Article

PAMJ Clinical Medicine

Images in clinical medicine

Multisystemic Langerhans cell histiocytosis with orbital involvement

Alexander Gerard Nino Lim Gungab, Gangadhara Sundar

Corresponding author: Alexander Gerard Nino Lim Gungab, Department of Ophthalmology, Orbit, Oculofacial and Lacrimal Surgery service, National University Hospital, Kent Ridge, Singapore. alexgungab@gmail.com

Received: 09 Dec 2022 - **Accepted:** 03 Jan 2023 - **Published:** 06 Jan 2023

Keywords: Langerhans cell histiocytosis, skull, lytic

Cite this article: Alexander Gerard Nino Lim Gungab et al. Multisystemic Langerhans cell histiocytosis with orbital involvement. PAMJ Clinical Medicine. 2023;11(7). 10.11604/pamj-cm.2023.11.7.38469

Available online at: https://www.clinical-medicine.panafrican-med-journal.com//content/article/11/7/full

Multisystemic Langerhans cell histiocytosis with orbital involvement

Alexander Gerard Nino Lim Gungab[1,&], Gangadhara Sundar[1]

[1]Department of Ophthalmology, Orbit, Oculofacial and Lacrimal Surgery service, National University Hospital, Kent Ridge, Singapore

[&]Corresponding author

Alexander Gerard Nino Lim Gungab, Department of Ophthalmology, Orbit, Oculofacial and Lacrimal Surgery service, National University Hospital, Kent Ridge, Singapore

Image in medicine

A 3-year-old male initially presented with low grade fever and scalp rash of 2 months' duration. Patient also showed macrocephaly with bilateral proptosis since birth. Work up revealed CD1a, S100, Langerin and BRAF mutation positive results. A diagnosis of multisystemic Langerhans Cell Histiocytosis (LCH) was confirmed. Multiple punched-out bony lytic lesions with geographic appearance predominantly in the frontal and parietal regions. These are classic radiographic finding for cranial vault lesions in LCH. A solitary lesion may already be suggestive. Langerhans cell histiocytosis is a rare disorder commonly affecting children with unknown etiology. It has multisystemic involvement (skin, lymph nodes,

bones, central nervous system, abdominal organs especially the liver and/or spleen). The bones are commonly involved in this disease. That's why biopsy-proven LCH should include a skeletal survey. Langerhans cell histiocytosis can involve any bone in the human body but mainly in the axial skeleton. Over 50% occur in the skull, ribs and pelvis.

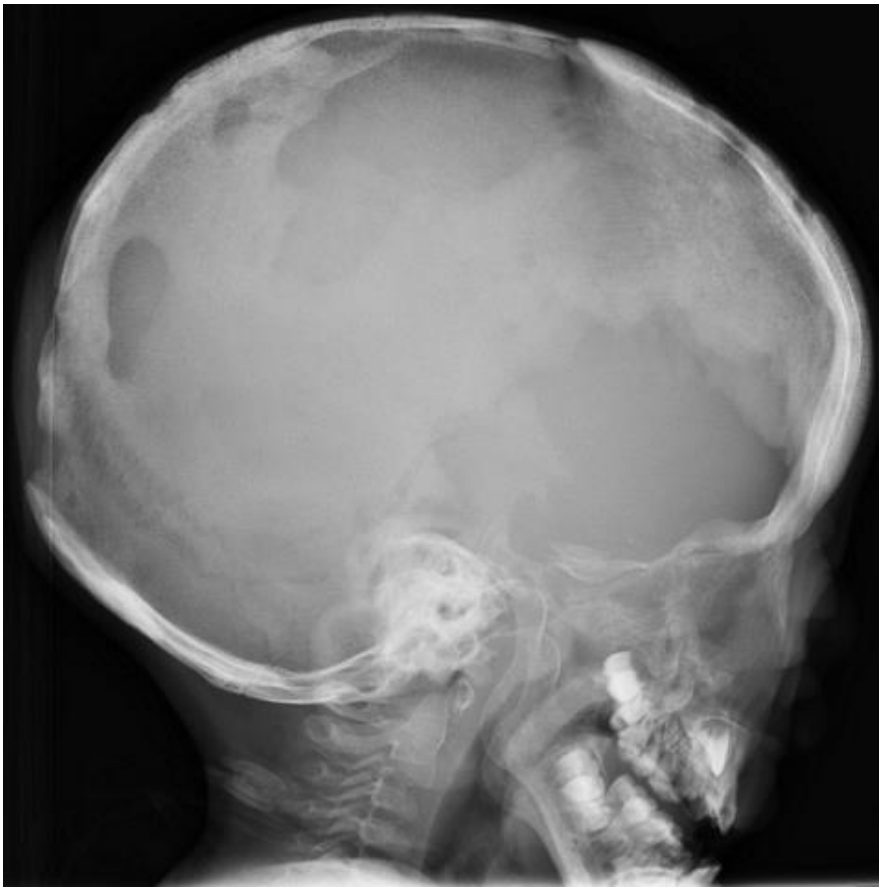

Figure 1: multiple punched out bony lytic lesions in the skull