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Clinical Image

Acquired Meningocoele due to Langerhans Cell Histiocytosis

Shreya Sengupta* and Sayantan Bose

Department of Pediatrics, RG Kar Medical College, India

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***Corresponding author**

Shreya Sengupta, Department of Pediatrics, RG Kar Medical College, India, Tel: +917278605066;
Email: shreya.95edcn@gmail.com

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Abstract

Langerhans cells histiocytosis is a relatively common ailment in pediatric age group. Common presentations include bone pain, pathological fractures, skin and scalp eruptions, features of pancytopenia and lymphadenopathy. Sometimes there can be chronic cough and shortness of breath due to lung infiltration. Acquired meningocoele is a rare presenting feature for this disease. Here we present one such case of unifocalpolyostotic Langerhans cells histiocytosis in a 16 year old male patient who had acquired a meningocele. Investigations showed multiple punched out lesions in skull and lytic lesions in several bones but the lung field was cleared. Biopsy and histochemistry confirmed the diagnosis. However the child was found to be HIV positive for which treatment was started but he developed renal failure due to hypercalcemia and died.

A 16 year old boy presented with dull, continuous headache for 2 weeks and a soft swelling over his forehead for 5 days. The child was pale, had significant weight loss, fever, growth retardation and hepatosplenomegaly. The swelling over his forehead was 2cm ×2cm, non tender, soft, fluctuant, transilluminant and showed expansible cough impulse. Palpation of rest of his head revealed multiple small depressed areas where the skull seemed to give way. He was thought to be having severe under nutrition along with vitamin D deficiency. Although Blood tests showed anaemia, his protein, glucose, electrolytes, vitamin D all were normal; whereas his calcium and alkaline phosphatase was high. X-ray of skull had multiple punched out lesions with non-sclerotic margins (Figure.1). The chest radiograph showed purelylytic areas in clavicle and ribs but the lung fields were clear. CT scan of head showed that his skull bones were completely dehiscent at certain areas through which dura was bulging out, one of which appeared as a meningocoele over his forehead (Figure.2). Biopsy showed abundant eosinophils and Langerhans cells with CD1a positivity which is pathognomonic of LCH [1]. A diagnosis of unifocal polyostotic Langerhans Cell Histiocytosis was established. LCH predominates in children and its annual incidence is estimated at 4.6 per million in children fewer than 15 years of age [2]. Bone is the most frequently affected tissue in children with LCH, encountered in about 75-80% of patients with LCH [2], with unifocal involvement being more common than multifocal involvement [3]. LCH can involve any bone, but there is a predilection for the axial skeleton, with more than 50% of bone lesions occurring in the flat bones (skull, ribs, pelvis) [4]. However this child was found to be HIV infected as well, which required prior attention. Antiretroviral therapy was scheduled to be started, but he rapidly developed renal failure with hypercalcaemia. He was shifted to ICU, but succumbed to death after 3 days.

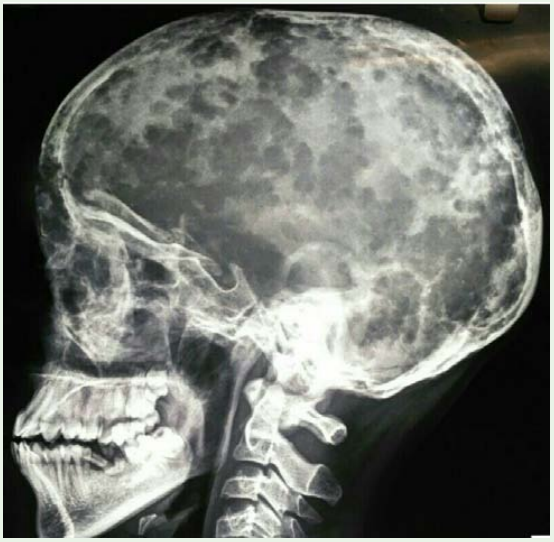

Figure 1: X-ray of skull had multiple punched out lesions with non-sclerotic margins.

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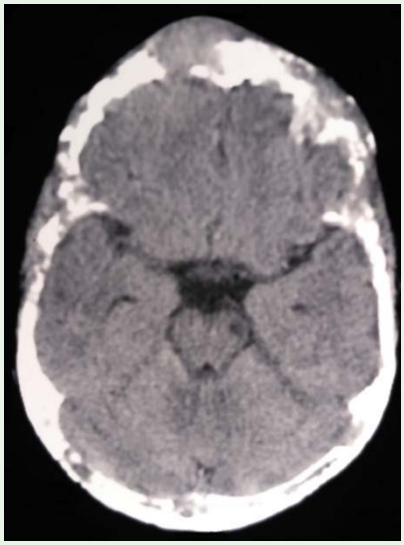

Figure 2: CT scan of head appeared as a meningocoele over his forehead.

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