University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2018, Vol. 4(2)

LANGERHANS CELL HISTIOCYTOSIS AS A RARE CAUSE FOR HEADACHE-A CASE REPORT

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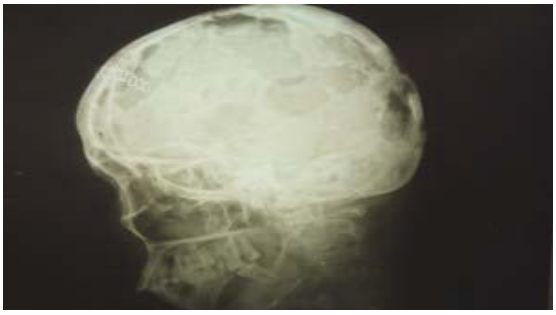
Abstract : 51 year old female presented with headache, physical examination and investigations were helpful in making a diagnosis of LANGERHANS CELL HISTIOCYTOSIS. This case is presented for its rarity of occurrence and also to emphasize the need for awareness among the physicians for
early diagnosis and successful outcome.
Keyword :LANGERHANS CELL HISTIOCYTOSIS, HISTIOCYTOSIS X, LCH



punched out lytic lesions-staplers revealing biopsy site

INTRODUCTION

Langerhans Cell Histiocytosis (LCH) is a rare disease characterized by aberrant proliferation of a specific dendritic (Langerhans) cell belonging to the monocyte macrophage system. LCH is encountered more often in children, with a peak age range of 1-3 years. LCH is rare in adults and the incidence may be underestimated due to the fact that many cases likely go undiagnosed.

CASE DESCRIPTION

A 51 yr old female admitted with C/O headache-8 months duration and multiple depressed softareas in scalp-6 months duration , h/o pain over the depression since 2 months .No H/O dysnoea/skin lesions/ear discharge/polyuria/polydipsia/ weight loss/visual blurring.No significant family history.On general examination, **Multiple tender cranial vault bone defects +** Left upper cervical nodes palpable ,two discrete nodes palpable,largest one measuring 1.5 cm × 1.5cm ,firm in consistency,mobile,non tender,No other significant lymph nodes palpable. Vitals and system examination normal. No lump in the breast.Fundus-normal study

WORK UP
urine:Bence jones protein-negative
Complete blood count-
- TC-7100,DC: P 74,L 26,E-0, Hb-12.1 gms,PCV-37.3: RBC-4.6 million /cumm; MCV- 79.5,MCH-25.8,MCHC-32.4, Platelet count-3.74 lakhs/cumm, ESR-5mm at ½ hour/10mm at 1hour
Serum protein electrophoresis: no M –band.
Ø Serum uric acid-5.5mg;S.calcium-10 mgs
Ø Liver function test:Total bilirubin- 0.5,direct-0.3, Indirect-0.2,SGPT-24 SGOT- 22 ,ALP- 190,PROTEIN- 7.6, Alb-4.6gm,glob-3 gm
X-ray skull-multiple punched out lesions in vault
X-ray-skeletal survey – revealed no other osteolytic lesions
Biopsy from osteolytic lesions revealed,Variable admixture of eosinophil,langerhans cell, non-specific histiocytes and lymphocytes. Impression : langerhans cell histiocytosis
Ø Bone Marrow study- Reactive marrow with lymphocytosis
Ø FNAC-cervical lymph node-Reactive lymphadenitis
Ø Peripheral smear:normocytic normochromic smear.
CT-CHEST-normal study
CT-BRAIN-**multiple osteolytic lesions over cranial vault** , no evidence of bleed/SOL MRI BRAIN-multiple T1 hypointensity lesion and T2 hypointensity lesion in skull giving hole within hole appearance. partial empty sella.
Patient was referred to higher centre for further management,where cladribine monotherapy was started.On follow up after 6 months, there was marked improvement of quality of life with decrease in headache and regression of bone lesions.

DISCUSSION

Langerhans cell histiocytosis (LCH) is a rare proliferative disorder in which pathological Langerhans cells (LCs) accumulate in a variety of organs. Overall male-to-female ratio is 1.5:1. Punched out lytic lesions are characteristic.1 These disorders can involve many organ systems but primarily affect the bone, skin, lymph nodes, lungs, liver and spleen, endocrine glands, and nervous system2 . Bone involvement is observed in 78% of patients with Langerhans cell histiocytosis and often includes the skull (49%), innominate bone (23%), femur (17%), orbit (11%), and ribs (8%). Lesions may be asymptomatic or may present with swelling over the affected bone, pain or pathological fracture. In adult, most common organ involved is lung followed by bone and skin3,4.Lung disease in adults is associated with smoking1. Management includes **1.**Single agent therapy with Purine analogs—cladribine or pentostatin

2.Multiagent therapy with cytarabine arabinoside , vincristine, and prednisolone.Bisphosphonates reduces skeletal complications.4,5 Length of time from the first symptom(s) to diagnosis is frustratingly long.Many patients wait one tofour years before the correct diagnosis is made, and others have symptoms for 5 to 20 years.6The difficulty in making an accurate diagnosis isreflected in the long time from symptom onset to diagnosis, lack of clinical suspicion, and the variable characteristics of the disease.

CONCLUSION

This case was unique as her symptoms of LCH were related only to bone involvement.Adult LCH without history of smoking is an uncommon occurence.Langerhan cell histiocytosis is one of the prominent causes for punched out lytic lesions of skull.The above case was worked up for headache.

Biopsy of the lytic lesions revealed an uncommon diagnosis of histiocytosis.The case was presented for importance of early diagnosis of histiocytosis and detection of visceral involvement before significant clinical complications occur.

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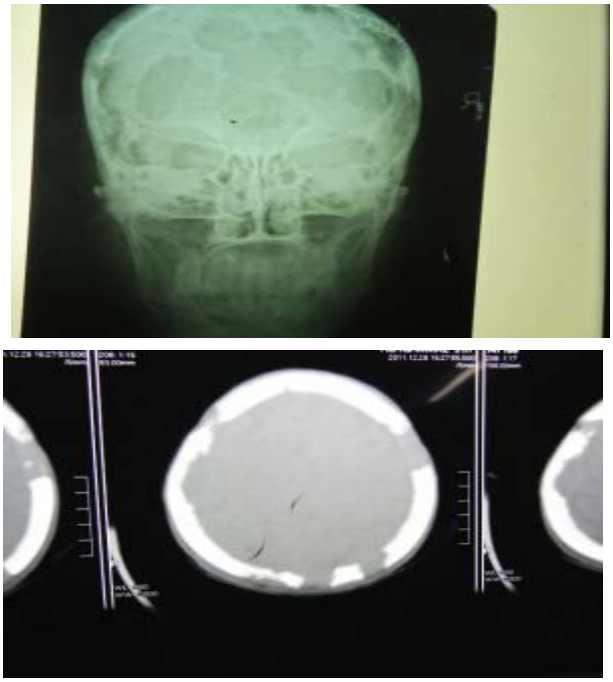
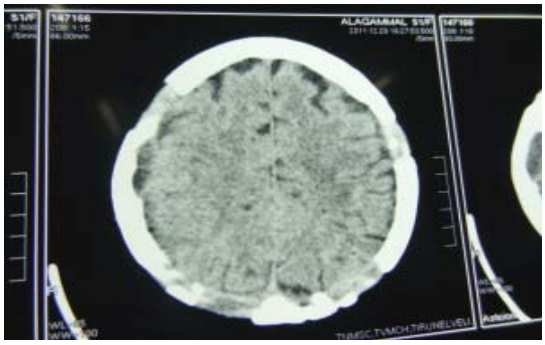
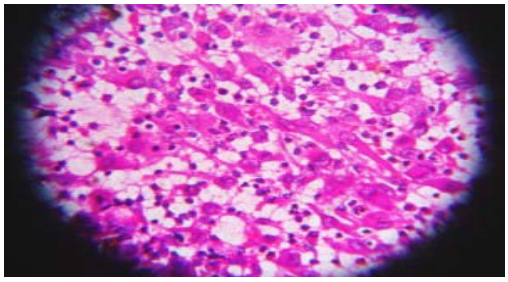


CT showing lytic lesions



CT BRAIN showing lytic lesions



microscopy revealing variable mixture of langerhan cell punched out lytic lesions