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PRIMARY BONE LYMPHOMA IN A 10-YEAR-OLD BOY

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Primary bone lymphoma has been defined as a solitary lesion in bone, without concomitant involvement of the extra osseous hematopoietic system, with no evidence of extra osseous disease within 6 months of the onset of symptoms. The vast majority of cases are of the large B-cell non-Hodgkin type. They are rare bone tumor. Distinguishing primary bone lymphoma from other bone tumors is important because the former has a better response to therapy and a better prognosis.

Key-words: Bone neoplasms, diagnosis – Children, skeletal system.

Case report

A 10-year old boy was admitted at the emergency department because of right arm pain without traumatic lesion. X-ray shows a lytic diaphyseally centered lesion associated to a periostal reaction (Fig. 1). On the same day, an MRI of the humerus is performed. T1-weighted images show an intramedullary 12 cm long lesion. Post gadolinium T1-weighted sequences reveal a nonhomogeneous enhancement of the lesion. Contrast uptake is only observed in the distal third (Fig. 2). Two biopsies are performed because of the non-homogeneous gadolinium uptake pattern. The first biopsy was obtained in the proximal third and was negative, and a second puncture was made in the distal third and made the diagnosis of B-cell lymphoma. No other localization was found on whole body MR, positron emission tomography and bone marrow analysis 15 months after the onset of pain.

Discussion

Bone mass is a frequent radiological finding on pediatric radiology. There are no specific clinical features. The patient presents with complaints of pain, local swelling, palpable mass or pathologic fractures. Primary lymphoma of bone is a rare malignant condition that accounts for less than 5% of all primary bone tumors (2). The radiographic appearance is variable and non-specific. Primary lymphoma of bone most often involves the diametaphysis of a major long bone and has an aggressive pattern of lytic bone destruction described as a «moth-eaten» or permeative pattern, and an associated soft-tissue mass (3). In young patients, the most frequent diagnoses include Ewing's sarcoma, metastatic neuroblastoma and osteomyelitis (1). Normal-appearing radiographs are reported (2). MRI characteristics are also non-specific findings. T1-weighted images reveal areas of low signal intensity within the marrow. Peritumoral edema and reactive marrow produce high signal intensity on T2-weighted images. Post gadolinium T1-weighted images demonstrate areas of enhancement within the lesion. Soft-tissue masses can be found, with or without extensive cortical destruction (2). Radionuclide bone scans show increased tracer uptake in 98% of patients (3).

Osseous involvement of disseminated malignant lymphoma, which is not uncommon, is indistinguishable from primary bone lymphoma by radiological or MR imaging characteristics. Moreover, patholgical findings cannot make the difference. The diagnosis implies the exclusion of any evidence of nodal or disseminated disease (2), which carries a poorer prognosis. The survey may include an entire body MR imaging, positron emission tomography, or even bone marrow analysis.

Primary lymphoma involving the bone has an excellent prognosis. A combined radiotherapy and chemotherapy treatment leads to better results than single modality treatment, with a 5-year cause-specific survival rate of 95% (4).

The differentiation of residual tumor and treatment-associated changes, including tumor necrosis and granulation tissue, may be challenging on MR images of bone tumors after treatment. FDG PET is the gold-standard for staging and

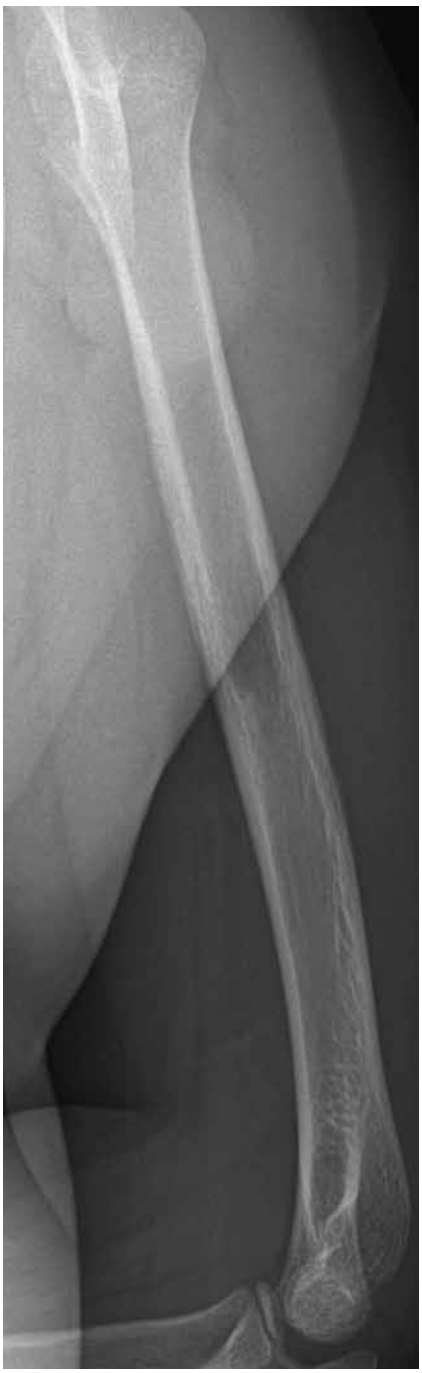

Fig. 1. — X-ray of the humerus showing a mediodiaphyseal lytic lesion with a «moth-eaten» cortical aspect.

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Fig. 2. — First MRI examination T1 (A), T2 (B): intramedullary lesion and periosteal oedema. Post-gadolinium T1 (C): nonhomogeneous enhancement, guiding the biopsy, which was positive only in the distal third.

evaluating treatment response. A recent study performed by Mengiardi et al. (5), analyzes MRI changes during and after treatment. They conclude that in a successfully treated patient, MRI shows a rapid decrease in tumor volume with complete disappearance of the soft tissue component. Minor signal abnormalities of bone marrow without clinical relevance may persist for up to 2 years (5).

In our case, MRI of the humerus in alternation with complete body MRI is performed every 3 months. The first control shows a decrease in tumor volume but persistence of bone signal anomalies. In all controls, decrease in volume and in signals anomalies is showed. The last control is performed 1 year and 6 months after first diagnosis.

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