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CASE REPORT

Langerhans Cell Histiocytosis of Cranium

Running Title: Histiocytosis of skull

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ABSTRACT

Introduction: Langerhans cell histiocytosis (LCH) is a rare idiopathic disease in which there is an accumulation of histiocytic cells in various tissues of the body. **Case Presentation**: We present a 3-year-old boy who reported bilateral swelling of the skull from one year. Even though Langerhans cell histiocytosis can happen in bones throughout the body, it especially happens in the skull, with a high frequency in the parietal and frontal bone. **Conclusion**: Our investigation varies to some extent from this finding in that in our patient there was a joint association of various skull bone injuries, not a singular one.

INTRODUCTION

Langerhans cell histiocytosis (LCH), a preferred term for the disease previously known as histiocytosis X, eosinophilic granuloma, Hand-Schüller-Christian disease, or Letterer-Siwe disease, is an uncommon entity involving histiocytes, recognized by heterogeneous lesions with an accumulation of CD207(+) histiocytic cells in many tissues of the body [1]. Langerhans cell histiocytosis (LCH) is a clonal proliferative disease featuring pathologic proliferating cells that show some characteristics of Langerhans cells [2]. It is most prevalent in the pediatric age, with a peak incidence between 1 and 3 years of age. There is a higher incidence in males, LCH commonly presents with osteolytic bone lesions and less commonly in the liver, spleen, lymph nodes, and bone marrow [3]. Limited data are available regarding the epidemiology of LCH, with an estimation of 2–5 cases per million inhabitants per year in the year 2009 [4]. Head and neck, particularly the base of the skull are involved in about 60% of the cases [5]. But the involvement of multiple bones of the skull is extremely rare. Almost no related articles reporting such characteristics have been published. The current study aims to illustrate the clinical presentations, diagnosis, and management of this disease. We report a male child with multiple osteolytic lesions of the skull.

CASE PRESENTATION

A 3-year-old boy from Karachi, Pakistan was brought by his father to the outpatient department (OPD) with a presenting complaint of swelling on both sides of the skull for one year. The swelling was gradual in onset, has increased in size over time, and was associated with pain. The pain was gradual in onset, intermittent in nature, non-radiating, and aggravates and relieves on its own. There was no past medical history of trauma or any other medical illness. He also denied any personal or family history of cancer. His father noticed some weight loss progress over the last year. No gross asymmetry or surface changes was noticed on clinical examination of the swelling, however, on palpation, a well-defined swelling was noticed on both side of skull which was hard and has an irregular surface, On general physical examination, the child was small for age, and mildly lethargic and his head circumference was larger than his age group. His vital signs on arrival were as follows: blood pressure, 120/80 mmHg; pulse, 70 bpm; respiratory rate, 18 breaths/minute; and axillary temperature, 36.5oC. On systemic review, there was no fever, chills, shortness of breath, abdominal pain, weakness, or paresthesias. We ordered a complete blood picture, metabolic profile, and urinalysis of the child, which was within normal limits. While in radiological investigations, an X-ray of the skull showed an osteolytic lesion of frontal and parietal bones (Figure1). Computed tomography (CT) scan of the brain with and without intravenous contrast revealed a 5-cm (AP dimensions) right parietal bone lesion with evidence of fluid level on current examination and left frontal bone small lytic lesion 3 x 2.3 x 5 cm AP x TR x CC dimensions involving the left orbital roof with intraorbital soft tissue extension abutting the globe. There was also a right infratemporal lesion eroding the greater wing of sphenoid superiorly with meningeal involvement, right zygomatic arch laterally, a posterior orbital wall with extraconal extension. Anteriorly, petrous part of sphenoid bone posteriorly infiltrating lateral pterygoid and temporalis

Figure1: Anterior, Anteroposterior and Lateral view radiograph of the skull shows multiple well-defined lytic lesions.

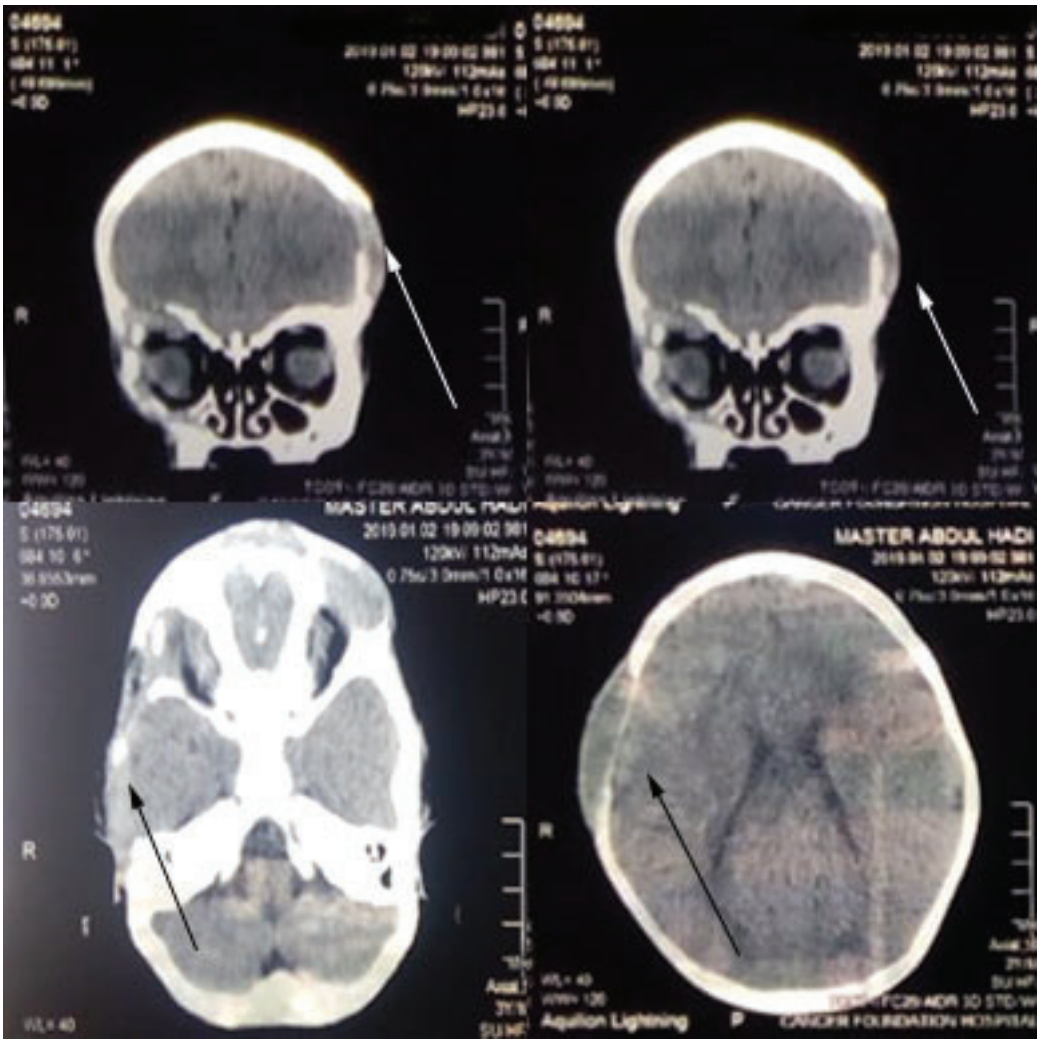


Figure 2: CT scan Brain demonstrates an osteolytic lesion in the right parietal and infratemporal Regions and also left frontal bone small lesion.

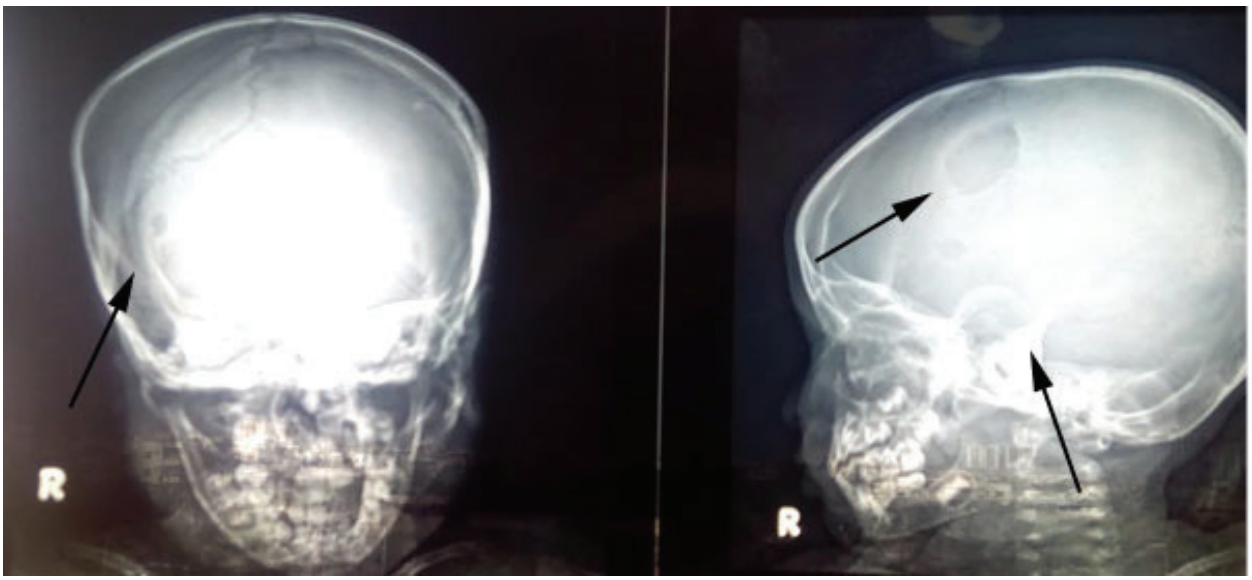
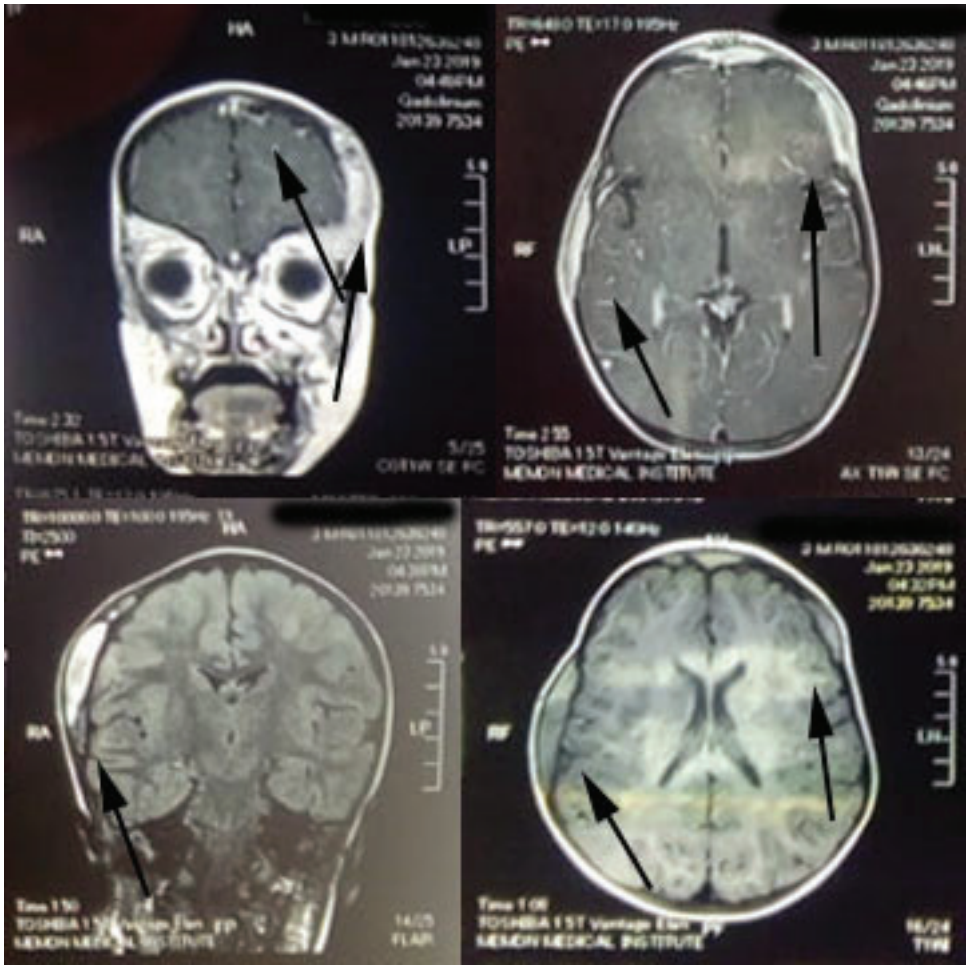


Figure3: MRI Brain an expansile mass in right parietal bone and heterogeneous lesion left frontal bone.



muscle. The soft tissue component measures 5 x 3 cm (Figure2). MRI Brain plain and contrast study was performed after one year which revealed an increase in right parietal bone lesion measuring 6.3 x 1 cm AP x TS and left frontal heterogeneous lesion which was extending into the left frontal sinus and also involving the lateral wall of the left orbit and supraorbital regions. It approximately measured 6.5 x 1.8 cm AP X TS which was slightly compressing the underlying brain parenchyma (Figure3). Computed Tomography (CT)-guided biopsy was performed. Two fine-needle aspiration and one core biopsy specimens were obtained from the parietal bone. Later, the examination of open biopsy samples revealed fragmented bone pieces of different sizes, which when studied microscopically, were composed of lymphocytes and macrophages, inconspicuous nucleoli, and slightly eosinophilic cytoplasm. The mentioned cells when checked for tumor markers, were found positive for S100 and CD1a. Based on the immunohistochemical and histopathological findings, a diagnosis of Langerhans cell histiocytosis (LCH) was reached.

DISCUSSION

LCH (histiocytosis X) is a malady complex that incorporates Letterer–Siwe illness, Hand– Schüller-Christian infection, and eosinophilic granuloma. While the previous two are foundational infections, the last is a limited type of histiocytosis. It is a non-neoplastic incessant illness of a granulomatous sort and obscure reason. Langerhans cell histiocytosis most often affects infants, while another study gave the predominant age to be 1-3 years [3,6]. Bone injuries are the most widely recognized appearances of LCH; they happen in 80%-95% of kids with LCH. A preference for hematopoietically dynamic medullary locales exists; the skull is now and again included [7]. Even though it can happen in any one of the entire body, it especially regularly happens in the skull, with a high frequency of an event in the parietal and frontal bone [8]. Our investigation varies to some degree from this finding in that in our patient there was a joint association of various skull bone injuries, not a singular one and these injuries were dynamic over a year without treatment as the patient was nonbearing. Treatment of LCH relies upon the degree of the sickness. Different types of treatment for a single lytic injury influencing bones have been endeavored, which incorporate curettage, nearby steroid infusion, radiotherapy, and chemotherapy alone or in a mix. The consequences of treatment of singular injuries are constantly acceptable, even though recurrence happens in certain patients [6].

CONCLUSION

The case of this male child described in the present case report suffered from frontal, parietal, and temporal bones LCH. Besides, the patient also progressed with an increase in the size of the lesion over one year without any kind of treatment. Although LCH is very critical, the prognosis is generally good if there is no vital organ involvement and if correct diagnosis and timely rational therapy are made. The indices of response, recurrence, and complications may vary due to the different schemes available.

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CONFLICTS OF INTEREST

None.

REFERENCES

1. Allen CE, Li L, Peters TL, Leung HC, Yu A, Man TK, et al. Cell-specific gene expression in Langerhans cell histiocytosis lesions reveals a distinct profile compared with epidermal Langerhans cells. J Immunol. 2010;184(8):4557–67.

2. Abla O, Egeler RM, Weitzman S. Langerhans cell histiocytosis: Current concepts and treatments. Cancer Treat Rev. 2010;36:354–359

3. Nicholson HS, Egeler RM, Nesbit ME. The epidemiology of Langerhans cell histiocytosis. Hematol Oncol Clin North Am. 1998;12(2):379–384.

4. Greenberg MS, Glick M, Ship JA. Burket's Oral Medicine. 11th ed. Hamilton: BC Decker; 2008. pp. 144–5.

5. Coleman MA, Matsumoto J, Carr CM, Eckel LJ, Nageswara Rao AA. Bilateral temporal bone langerhans cell histiocytosis: radiologic pearls. Open Neuroimag J. 2013; 7: 53-7.

6. Howarth DM, Gilchrist GS, Mullan BP, Wiseman GA, Edmonson JH, Schomberg PJ. Langerhans cell histiocytosis: diagnosis, natural history, management, and outcome. Cancer. 1999; 85(10):2278-90.

7. Bollini G, Jouve JL, Gentet JC, Jacquemier M, Bouyala JM. Bone lesions in histiocytosis X. J Pediatr Orthopaed. 1991;11(4):469-77.

8. Yunoki M, Hirashita K, Gohda Y, Yoshino K, Fujimoto S. An operative case of eosinophilic granuloma of the skull: Case report –special emphasis on surgical procedure, CP Neurosurg. 2007;17:358–64.