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Tuberculosis: the great mimicker. This time resembling Langerhans cell histiocytosis

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Abstract

The clinical case of a previously healthy 8 year old boy who presented to the emergency room with constitutional symptoms, along with multiple osteolytic lesions of the skull, ribs and vertebrae secondary to Tuberculosis infection resembling Langerhans cells Histiocytosis (LCH) is presented. Diagnosis was clarified with a biopsy of the frontal bone which revealed multinucleated cells with granulomas. Negative S100 and CDA1 immunohistochemical markers helped exclude. Also, drainage of a paracostal soft tissue abscess reported acid-fast bacilli seen in the Ziehl-Neelsen stain of pus. Bone marrow biopsy and aspirate were both normal. Similarities in clinical presentation as well as differences between both conditions that can help distinguish between these diseases and define the diagnosis are shortly discussed, along with the description of treatments and clinical evolution of the reported patient.

Introduction

LCH mimics many other conditions and a definitive diagnosis requires a combination of clinical manifestations, histological findings and specific immunohistochemistry markers. Tuberculosis is one of the most frequent differential diagnosis, but still, the clinical and radiological manifestations overlap between these two diseases. A high index of suspicion is necessary to clarify the diagnosis and offer timely treatment.

Case report

An 8-year-old, apparently previously healthy boy, presented to the emergency room with 3 months of a left paracostal abscess and a frontal subcutaneous nodule, accompanied by weight loss and intermittent non quantified fever. Skull radiography revealed osteolytic lesions of the frontal bone (Figure 1) and chest X rays showed osteolytic lesions of the left tenth and eleventh ribs, without pulmonary compromise. A 3-phase bone scintigraphy, reported abnormal cranial, costal and vertebral (T2, T4 and T9) capitation (Figure 2). Suspecting Langerhans cell Histiocytosis (LCH), a biopsy of the frontal bone along with drainage of the paracostal soft tissue abscess were performed. The Ziehl-Neelsen stain of pus drained from the paracostal abscess reported acid-fast bacilli (Figure 3). After growth in culture, a rapid test (SD BIOLINE TB Ag MPT64 Rapid TEST) confirmed *M. tuberculosis complex*, later identified as *M. tuberculosis* by PCR (GenoType Direct mycobacterial assay). Frontal bone biopsy revealed multinucleated cells with granulomas. S100 and CDA1 immunohistochemical markers to exclude LCH came back negative. The bone marrow biopsy and aspirate were both normal, as well as a negative 4th generation HIV test. No different organic compromise was identified except the multiple bones previously mentioned. The patient was started on anti TB treatment with HRZE during the first two months of therapy and continued on Isoniazid/Rifampicin for 10 months 3 times weekly. He completed one year of therapy with clinical improvement and resolution of symptoms. He was referred to immunology to exclude primary immunodeficiency due to his unusual clinical presentation with multiple TB osteomyelitis foci and a family history of a sister with previous Pott's disease.

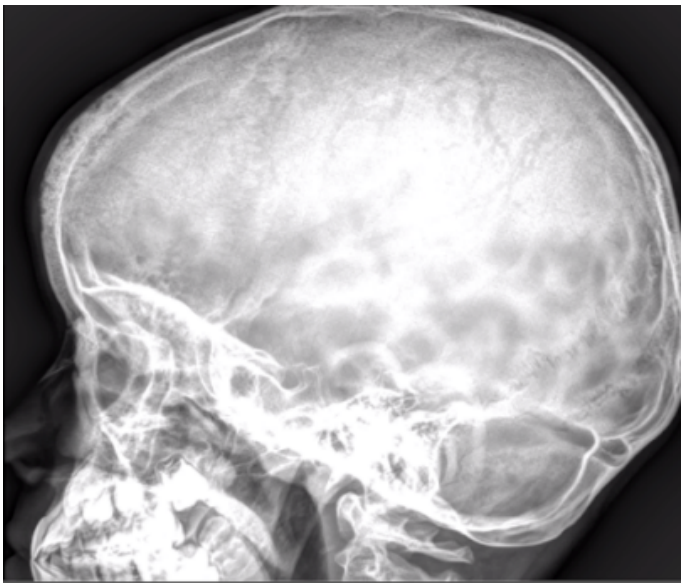

Figure 1. Osteolytic lesions of the frontal bone on skull radiography

Discussion

LCH of the bones is a relatively rare disorder of unknown etiology, most commonly reported to occur in children from one to three years of age [1]. A single (monostotic) bone lesion is a more frequent presentation than multiple (polyostotic) osseous compromise. In children, the skull is the most commonly affected bone. Conventional radiographs serve as the primary imaging method used to identify LCH lesions, since skull

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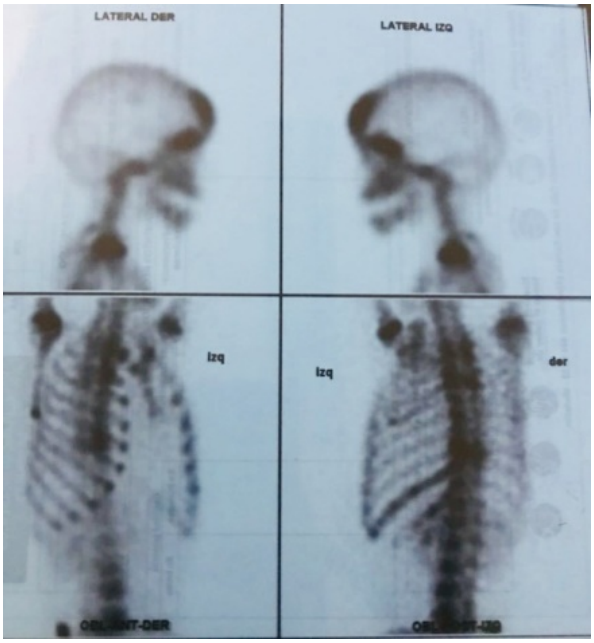

Figure 2. 3-phase bone scintigraphy abnormal cranial, costal and vertebral (T2, T4 and T9) capitation

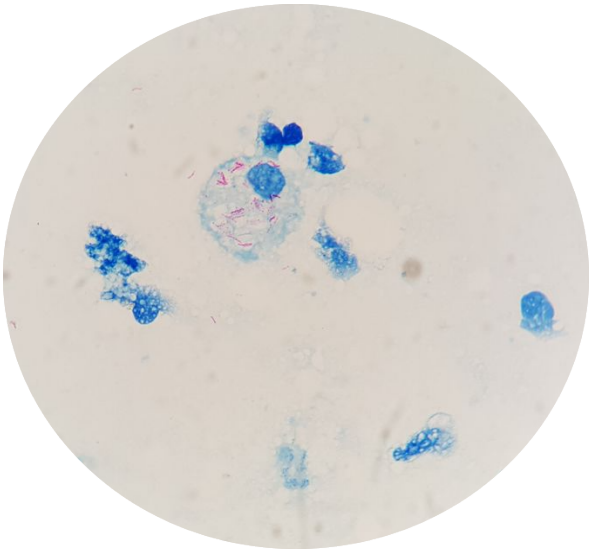

Figure 3. The Ziehl-Neelsen stain of pus drained from the paracostal abscess reported acid-fast bacilli

lesions classically have a punched-out, lytic appearance. For diagnostic confirmation, the histopathology is necessary, since the clinical and imaging characteristics of LCH may overlap with several other diseases, including infections like tuberculosis [2].

Tuberculosis (TB) has always been called the great mimicker due to its various clinical presentations, sometimes with nonspecific symptoms. It can affect almost any organ including bones; of these, the most frequently affected are the vertebrae in 41% of cases [3], Non-weight-bearing bones, such as the skull, clavicle, and mandible, are less often involved. About 50% of children with skeletal TB have abnormal chest radiographs. Confirmation of this diagnosis is based on microbiological identification in stains, cultures, pathology, or molecular methods [4].

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