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Case Report

Primary lymphoma of the distal radius of a child: imaging features

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ABSTRACT

Primary lymphoma of bone (PLB) is a rare entity, defined as a lymphoma confined to the bone without evidence of systemic involvement. The disease commonly affects middle-aged to elderly population and it accounts for less than 1% of all malignant lymphomas. We present a case of a 10-year-old child affected by PLB of the forearm and the frontal bone. Characteristic imaging features of PLB and the main differential diagnosis were discussed.

Introduction

Primary lymphoma of the bone (PLB) is a rare variant of extra-nodal non-Hodgkin lymphoma [1–3]. According to the World Health Organization classification, lymphoma involving bone can be divided into four groups: (1) single skeletal site without regional lymph node involvement; (2) multiple-bone involvement without visceral or lymph node involvement; (3) bone lesion with involvement of visceral sites or multiple lymph nodes at multiple sites; and (4) patient with known lymphoma and bone biopsy confirming involvement of bone. Groups 1 and 2 are considered primary lymphomas of the bone [3,4]. PLB account for less than 1% of all malignant lymphomas and 7% of malignant bone tumors. Only 4%-5% of extra nodal non-Hodgkin's lymphomas manifest as PLB [2, 5] and histologically most primary bone lymphomas are primary bone diffuse large

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B-cell lymphomas (PBDLBCLs). The disease commonly affects middle-aged to elderly population, with a median age of 48 years [2]. In this report, we describe the case of a 10-year-old boy affected with PLB presenting with a rare double involvement of the forearm and the frontal bone, focusing on PLB imaging features and differential diagnosis.

Case report

A 10-year-old boy came to our attention with a 2-month history of pain in the left forearm, in absence of trauma. On physical examination, a subcutaneous swelling was note in the frontal region. No other symptoms or signs were present; no history of fever, weight loss or sweating was reported. All blood tests were negative. A radiography showed structural alteration of the distal diaphysis of the radius, consisting in alternated areas of hyperlucency and sclerosis, associated with focal interruption of the cortex (Fig. 1a and b). The patient was admitted to the Pediatric Orthopedic department for further investigations. Considering the radiography findings, CT was performed. CT examination confirmed a 7-cm long structural alteration of the distal diaphysis of the radium with a multilayered periosteal reaction (lamelated or "onion skin" periosteal reaction), with bone rarefaction within the lesion, without involvement of the epiphysis, joint and adjacent bones. A CT of the frontal region was performed showing a second osteolytic lesion, with lenticular morphology, superficially spreading into soft tissues and deeply into the subdural bone, with enhancement after iodinate contrast medium administration (Fig. 2a-d). A 1.5T MRI was performed to evaluate the radial lesion that appeared markedly hypointense on T1-weighted images and slight hyperintense on T2-weighted images. There was also periosteal reaction with soft tissue edema. There was no soft-tissue mass (Fig. 3a-c). A Technetium bone scan was performed for staging of the disease. It showed focal uptakes in the radial lesion and in the frontal bone (Fig. 4). Additional investigation including whole body CT with contrast medium administration, spinal tap, a bone marrow aspiration and biopsy were all negative. An open surgical biopsy of the radial lesion was performed, revealing atypical large cells diffusely infiltrating the bone. Immunophenotyping showed positivity for CD20 staining, weak expression of BCL2, and negativity for CD3, CD10, CD30, BCL6, TdT (terminal deoxynucleotidyltransferase) and broad-spectrum cytokeratins; in situ hybridization for Epstein-Barr virus was also negative. Tumor cells showed a high proliferation index (90%) as determined by evaluation with anti-Ki67 antibody (Fig. 5a-d). The patient underwent four cycles of polychemotherapy according to the non-Hodgkin lymphoma (NHL) 97 protocol (consecutive blocks of polychemotherapy containing dexamethasone, cyclophosphamide, methotrexate, vincristine, cytarabine, etoposide, ipofosfamide, ara-c, daunomycine, and intrathecal therapy with methotrexate, cytarabine, and prednisolone). After two cycles of chemotherapy, the child underwent re-staging that showed partial regression of both radial and frontal lesions. After three more cycles of chemotherapy, a second open surgical biopsy was performed; showing complete remission of the disease, and MRI of the frontal bone

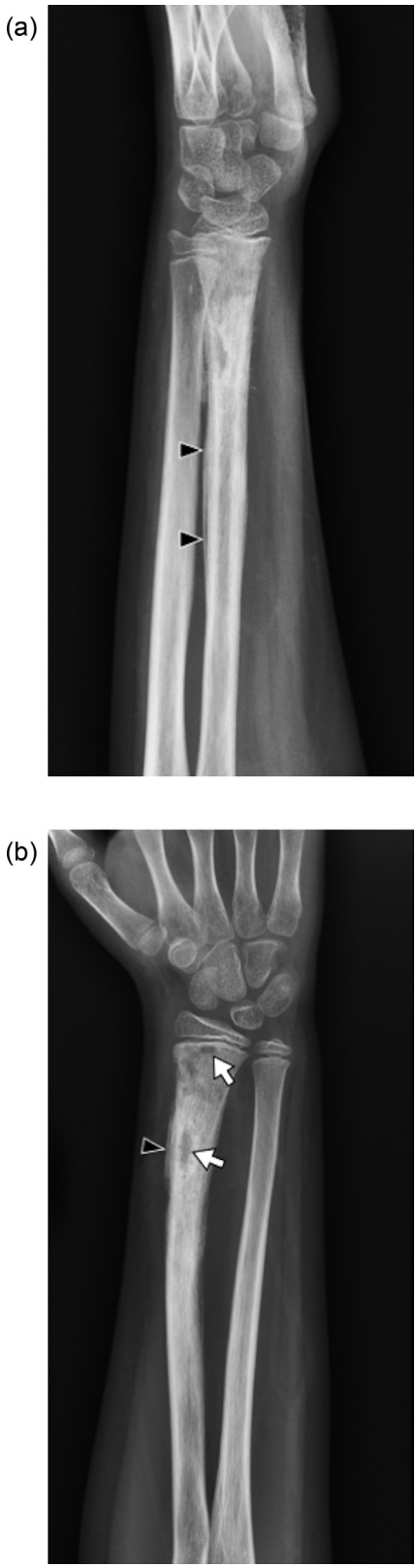

Fig.1 – Laterolateral projection (a) and anteroposterior projection (b) radiographs of the forearm showing a structural alteration of the distal diaphysis of the radius with alternated areas of hyperlucency (arrows) and sclerosis. A regular periosteal reaction is evident (arrowheads).