignancy. Bony remodeling, periosteal change, and marrow edema were evident on MRI (Fig. 1A-B). The bone scan showed increased uptake only in the region of the proximal left tibia.

2.2. Pathology

The child underwent a biopsy with intraoperative consultation, and a diagnosis of "suspicious for sarcoma" was rendered based presumably on the atypia associated with the cartilaginous zones. The histologic material was sent in consultation. Based on this, later revised, diagnosis, a wide marginal excision was performed. Grossly, the original material was described as bony and gelatinous, red-tan to white in color and measuring in aggregate 3 × 1.5 × 1 cm. The reexcision included both tumor, bone, and surrounding soft tissues and skin. Histologically, at lower power, a (pseudo) lobular pattern was appreciated with cellular condensation at the periphery (Fig. 2). At higher magnifications, stellate to