Langerhans cell histiocytosis (LCH) is a rare proliferative disorder with unknown etiologies. Immunoregulatory defects leading to abnormal maturation and migration of the Langerhans cells to the central nervous system, lungs, skin, bone, bone marrow, lymph nodes, thymus, liver, and spleen were postulated to be the causative factors for LCH. The incidence of LCH is about five to six cases per one million children. Males are more frequently affected than females, and the average age of presentation varies from a few months to 15 years. It can present as solitary or multifocal lesions and may involve multiple systems. Head and neck involvement is found in almost 60% of cases and is the most common site for LCH. The incidence of LCH is about five to six cases per one million children. Males are more frequently affected than females, and the average age of presentation varies from a few months to 15 years. It can present as solitary or multifocal lesions and may involve multiple systems. Head and neck involvement is found in almost 60% of cases and is the most common site for LCH.

Clinical examination revealed a firm, warm, tender, and erythematous swelling over the left postauricular region [Figure 1]. The swelling measured 2 × 2 cm and displaced her left pinna anteroinferiorly. Bilateral otoscopic examination displayed granulation tissue mixed with a copious amount of mucopurulent discharge, which was obscuring the tympanic membrane view. She also had a small ulcerative lesion on the hard palate. Multiple firm, non-tender subcentimeter cervical lymph nodes were palpable bilaterally.

CT imaging was suggestive of bilateral mastoiditis with collections over the left mastoid region [Figure 2]. The swelling measured 2 × 2 cm and displaced her left pinna anteroinferiorly. Bilateral otoscopic examination displayed granulation tissue mixed with a copious amount of mucopurulent discharge, which was obscuring the tympanic membrane view. She also had a small ulcerative lesion on the hard palate. Multiple firm, non-tender subcentimeter cervical lymph nodes were palpable bilaterally.

Concurrent Mastoid Cellulitis and Langerhans Cells Histiocytosis: A Challenging Diagnosis

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare proliferative disorder, which commonly arises in the bone and may involve other systems. To date, the diagnosis of temporal bone LCH remains a challenge as it may masquerade as a common ear infection. We report a case of a child who presented to us with persistent bilateral ear discharge for four months and was not responding to treatment. Her condition subsequently worsened, with clinical features and radiological findings suggestive of mastoid cellulitis. Nevertheless, further histopathology study revealed LCH.
a soft tissue mass occupying the mastoid cavity with serous fluid collection. Tissue biopsies taken from the mass, external ear canal, and the hard palate eventually confirmed the diagnosis of LCH with the detection of CD1a and S-100 protein [Figure 3]. Treatment modalities, including chemotherapy, were discussed. Unfortunately, her parents refused further intervention and opted for conservative management. They did not present for follow-up subsequently.

**Differential diagnosis**

A differential diagnosis of mastoiditis and cholesteatoma with mastoid abscess was made. The patient, unfortunately, succumbed to the illness a few weeks after the diagnosis. The cause of death was not ascertained as the family members refused a post-mortem.

**DISCUSSION**

LCH has been well known to mimic a variety of common diseases and misdiagnosis is not uncommon.⁷ There is a wide clinical spectrum, ranging from isolated lesions to multisystem involvement with the bone tissue most commonly affected.⁵ Moreover, studies have shown that bone marrow involvement is more likely to occur in patients with multisystem involvement resulting in cytopenia.⁵ Our patient had bilateral temporal bone involvement and hard palate involvement. Oral involvement of LCH usually presents as swelling or ulceration, with or without the involvement of the underlying bone.⁷

The presence of atypical otitis media, otorrhoea, aural polyps, granulations tissues, or a postauricular mass should alert to the possibility of LCH.⁶ Our patient presented initially with intermittent bilateral otorrhoea for four months and therefore was treated as a case of otitis media. However, she was resistant to treatment. She subsequently developed a left

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**Figure 1:** Swelling over the left postauricular region suggestive of acute mastoiditis or mastoid abscess.

**Figure 2:** (a) Axial and (b) coronal view of the computed tomography scan done on the patient showing an abscess formation on her left temporal region with erosion.
postauricular swelling with spiking fever that was suggestive of mastoid cellulitis. CT imaging revealed bilateral temporal bone involvement with sparing of the bony labyrinth, which is consistent with the diagnosis of LCH. Another important feature that may be strongly suggestive of LCH, is the presence of lytic or ‘punched out’ lesions of the temporal bone, which was not seen in our case. Magnetic resonance imaging (MRI) should be employed following CT imaging for further characterization of the lesion and to evaluate intracranial involvement. MRI was not done in this case due to the request of the family members for conservative management. The diagnosis of LCH was made based on histopathology results. It was supported by the detection of CD1a and S-100 protein. These markers are specific and helpful in differentiating LCH from granulomatous osteomyelitis or malignant lymphoma, which have similar histological properties. This implies that clinical and imaging tools are not adequate for accurate diagnosis and histopathological studies play an important role in the diagnosis.

Various treatment modalities have been used for LCH including close monitoring, surgical intervention, local steroids injections, high-dose systemic corticosteroids, low-dose radiotherapy, chemotherapy, bone marrow transplantation, and antibody therapy. Nonetheless, the combination of steroids and vinblastine has been accepted internationally as the standard treatment for LCH, even in patients with multisystem involvement. Generally, the overall prognosis is good but patients with multisystem involvement and active disease, such as seen in this case, tend to have a poorer outcome. In fact, the Histiocyte Society has categorized LCH patients into low- and high-risk groups, based on the outcomes related to the extent and the site of the LCH lesions. Patients with lungs, liver, spleen, and bone marrow involvement are considered to have higher risks and poorer outcomes. Furthermore, the presented case had features of tegmen tympani erosion, which is suggestive of a central nervous system risk lesion. This predisposes the child to severe, irreversible neurodegenerative disease.

Obtaining a diagnosis of LCH can be challenging as its presentation may mimic a large number of diseases. In its early stages, a child may present with non-specific symptoms. Furthermore, tumors of the middle ear and temporal bone are uncommon and may be missed. A strong index of suspicion is important especially in children who present atypically for common diseases and are resistant to preliminary treatment. Timely diagnosis may ensure a better outcome as the prognosis of LCH is good when detected early.

**CONCLUSION**

The diagnosis of LCH should be considered in children as a differential when they present with chronic otorrhea with the presence of a mass or granulation tissue, which does not respond well to conventional medical treatment. Full systemic examination is warranted in children who do not respond to medical treatment to exclude...
multisystemic involvement of the disease and to detect if the child is immunocompromised. As CT imaging poses children to radiation exposure, MRI may be considered as an alternative. A deep tissue biopsy is indicated in suspicious lesions or cases resistant to treatment.

Disclosure
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