

## Langerhans' cell Histiocytosis of Temporal Bone

### -Case Report-

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### Abstract

A 3 year old child presented with a history of progressive abdominal distention associated with low grade fever and discharging left ear for the last 6 months. He was diagnosed as a case of chronic suppurative otitis media. He was not responding to the medical treatment. He then developed left post-auricular soft not tender swelling with aural granulation tissue and hepatosplenomegaly. Clinical and histological examination confirmed the diagnosis of Langerhans' cell histiocytosis. His condition responded very well to steroids and chemotherapy. This case report to highlight this condition as a possible cause of chronic suppurative otitis media and requires imaging and histological examinations for definitive diagnosis and appropriate treatment.

**Key words:** Langerhans cell histiocytosis (LCH), temporal bone, chronic suppurative otitis media, hepatosplenomegaly. Complete Blood Picture (CBP)

### Introduction

Langerhans' cell histiocytosis (LCH), formerly called histiocytosis X, is a condition that is characterized by the proliferation of histiocytes that share the characteristic of Langerhans' cells (normally located in the dermis). These cells are recognized by the presence of inclusion bodies in the cytoplasm, called Birbeck granules. Proliferation and infiltration of these cells may develop in any organ in the body, including bone, skin, lung, liver, spleen, and nervous system. <sup>(1)</sup> LCH has historically been divided into three clinical entities: eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease. Eosinophilic granuloma is the localized and most benign form LCH, whereas Letterer-Siwe disease is the most aggressive form. Hand-Schuller-Christian disease is characterized by cranial bone involvement with exophthalmos and diabetes insipidus, and has a more chronic and milder clinical course than Letterer-Siwe disease. All three disease share

similar histological features, and the disease concept of histiocytosis X was introduced in the early 1950s by Lichtenstein.<sup>(2)</sup> The term LCH was assigned to this disease by the Writing Group of the Histiocyte Society in the late 1980s.<sup>(3)</sup> LCH predominantly occurs in children and adolescents, but is relatively rare in adults. The most frequent site of occurrence is the bone. The temporal bone is involved in 15-61% of all LCH cases.<sup>(4)</sup> LCH may also arise in the skin, lymph node, soft tissue, liver, spleen, lung, thymus, and bone marrow. Symptoms and signs of ear and temporal bone involvement are often indistinguishable from that of chronic suppurative otitis media and may include a conductive or sensorineural hearing loss. Thirty percent of patients with temporal bone LCH have bilateral involvement. The characteristic radiological appearance of the temporal bone is that of a 'punched out' or lytic appearance. While extensive destruction of the temporal bone can take place, there are only limited reports of otic capsule involvement with disease often confined

to the external and middle ear.<sup>(1)</sup> When soft tissues and the reticulo-endothelial system are involved, it can present with fever, lymphadenopathy, hepatosplenomegaly, pancytopenia. or skin lesions.<sup>(3)</sup> The ultimate prognosis of the patients with LCH depends on whether the disease is uni-focal, multi-focal or disseminated. Age at presentation of less than two years

with organ dysfunction is an indicator of a much poorer prognosis. Uni-focal disease, on the other hand, has a very good prognosis responding to local treatment, which may include local resection or radiotherapy. Most series have a 95-100 percent cure rate. Multifocal disease also has a good response to treatment, but disseminated disease can be fatal.<sup>(1)</sup>

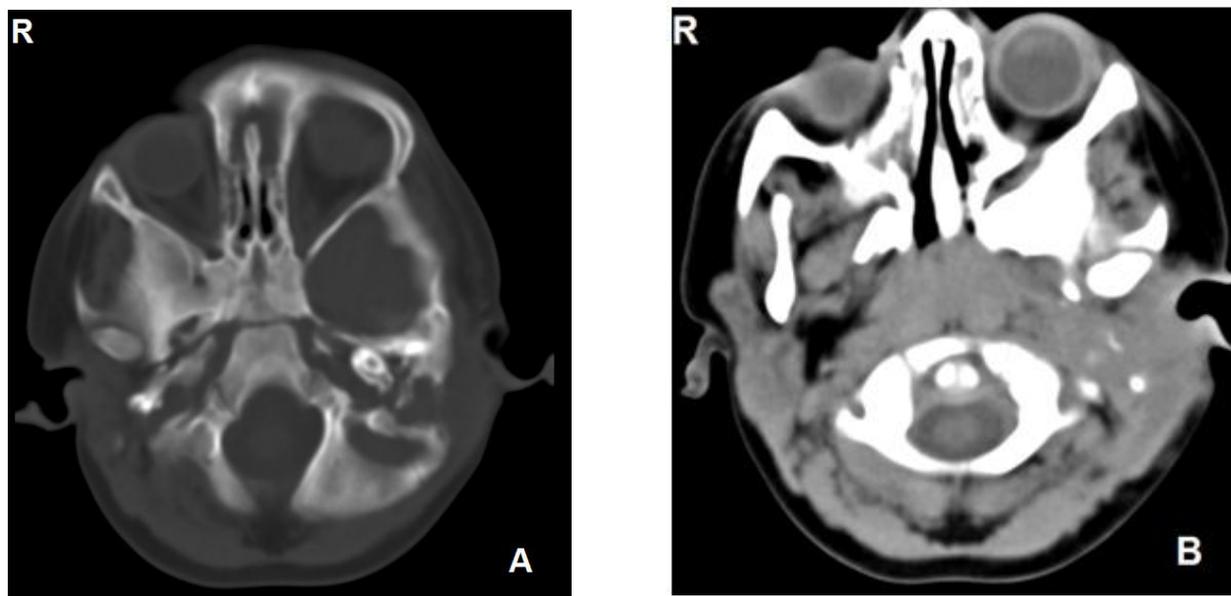


Fig.1 (A,B): computerized tomography scan

## Case Report

A 3 year old child presented with a history of progressive abdominal distention with a low grade fever and discharging left ear for the last 6 months not responding to medical treatment. Examination revealed evidence of chronic suppurative otitis media with tympanic membrane perforation and granulation tissue.

General examination showed hepatosplenomegaly. There was no lymphadenopathy. Later he developed left post-auricular swelling, soft, not tender, with no erythema of the skin. The child had no fever, and CNS examination was normal. CBP, liver function tests, blood urea, serum creatinin, serum electrolytes, and blood sugar were all normal. He had negative Comb's test and his ESR was 28mm/hr. The CT scan showed enhancing soft tissue mass in the left mastoid and

petrous temporal bone. The mass is causing local bone destruction with involvement of the middle ear cavity and left lateral semicircular canal. It extends to the left post-auricular area and posterior cranial fossa near the left sigmoid sinus. The middle ear ossicles are surrounded by the lesion but not destroyed. Doppler shows moderate vascularity. The picture is of a destructive tumour mass rather than cholesteatoma (Fig.1). US guided FNA from the left mastoid bone mass revealed a smear of adequate cellularity, polymorphic cell population with predominance of round-polygonal cells with adequate cytoplasm and kidney shaped nuclei some with nuclear groove (Langerhans like cells), variable mixture of inflammatory cells, with eosinophilic components and degenerated background. The picture is highly suggestive of Langerhans histiocytosis (Fig.2). A biopsy was taken

from the external meatus granulation tissue to confirm the diagnosis, and it was positive for S100, CD1a tumour markers (Fig.3). The patient was referred to the pediatric oncology unit and a bone marrow biopsy was done. The child developed a hard mass (3x3 cm) in left frontal area in this period. He was given chemotherapy (vinblastine, etoposide, and

prednisolone) for 6 weeks as the initial treatment. His condition improved, the frontal mass disappeared in the third week of his treatment. His maintenance treatment then continued for 19 weeks with vinblastine, etoposide, prednisolone, 6MP, and co-trimoxazole as per the protocol.

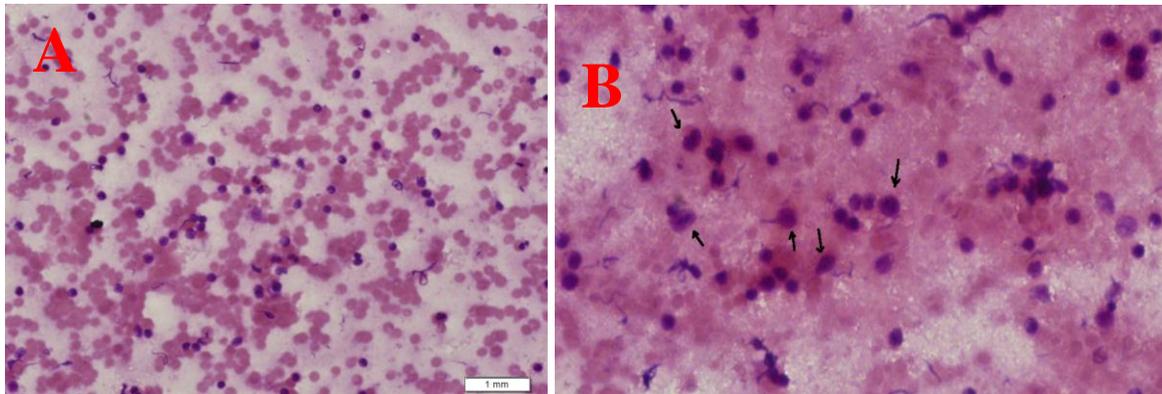


Fig.2 FNA cytology A: smear show mixed inflammatory cells in the background with characteristic Langerhans cells X 200. B: smear show the characteristic Langerhans cell with pale cytoplasm and kidney shape nucleus (arrows) X400.

## Discussion

There are three key subtypes of LCH: uni-focal, multi-focal, and systemic LCH. Uni-focal LCH (eosinophilic granuloma) is the most common. Bones are mostly affected, usually the skull. Uni-focal LCH is found most commonly in older children and adults and presents as a very aggressive tumor, with large areas of tissue destruction and rapid expansion, but still carries an excellent prognosis due to its confined and self-limiting growth.<sup>(5)</sup> Multi-focal LCH (Hand-Schuller-Christian disease) involves multiple sites and is most common in young children.<sup>(3)</sup> Lastly, diffuse (systemic) LCH (Letterer-Siwe disease) is typically found in newborn babies and carries a very poor prognosis. When systemic disease causes organ dysfunction, mortality can be as high as 50%.<sup>(6)</sup> There are different ways to treat the uni-focal LCH and there have been four approaches described in the literature. The first option is not to give any treatment at all, because some

eosinophilic granulomas are self-limiting and spontaneously regress.<sup>(7, 8)</sup> We think this option is risky, because we feel a biopsy is warranted to rule out other differential diagnosis of concern. The second option is to attempt at least a partial resection or to perform a complete excisional biopsy of the lesion. The third option is a biopsy, followed by low-dose radiation to the lesion with 6–10 Gy with a local control rate of approximately 80%.<sup>(9)</sup> The fourth option, (in uni-focal eosinophilic granuloma) can be treated with intra-lesional corticosteroids, usually methylprednisolone, 30-125 mg to limit the immunoresponse, followed by sequential imaging.<sup>(6,10,11)</sup> Finally, systemic chemotherapy may be indicated when LCH progresses systemically or is recurrent. A popular agent is 2-chlorodeoxyadenosine, which can block cell proliferation in histiocytes and eosinophils.<sup>(12)</sup> Tumor recurrences typically occur within 2 years of the primary diagnosis, reports have been made of LCH tumors recurring up to 16 years

later, which advises us to follow these patients long-term with serial imaging.<sup>(13)</sup>

Systemic chemotherapy was recommended in our patient because his

condition had progressed systemically and there was multifocal skull lesions, temporal and frontal

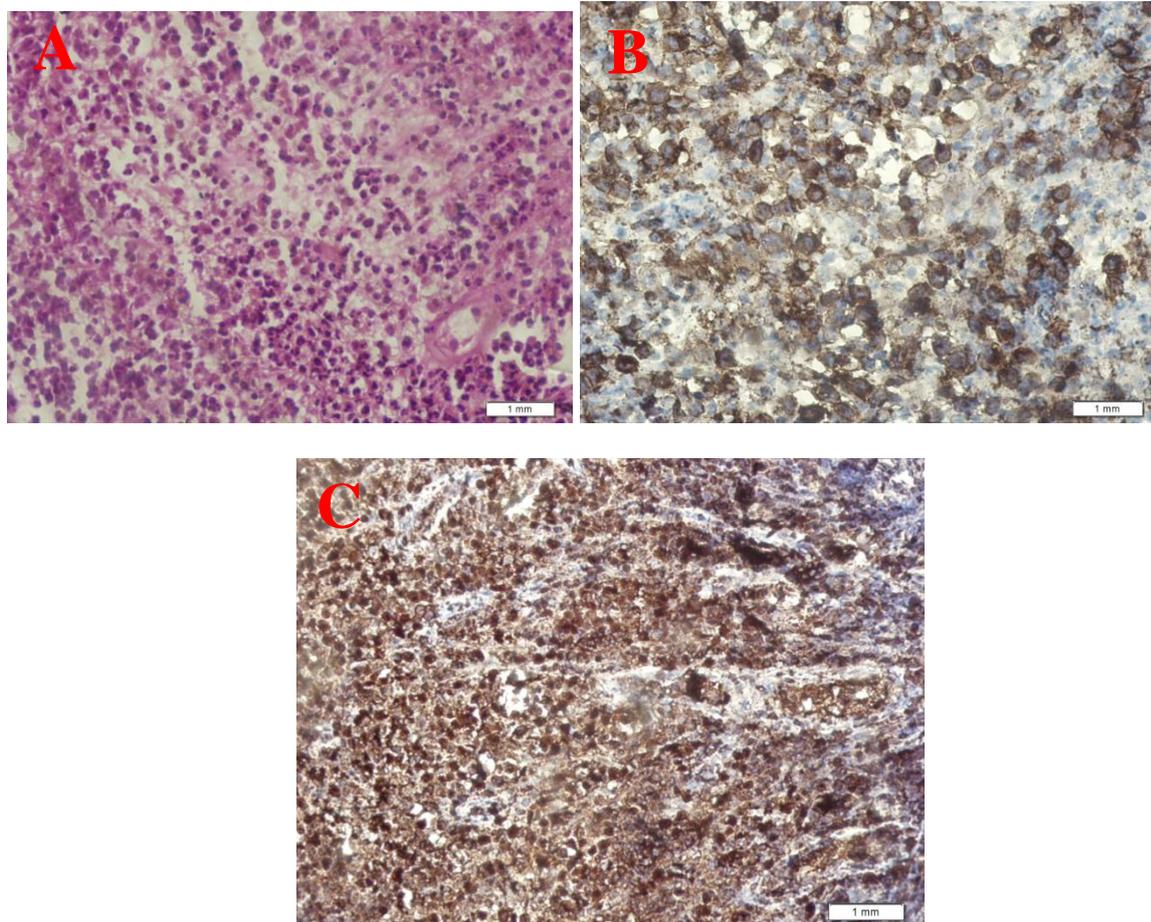


Fig.3 A: photomicrograph for tissue section show diffuse tissue infiltration by mixed inflammatory cells rich in eosinophils and Langerhans cells. B,C: histiocytic positive for CD1a and S-100 protein.

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