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RESEARCH ARTICLE

SCAPULA LANGERHANS CELL HISTIOCYTOSIS IN A CHILD : A CASE REPORT.

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Abstract

Langerhans cell histiocytosis (LCH) is a proliferative histiocytic disorder relativily rare in the perdiatric group. This complex disease has no specific clinical or radiographic presentation. The scapula is one of the rarest site of LCH. Imaging and specially MRI are particularly helpful for characterizing the lesion and delineating the local and systemic extent. In this report, we present a rare case of Langerhans cell histiocytosis of the scapula in a 07 month-old baby.

Introduction:-

Langerhans cell histiocytosis (LCH) is primarily a disease of childhood . It affect predominantly a population between 5 and 15 years with an average age of 10-14 years.(1,2) The disease course has a spectrum from spontaneous resolution to progressive multisystem disorder. The lesions share the common histology of CD1a+/CD207+ dendritic cells. Radiography remains important in the diagnostic and staging procedure of bone lesion. Computed tomography (CT) or magnetic resonance imaging (MRI) may be necessary to assess precisely the degree of cortical bone destruction or to assess the degree of soft tissue infiltration.(3,4).

Case report:

A 07 month-old baby was reported with 02 month history of progressive pain in the right shoulder and restriction of movements. No fever, chills, night sweats or weight loss, were seen. The patient received analgesics on and off for 01 month and again complained of pain and presents a shoulder swelling with volume increase. X-RAY showed ill-defined osteolytic lesion without sclerotic margin in both scapula with cortical blow (Figure 1). MRI demonstrate an heterogeneous hyperintense signals involving the scapula on STIR and T2 weighted images , with enhancement after gadolinium-based contrast agent administration (figure 2). An associated large extraosseus soft tissue component was seen. CT scan demonstrates a geographic osteolytic lesion with subtle areas of marginal sclerosis. Biopsy taken from the bony lesion showed a lesion composed of sheets of histiocytes.

Discussion:-

Langerhans cell histiocytosis (LCH) is a rare disease, divided into three groups on the basis of the number of lesions and systems involved. (2,7)

In the year 1868, Paul Langerhans discovered the epidermal dendritic cells.However, the Birbeck granule was not identified until the 1970s by Nezelof.(1)

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Diagnosis of LCH is made by corroborating clinical features, histopathology, immunohistochemistry, and radiologic findings. No specific clinical and radiographic presentation of LCH is described in the literature.
LCH may present as a single bone lesion, skin rash, or as invasive disseminated disease and occurs typically in the pediatric and adolescent population,affecting both males and females.(1,2,8,6)

Although long bone involvement is more common in children, isolated flat bone involvement is more commonly seen in adults and is consistent with the case report findings described above. The scapula is the site of 3% of bone tumours, while for LCH it is the least common site. (1,2)

The radiographic appearance of the lesions depends on the site of involvement and the phase of the disease. Bone involvement usually presents as a single or multiple osteolytic lesions, with a sclerotic rim and surrounding sclerosis. This is concordant with the CT scan findings of our patient. On MRI, a focal lesion with extensive soft tissue and marrow edema is most commonly found as hypointese areas in T1W images and hyperintense area on T2W and STIR images with enhancement ,also consistent with findings in our case report.(3,5,6). An endosteal rim of low signal intensity can be found in LCH, which may be an early sign of healing.

The peritumoral edema is less extensive than that of Ewing's sarcoma and osteomyelitis, two most common differentials of LCH.. (5,8)

Bone scintigraphy may be a complementary technique to the radiographic skeletal but its sensitivity is limited. Recently, new imaging techniques, such as positron emission tomographycomputed tomography (PET-CT) and whole-body MRI, have developed in view of an improved assessment of the extent and severity of the disease (3,7),
The differential diagnostic considerations including metastasis, plasmacytoma, unicameral and aneurysmal bone cysts, and chondromyxoid fibroma among other disease processes (1,4)

The treatment of Langerhans cell histiocytosis (LCH) is still controversial and has changed over the past decades. With advances in pathogenesis, the trend now is towards radiation therapy and cytotoxic chemotherapy. (2)

Conclusion:-

Langerhans cell histiocytosis (LCH) is a rare disorder, mostly affecting children. The scapula is uncommonly affected in young children. Imaging specially MRI is presently the most informative imaging tool in the management of bone LCH.

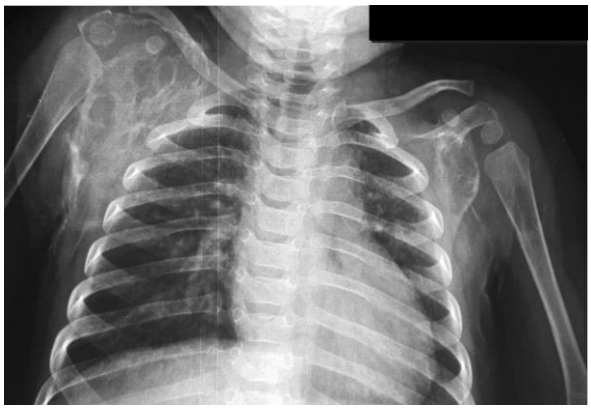

Figure1:-multiples osteolytic lesions in both scapula and left humerus.

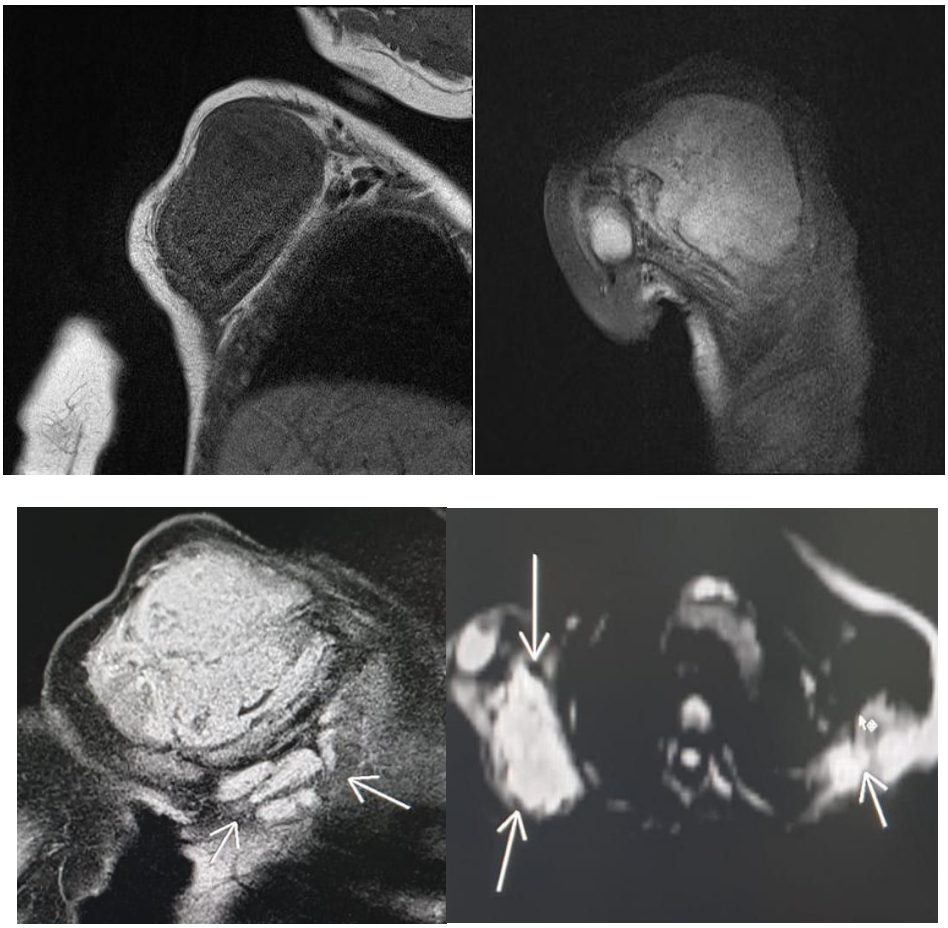

Figure 2:-MRI showed heterogeneous hypointense T1 hyperintense on diffusion and T2 weighted images. Heterogeonous enhancement (arrow). Lymphadenopathy was also noted in right axillary region.

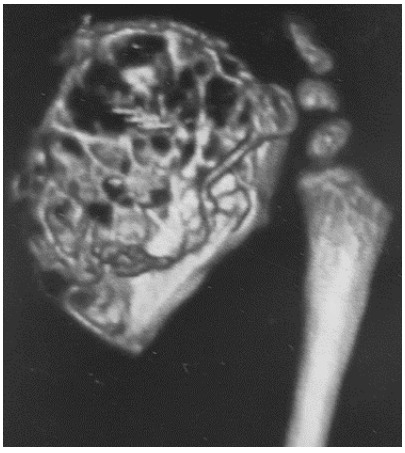

Figure 3:-3D CT image showd multiples lytic lesions

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