Tissue is the Issue- A rare case of Langerhans Cell Histiocytosis

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Background:

Langerhans cell histiocytosis (LCH) is a rare disease characterized by accumulation of CD1a cells and wide-ranging organ involvement. It is the condition characterized by proliferation and uncontrolled stimulation of antigen presenting cell called Langerhans cells. Incidence of the disease is around 3-5 /million, majority of the patients are younger than 3 years old, while in adult's incidence is around 1-2/million (1) Langerhans cell histiocytosis pathogenesis is due to the gain of function mutation in BRAF (V600E) as this mutation was seen in fifty percent of patients. We describe a case of an adult presenting with LCH.

Summary:

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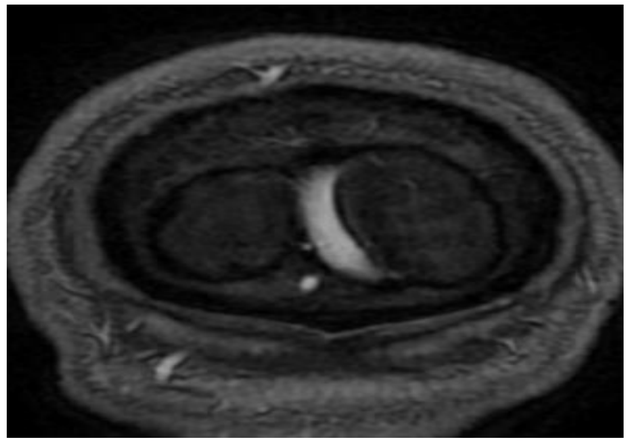
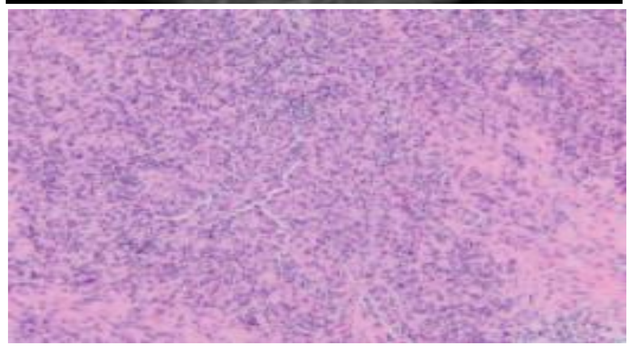
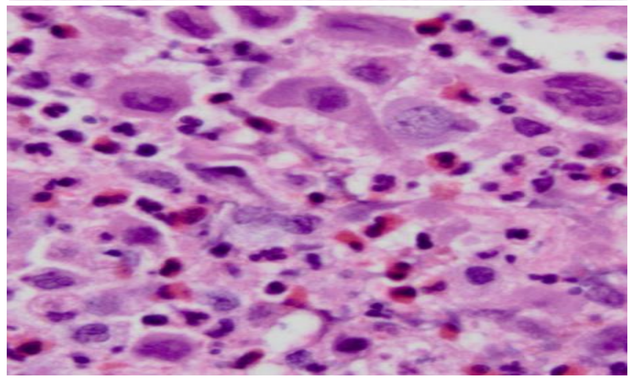
- 21-year-old male presented with the complaints of tender swelling on the right forehead for several days, associated with intermittent vision changes.
- Physical examination was unremarkable except palpable firm tender mass on right forehead without erythema and fluctuance.
- CAT Scan of the head showed right frontal bone osteomyelitis with epidural abscess and pansinusitis. He was started on broad spectrum antibiotics.
- MRI Brain showed right frontal bone osteomyelitis with underlying epidural abscess.
- Infectious disease workup was negative.
- Craniectomy was done and bone biopsy showed Langerhans cell histiocytosis. Antibiotics were discontinued.
- CAT scan chest, abdomen, and pelvis was negative for any abnormal findings. Hormonal work up was with in normal limits. He was discharged with follow up with endocrinology/oncology.







Significance :

Langerhans cell histiocytosis (LCH) is a difficult disease to diagnose because of its overlap with different diseases. Biopsy showing histiocytes with expression of cd 207 and Cd1a is pathognomonic of Langerhans histiocytosis. This case highlights a rare, primarily pediatric disease affecting an adult patient. With a broad spectrum of clinical presentations, LCH requires a high suspicion for disease; Diabetes insipidus is the most common complication of LCH in CNS (2). Initial work up for staging of LCH includes MRI of brain, comprehensive metabolic panel, CBC with differentials, LDH, T4 , TSH, cortisol , ACTH, testosterone , prolactin , IGF-1,FSH, LH, morning urine and serum osmolarity(3).Treatment of LCH include surgery, radiotherapy, topical corticosteroids and chemotherapy depending on the extent of disease. Prognosis is unpredictable and long term follow up is required to identify disease reoccurrence and LCH patients are at risk for secondary malignancies.

First image shows bone osteomyelitis with abscess . 2nd and 3rd image shows histological section of Langerhans cell histiocytosis showing multinucleated giant cells and prominent nuclear grooves.

References:

1)Carstensen H, Ornvold K. The epidemiology of LCH in children in Denmark. Med Pediat Oncol. 1993; 21:387-388.

2)Dunger DB, Broadbent V, Yeoman E, et al. The frequency and natural history of diabetes insipidus in children with Langerhans-cell histiocytosis. N Engl J Med. 1989;321(17):1157–1162. [PubMed]

3)The Mayo Clinic Histiocytosis Working Group Consensus Statement for the Diagnosis and Evaluation of Adult Patients With Histiocytic Neoplasms: Erdheim-Chester Disease, Langerhans Cell Histiocytosis, and Rosai-Dorfman Disease Gaurav Goyal, MBBS, Jason R. Young, MD Matthew J. Koster, et al