Journal Homepage: -www.journalijar.com
INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)
Article DOI:10.21474/IJAR01/6992
DOI URL: http://dx.doi.org/10.21474/IJAR01/6992

RESEARCH ARTICLE

A RARE CASE REPORT ON LANGERHANS CELL HISTIOCYTOSIS(LCH) IN A 4 YR OLD CHILD.

Sunil ku Agarwalla[1], Poonam Agrawal[2], Rina Meher[2].
1. Associate Professor, Department of Paediatrics, MKCG Medical College and Hospital, Berhampur, Odisha
2. Senior Resident, Department of Paediatrics, MKCG Medical College and Hospital, Berhampur, Odisha.

Manuscript Info

Manuscript History

Received: 19 February 2018
Final Accepted: 21 March 2018
Published: April 2018

Keywords:-
Langerhans cell histiocytosis, Histiocytosis X, APC.

Abstract

Langerhans cell histiocytosis (LCH), previously known as histiocytosis X, is an uncommon haematological disorder affecting infants and young children. It is a condition characterized by uncontrolled stimulation and proliferation of normal antigen presenting cells (APC), Langerhans cells. Its aetiology is unknown but it could be due to antigenic stimulus of an infection, genetic abnormality, deregulated immune response, or even clonal origin. Clinical presentation may be localized or systemic, invading skin, lungs and bone in adult, and bone marrow and lymph node in children. The purpose of this paper is to report a case of LCH in a 4yr old child with multisystem involvement.

Introduction:-

Langerhans cell histiocytosis (LCH) is a group of idiopathic disorders characterized by the proliferation of specialized bone marrow-derived Langerhans cells and mature eosinophils. The etiology is still unclear, common hypotheses include uncontrolled and irregular multiplication of lymphocytes and histiocytes after a gene mutation or infection, some fragmentary manifestations are not believed to be of a reactive pattern though but effectively represent real neoplasms [2, 3]. The nomenclature-Histiocytosis X was coined by Lichtenstein in 1953 to account for three clinical varieties which showed some histological characteristics in common: Eosinophilic granuloma, Letterer-Siwe syndrome and Hand-Schuller-Christian syndrome. The term ‘histiocytosis’ refers to the proliferation of histiocytes and other inflammatory cells, whereas the letter ‘X’ was added to denote the unknown etiology of the disease. The recent adoption of the terminology ‘Langerhans cell histiocytosis’ is due to the fact that the histiocytes involved in the disease present a phenotype which is similar to that of Langerhans cells found in normal mucosa and skin. A new proposed classification creates two categories of LCH- nonmalignant disorders such as unifocal or multifocal eosinophilic granuloma and malignant disorders including Lettere-siwe disease and variants of histiocytic lymphoma.

Any organ or system of the human body can be affected, but those more frequently involved are the skeleton (80% of cases), the skin (33%), and the pituitary (25%). Other organs involved are the liver, spleen, the hematopoietic system and the lungs (15% each), lymph nodes (5–10%), and the central nervous system excluding the pituitary (2–4%). In LCH, the long bone such as femur, humerus, and tibia are most commonly involved. The clinical course may vary from a self-limiting disease to a rapidly progressive one that might lead to death.

Corresponding Author:- Sunil ku Agarwalla.
Address:- Associate Professor, Department of Paediatrics, MKCG Medical College and Hospital, Berhampur, Odisha

Case Report:-

A 4yr old Fch was admitted to paediatrics ward with complaints of fever for last 4 months and pain in right knee joint for 2.5 months. Pain was dull aching in nature and was associated with limping. Patient was prescribed several antibiotics for fever and NSAIDs. Patient was diagnosed as a case TB arthritis (Rt knee joint) by local physician and was on ATT for past 2 months but symptoms didn't improve. Examination revealed severe pallor, right posterior cervical lymphadenopathy, multiple bony swellings on scalp, seborrhoea of scalp and hepatomegaly.

CBC revealed severe anemia without any evidence of bicytopenia or pancytopenia. Detailed investigations showed dimorphic anemia with Hb-4.2gm%, TLC-8500/cmm, TPC-2,80,000/cmm, retic count- 2%, ESR-90mm/1st hr, LFT & RFT within normal limits, s.triglyceride-83mg%, s.ferritin-238.7ng/ml, HPLC- normal, X-ray of right knee joint revealing few punched out lesions in lower part of right femur and epiphysis and metaphysis of right tibia, X-ray of skull bone showing defects in outer table of cranium, bone marrow aspiration study showing increased number of macrophages showing evidence of hemophagocytosis, s/o hemophagocytic syndrom. Non palpable spleen and normal serum ferritin ruled out the provisional diagnosis of HLH. Histopathological examination of right cervical nodes revealed features of LCH. Patient was diagnosed with LCH with multisystem involvement and chemotherapy was started (vinblastine + corticosteroids). On follow-up, now after 2 months bony swellings on scalp have subsided and the osteolytic lesions in right tibia have disappeared.

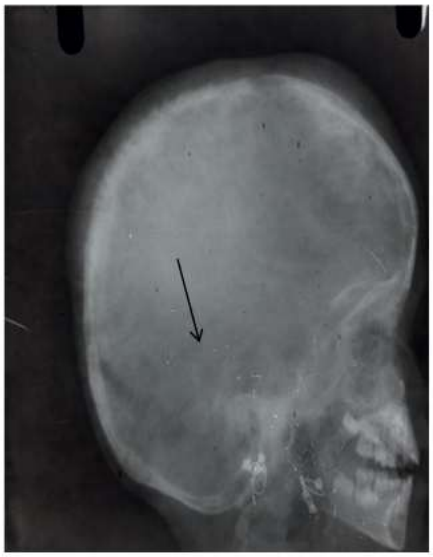
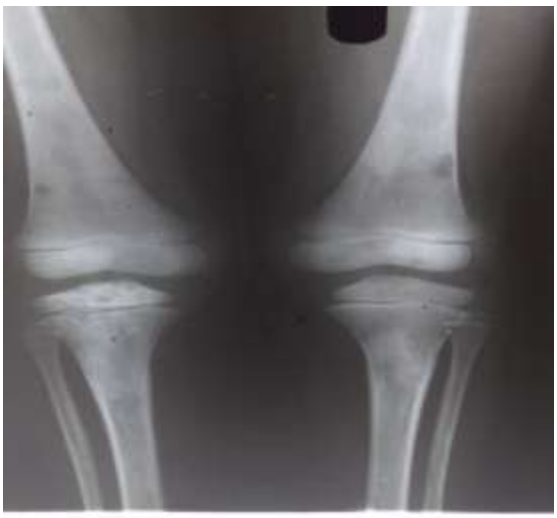


Figure 1:-Lateral x-ray of skull showing multiple osteolytic lesions

Figure 2:- Multiple swellings on scalp



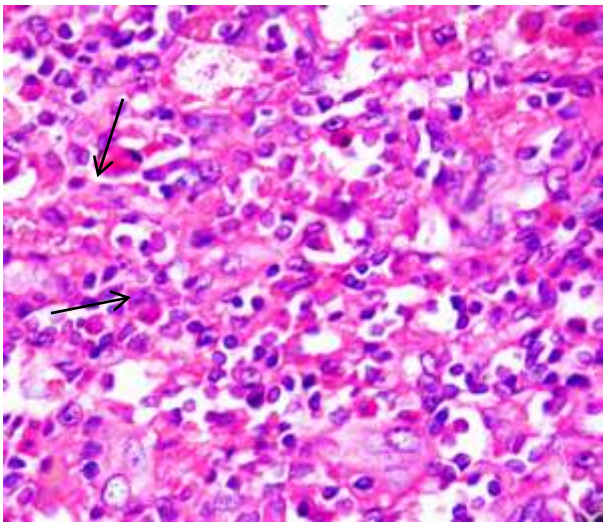


Figure 3b:- Histopathology of cervical lymph node showing features of LCH.

Discussion:-

LCH, formerly called histiocytosis X, was introduced as a collective term to represent a spectrum of clinicopathological conditions that are characterized histologically by a monoclonal proliferation of histiocyte-like cells accompanied by varying numbers of eosinophils, lymphocytes, plasma cells and multinucleate giant cells. The histiocytes present in this lesion have been identified as Langerhans cells. In LCH, literature review suggests a monoclonal proliferation of Langerhans cells leading to destruction of hard and soft tissues. For many years, it has been considered as a reactive disorder of immune regulation and not true neoplasm. However, recent evidence has shown monoclonal proliferation of lesion cells, findings that is indicative of neoplastic lesion. [7]

LCH usually encountered in children between 1 and 15 years old with a peak incidence between 2 and 4 years of age, with male predilection twice that of female. Among children under the age of 10 years, yearly incidence is thought to be 1 in 200,000. The present case was reported in a 4-year-old female child. Solitary or multiple bone lesions are the most common clinical presentations seen in the majority of cases, involving skull, jaws, ribs, long bones, vertebrae and pelvis. In our case, skull bone and tibia were involved.

LCH has a broad clinical spectrum, wherein clinical course varies considerably according to the age of the patient, extent and pathological infiltration of Langerhans cells in various organ systems of body such as bone, skin, lymph nodes, bone marrow, liver, spleen, lung, endocrine system, ear and brain. Bones of the skull commonly involved are orbital and temporal bones, the sella turcica.

For therapeutic purposes, multisystem LCH is divided into two categories based on the risk of mortality from disease. Risk LCH includes all patients with disease in two or more organs including a risk organ, defined until recently as involvement of liver, spleen, lung and hematopoietic system. The latter is defined by the presence of anemia, neutropenia and/or thrombocytopenia, and is not excluded by the absence of morphologic infiltration of bone marrow. Hematopoietic disease may be associated with secondary hemophagocytosis – a common finding in young patients who died from disease in a French study (Donadieu, personal communication). [6]

Any child presenting with prolonged fever and bony swelling particularly of skull bone, we have to suspect LCH. However, neuroblastoma can present with similar features due to metastasis to skull bones. Early diagnosis and specific chemotherapy gives good outcome in LCH. Our patient who is under treatment is doing well and need few more cycles of chemotherapy. We are reporting this case as this is a rare diagnosis in children.

References:-

1. Greenberg MS, Glick M, Ship JA. Burket's Oral Medicine. 11th ed. Hamilton: BC Decker; 2008. pp. 144–5.
2. Postini AM, Andreacchio A, Boffano M, Pagano M, Brach Del Prever A, Fagioli F. Langerhans cell histiocytosis of bone in children. J Pediatr Orthop B 2012; 21: 457-62.
3. Aricò M, Danesino C. Langerhans' cell histiocytosis: is there a role for genetics? Haematologica 2001; 86: 1009-14.
4. Saliba I, Sidani K, El Fata F, Arcand P, Quintal MC, Abela A. Langerhans' cell histiocytosis of the temporal bone in children. Int J Pediatr Otorhinolaryngol 2008; 72: 775-86.
5. Nicollas R, Rome A, Belaïch H, Roman S, Volk M, Gentet JC, Michel G, Triglia JM. Head and neck manifestation and prognosis of Langerhans' cell histiocytosis in children. Int J Pediatr Otorhinolaryngol 2010; 74: 669-73.
6. Riccardo Haupt MD,Milen Minkov MD,Itziar Astigarraga MD, Eva Schäfer MSc ,Vasanta Nanduri MD ,Rima Jubran MD; Langerhans cell histiocytosis (LCH): Guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years*
7. Willman CL. Detection of clonal histiocytes in Langerhans cell histiocytosis: Biology and clinical significance. Br J Cancer Suppl. 1994;23:S29–33. [PMC free article] [PubMed]
8. Henry RJ, Sweeney EA. Langerhans' cell histiocytosis: Case reports and literature review. Pediatr Dent. 1996;18:11–6. [PubMed].