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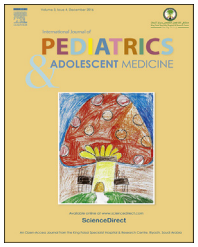

Images in Pediatrics

Extra-abdominal "skeletal" presentation of metastatic neuroblastoma

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We present a twenty-month-old boy who presented with right hip pain, limping gait, and fever. He was initially managed by orthopedic team in privet sector with provisional diagnosis of osteomyelitis based on the clinical and x-ray findings of poorly marginated osteolytic focus along the medial aspect of proximal right femoral metaphysis (Fig. 1). Child was then referred to our hospital because his symptoms did not improve despite the intravenous coverage with antibiotics. The follow up x-rays done for pelvis, both lower limbs, and chest showed multiple ill-defined osteolytic foci involving metaphyseal aspect of most of the long tubular bones bilaterally, some of which causing cortical disruption (Fig. 2 a&b). Based on these findings, patient underwent further clinical, blood workup, and cross sectional imaging including abdominal ultrasound and computed tomography which disclosed presence of left large suprarenal mass of heterogeneous texture displacing left kidney downward, splaying splenic vessels anterosuperiorly, and the ipsilateral renal vessels anteroinferiorly (Fig. 3 a&b). The findings were subsequently found to be bony metastases from abdominal neuroblastoma that was confirmed by Meta-iodobenzylguanidine (MIBG) isotope scan and histopathology. The pediatric radiologist first proposed the correct diagnosis.

Neuroblastoma is one of the most common solid extra-cranial malignant neoplasms in children accounting for almost 7% of all childhood cancer [1]. It arises from primitive neuroblasts of the embryonic neural crest, therefore, it can occur anywhere within the sympathetic nervous system [2]. The most common site of the primary tumor occurs within the abdomen, presenting an abdominal mass (65%) [3]. About half of these tumors arise from the adrenal medulla while the neck, chest, and pelvis represent other common sites [3]. Extra-abdominal presentations occur in about (50%) of cases and result from disseminated hematogenous metastatic disease to different sites such as bones (skull, long bones, ribs, and vertebrae), bone marrow, liver, skin, and non-regional lymph nodes; or from associated paraneoplastic syndromes.

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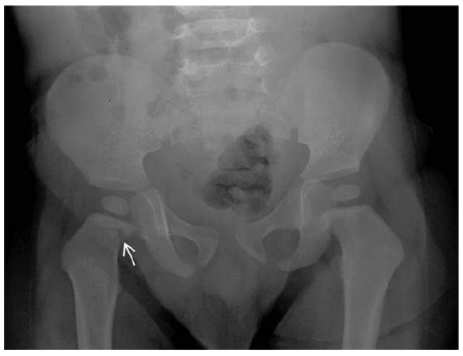

Figure 1 This is AP view of the pelvis x-ray demonstrating poorly marginated osteolytic lesion seen along the medial aspect of proximal right femoral metaphysis (arrow). Adjacent cortex appears ill-defined. No clear periosteal reaction. No other osseous lesion.

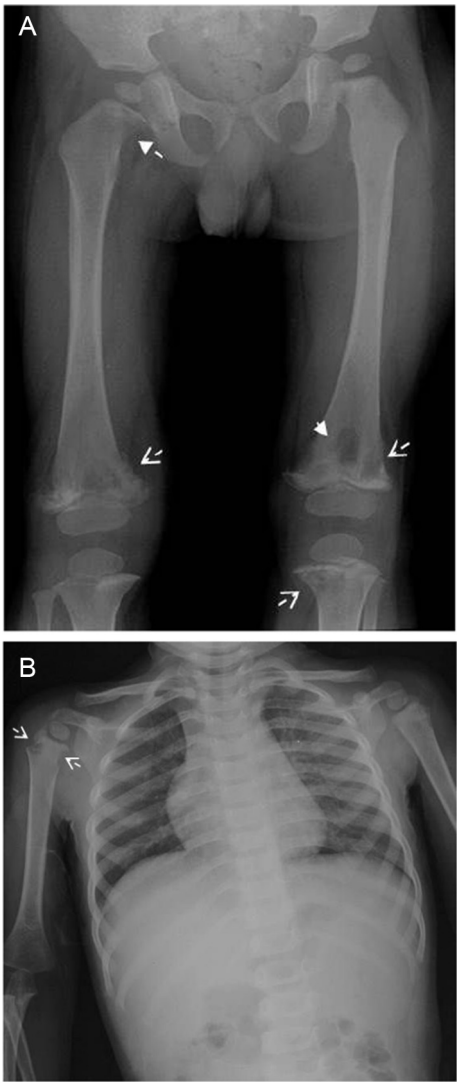

Figure 2 a&b: X-ray of pelvis and both lower limbs (a) and chest X-ray (b) show multiple osteolytic foci involving metaphyseal aspect of most of the long tubular bones (solid arrows), some of which appear ill-defined with cortical disruption (dashed arrows).

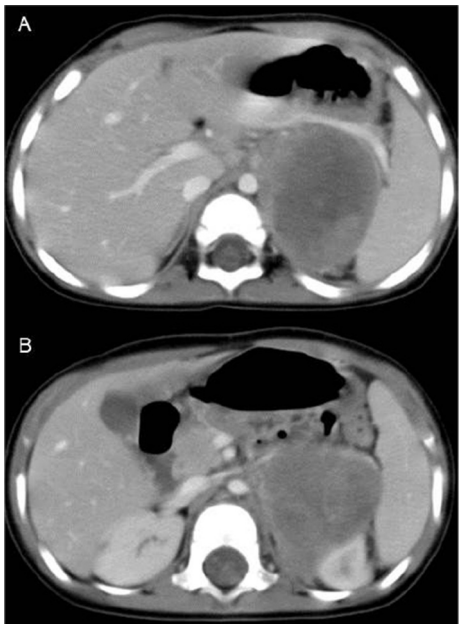

Figure 3 a&b: Two selected images from CT scan study of abdomen, (A&B) demonstrating large left suprarenal mass of heterogeneous texture displacing left kidney downward and laterally, splaying splenic vessels anterosuperiorly, and the ipsilateral renal vessels anteroinferiorly.

Widespread bone and bone marrow disease may present clinically with limping due to bone pain or irritability in younger children. Anemia, bleeding, or infection may be also seen due to bone marrow replacement. The constitutional symptoms associated with disseminated skeletal metastases may include failure to thrive and fever. Skeletal metastases occur in up to 60% of cases with a variable radiological appearance [1]. Skeletal lesions in long bones may present radiographically as single or multiple poorly marginated osteolytic foci with or without periosteal reaction, lucent areas of irregular mottling, lucent horizontal metaphyseal line, or vertical linear radiolucent streaks in the metadiaphysis [3—6]. Some skeletal lesions may present as a pathological fracture. Vertebral collapse might be the only findings seen in patients with spinal metastases presentation, while sutural diastasis is often seen in metastases to skull owing to subjacent dural metastases, another common location is the bony orbit which results in periorbital ecchymosis (''raccoon eyes'') and proptosis [1,5]. Early skeletal lesions may be missed both clinically and radiologically when cortical destruction is limited. The imaging findings might be misinterpreted as infective "osteomyelitis/septic arthritis" or inflammatory process such as "juvenile idiopathic/rheumatoid arthropathy" in particular when the clinical presentation overlaps with arthritic symptomatology and concurrent fever. The imaging findings may remain non-specific even with utilization of more sophisticated modalities such as the use of MRI in the evaluation of the conventional radiographic findings in particular when these are associated with presence of joint effusion, soft-tissue edema.

The purpose of this presentation is to highlight the pitfalls in making the correct diagnosis and alert both the

general radiologist and concerned physician to the possible extra-abdominal presentations and the atypical manifestations of this common childhood malignancy to derive early detection and diagnosis prompting proper treatment that help in increasing survival rates and minimizing irreversible damage.

Conflict of interest

The authors have no conflict of interest to report.

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