Eosinophilic granuloma is a solitary, non-neoplastic proliferation of Langerhan’s cell (LC). It was first described by Paul Langerhans in 1868. It occurs most commonly in children aged from 5 to 10 years and is uncommon in African population. The male to female ratio is two to one. The term histiocytosis X concerns the multiple organ involvement in disseminated Hand Schuler Christian disease. Neither the clinical nor the radiographic presentations of EG are specific, and the differential diagnosis is often difficult (1, 2).

Case report

A 2-year-old girl with no significant medical history presented with pain, swelling and restricted movements of the right forearm for two weeks. Physical examination revealed no history of fever, weight loss, or rash. Pain and deformity were not present at any other bony site or joint and there were no other soft tissue swellings.

Laboratory values showed a total white cell count of 14500/mm³ among which there was 61% Polynuclear Neutrophils and 30% Lymphocytes; an elevated Erythrocyte Sedimentation Rate of 45 mm/h with a slightly elevated C-reactive protein at 5.6 mg/dl.

The plain radiography of the affected forearm showed a globular aspect of the proximal part of the diaphysis of the radius bone associated with cortical irregularity, soft tissue swelling and periosteal reaction (Fig. 1).

An MRI imaging of the forearm was performed and demonstrated a hyper-intense bone lesion on T2-weighted sequences (Fig. 2). The bone lesion was hypo intense on T1 weighted sequences. Sequences with administration of gadolinium showed an intense cortical bone, periosteal and soft tissues enhancement (Fig. 3). A small fluid collection was protruding from the proximal radius which presented a broadened medullar cavity and cortical bone destruction (Fig. 4).

A biopsy of the bone lesion was performed and the histological examination revealed Langerhan’s cell histiocytosis characterized by Langerhan’s cell proliferation accompanied by giant cell and inflammatory infiltrate. Langerhan’s cells were positive for S-100 protein and CD1 (Fig. 5) (3, 4). Isotopic investigations enhanced the known lesion in the forearm but also a focal skull area. A head CT scan was performed and confirmed the presence of a solitary lytic lesion on the skull bone (Fig. 6).

The child did not receive any treatment. A subsequent every 6-month follow up by biological data and abdominal ultrasonography was programmed to exclude any further visceral involvement.

Discussion

Histiocytosis can present with various clinical manifestations (5-8).

Eosinophilic Granuloma (EG), 80%; EG is a self limited lesion in bone or lung. EG may be found in the skull, mandible, spine and long bones. EG can convert to more aggressive systemic forms described below.
Hand-Schuller-Christian disease (HSC) -15-20% is a chronic disseminated form of Langerhan’s histiocytosis occurring in older patients. There is a well known triad of HSC which is diabetes insipidus, exophthalmos and skull lesions.

Letterer-Siwe disease - 10% is a fulminant systemic disease that occurs in children under 3 years and is rapidly fatal.

The radiologic appearance of EG is non-specific and differs by location. Cranial lesions may appear on plain X-ray with sharp, punched out borders. In the diaphysis or metaphysis of long bones, the lesions develop in the center of the medullary cavity. The lesion may then cause endosteal scalloping and periosteal reaction (9).

Bone scintigraphy scanning is not useful in defining the EG by itself but is useful in detecting any other lesions after the diagnosis has been established (9).

CT scan and MR-imaging are useful to delineate the extent of the intramedullary and cortical extension (7-9).

The LCH imaging characteristics range between geographic non aggressive and highly aggressive appearance. Our case was the aggressive type of LCH with permissive and periosteal reaction. A fluid lesional component growing from the radius into the surrounding soft tissue was present resembling to an abscess formation.

The radiologic differential diagnosis includes any aggressive bony lesion like Ewing’s sarcoma.

**Fig. 2.** — Transverse section in T2-wt showing hyper-intense signal in the right upper radius surrounded by hyper-intense soft tissue infiltration (arrow).

**Fig. 3.** — Transverse section of the forearm in T1-wt sequences showing hypo-intense radius (A) and after gadolinium injection (B) it showed broadening of the medullar cavity and destruction of the cortical bone. The surrounding soft tissue is also enhanced by gadolinium.

**Fig. 4.** — Sagittal section of the forearm in T1-wt sequences showing broadening of the medullary cavity and destruction of the cortical bone of the proximal radius (A); after Gadolinium injection a collection raising from the radius towards the surrounding tissue is well depicted (B) (arrow).

**Fig. 5.** — Langerhan’s cell histiocytosis characterized by a proliferation of Langerhan’s cell CD1a positive (CD1a immunohistochemistry, original magnification [x200]).
osteosarcoma, metastases, leukemia and osteomyelitis. In bone tumours the soft tissue involvement is less common except for osteosarcoma. Leukemia is usually polyostotic. Osteomyelitis is most often metaphyseal in children (diaphyseal locations may be secondary to direct trauma). It is also very aggressive and permissive, and can be associated with reactive sclerosis and periosteal reaction. These features are very similar to LCH (10).

The commonest sites of EG are skull, legs, ribs, pelvis, spine, jaw, humerus, and tibia (9, 10). Radial involvement is less frequent. Due to this uncommon location and its highly permissive nature leading to infiltration of the surrounding soft tissue, the diagnosis of osteomyelitis with an abscess formation was proposed. Only when the biopsy findings fail to show any germs in bacteriology, histiocytosis can be suspected. In our case, the MRI features were very much similar to osteomyelitis. The elevation of the periostium with presence of liquid around the damaged bone could be misleading. When the spine is involved, preservation of disc space above and below the vertebral body helps to differentiate lesion from osteomyelitis (9).

**Treatment options and outcome**

Treatment of EG depends on the extent of the disease (9, 10). Treatment is planned after thorough evaluation of the patient to determine the extent of involvement. In localized disease, a biopsy alone is often enough to incite healing. In some cases the disease will regress without any treatment at all. Other treatment modalities of EG include curettage, excision, steroid injection, radiation (9). Chemotherapy (vinblastine) is recommended for systemic disease (8, 9). It is recommended to deliver the less possible aggressive treatment to keep the lesion under control and allowing it to heal without treatment. In our case, the affection was polyostotic without systemic involvement; therefore, it was decided to follow the child without treatment. The follow-up includes biological data and abdominal imaging to exclude visceral involvement. Until now, no new lesions have been detected.

**Conclusion**

The pathology of Langerhan's cell histiocytosis is not clearly understood (10). The disease having an unpredictable outcome and a heterogeneous clinical presentation makes the diagnostic work-up one of the most essential step in order to do a correct follow-up of patients.

**References**