Recurrent Multisystem Langerhans Cell Histiocytosis with Skull-Predominant Manifestations: Case Report and Review of the Literature

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Background
- Langerhans cell histiocytosis (LCH) is a hematologic disease process where Langerhans cells proliferate as a localized or disseminated disease process, most commonly occurring in children under 15 years of age with an incidence rate of 4.5 per million children.
- One variant of LCH, eosinophilic granuloma, is most commonly identified in lesions within the skull as a single or multiple osteolytic lesions which may be painful.
- We present a case of a child with a history of disseminated LCH with recurrence in the skull.

Materials & Methods
- A review of the patient’s electronic medical record was undertaken and radiologic and histologic findings were reviewed.
- A literature search was conducted using appropriate key words.

Case Report
- The patient is a 5-year-old boy. He presented initially at age 2 with left eye swelling.
- Imaging of his head revealed (1) a lesion in the right orbit, 2.6 cm, with mass effect on the left globe, and (2) an area of lytic bone loss, 2.1 cm, suspicious for an eosinophilic granuloma, with a subtle asymmetric T2 hypointensity within the left orbit. Treatment was pursued with LCH III on 6/7/2021 (Arm B). He completed chemotherapy on 9/14/2022.
- Scans showed no clear evidence of recurrent or persistent disease, although there was a subtle asymmetric T2 hyperintensity within the left orbit.
- He subsequently, he developed frequent headaches and fatigue along with a new, palpable bump on his head.
- CT and MRI imaging of his skull in 06/2024 revealed (1) a new left parietal skull lesion, lytic, and (2) a lesion in the right orbit, 2.6 cm, with mass effect on the left globe. A. Standard CT scan; B-C, CT with Bone Windows, all coronal sections.

Results

Literature Review Summary
- Langerhans cells are specialized dendritic cells which present antigen to naïve T cells and are predominantly found in the epidermis.
- Langerhans cell histiocytosis (LCH) is a hematologic disease process where Langerhans cells proliferate as a localized or disseminated disease process.
- The disease is characterized by Birbeck (tennis racket) granules on electron microscopy as well as CD1a and S-100 positive by immunohistochemistry.
- It most commonly occurs in children under 15 years of age with a slight bias towards males with reported sex biases 1.6:1 (male: female).
- Langerhans Cell Histiocytosis has an incidence rate of 4.5 per million children. Among children, 63% of LCH neoplasms arise from bone or bone marrow.
- The single system Langerhans Cell Histiocytosis is the most prevalent type in children, accounting for 56-85% of all LCH recorded cases, sources provide statistics that multiple bone lesions occur approximately 19% of the time.
- The recurrence rate is relatively low, with a 5.7% recurrence in the unifocal bone disease, 12.5% reactivation in single system bone disease and 23.8% recurrence in multifocal bone disease.

Conclusions
- LCH can present in the skull as a localized lesion or as part of a disseminated disease process.
- The recurrence rate after treatment varies from 5.7% recurrence in unifocal bone disease, 12.5% reactivation in single system bone disease, and 23.8% recurrence in multifocal bone disease.
- When disseminated, with involvement of bone marrow, liver, or spleen, treatment is more challenging and survival is reduced.
- This case illustrates the challenges of effective treatment of LCH, particularly when disseminated, and the need for vigilant follow-up.

References