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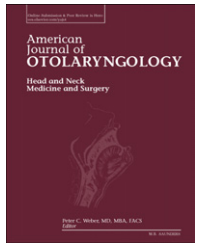

A rare disorder mimics otitis media: Langerhans cell histiocytosis of the temporal bone in a child with interstitial pulmonary fibrosis☆

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare disease ranging from a benign to a rapidly fatal condition affecting young children predominantly, and is characterized by an abnormal clonal proliferation of Langerhans cells. We report a case of a 3-year-old child presenting with a 1-year history of otorrhea and otorrhagia followed by a 6-month history of postauricular swelling in the right ear. Imaging demonstrated a large mass of organized tissue. A biopsy was conducted, and the diagnosis of LCH was confirmed by histopathological and immunohistochemical examination. The child was treated with a 12-month course of vinblastine chemotherapy with prednisolone. No clinical evidence of recurrence was noticed after 3 years of follow-up. This rare case highlights the importance for otolaryngologists to keep LCH in mind for differential diagnosis in very young patients with symptoms and signs suggestive of acute mastoiditis or chronic otitis media.

1. Introduction

Langerhans cell histiocytosis (LCH) is characterized by an abnormal clonal proliferation of Langerhans cell in single or multiple organs [1]. It is generally considered to include a spectrum of disorders as eosinophilic granuloma (EG), Hand–Schüller–Christian (HSC) disease and Letterer–Siwe (LS) disease which vary in the prognostic results [2]. The incidence of LCH is around 5.4 per million with a male predominance [3]. Its peak incidence period is noted in children aged from 1 to 4 years old, however, it can present at any age from the neonatal period to old age [4,5]. Inflammation, autoimmunity and loss of controlled proliferation of Langerhans cells are the assumed etiologies [5–7]. The clinical presentation of LCH is very heterogeneous, ranging from a single-system involvement with bones and skin as the most commonly involved sites, generally benign [8], to a multisystem life-threatening disease. The head and neck are the most common site of involvement, occurring in approximately 60–75% of LCH patients [8,10]. Moreover, the temporal bone is involved in 19–25% of cases and the involvement is bilateral in a third of all cases [9,10]. Head and neck manifestations are diverse and

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include cervical lymphadenopathy, skin rush, skull and temporal bone lesions, lung and larynx lesions [11,12]. Diagnosis can be difficult as these lesions mimic other common conditions seen by the otolaryngologist, including otitis media, otitis externa, acute mastoiditis, and gingivitis.

We present a case of a child at the age of 3 years old who developed LCH at the temporal bone with interstitial pulmonary fibrosis, mimicking as chronic suppurative otitis media.

2. Case report

A 3-year-old boy was referred to the Otolaryngology Head and Neck Surgery Clinic of the Second Xiangya Hospital, complaining of a 1-year history of otorrhea and otorrhagia followed by a 6-month history of postauricular swelling in the right ear. One year ago, he was diagnosed with interstitial pulmonary fibrosis, however, no therapy was conducted for the disease. Physical examination disclosed an obvious swell on the right pars temporalis (Fig. 1). Measuring 4 × 7 cm, the swell was soft with no tenderness, redness nor ulcer. There was a kermesinus discharge from the right ear and an apparent red mass in the right external auditory canal (EAC). Also, a large amount of cerumen was observed in the left EAC. The patient did not present any paresis of facial nerve. There was no history of trauma or inflammation in the region, and the medical history was noncontributory.

During the hospital stays, relevant examinations were performed on this patient. An electrocardiogram was unremarkable. Complete blood count, coagulant function and serum electrolytes serology were normal. His hearing level (trichotomy) was 20 dB for the left ear and 50 dB for the right ear. On ultrasound, no abnormality was observed in liver, gallbladder, pancreas, spleen, portal vein and kidney. A chest X-ray displayed diffuse high density shadows with hazy border in the bilateral upper and middle lung field, multiple thin-walled cystic shadows in the middle and lower lung field, and a fuzzy pulmonary hilum (Fig. 2A). A computed tomography (CT) scan showed a large soft tissue mass in the right mastoid region, destroyed right EAC, and extensive destruction of the right mastoid bone. Meanwhile, CT scan also revealed that for the left ear, soft tissue in the mastoid cavity, thickening tympanum, diploic mastoid, and bone destruction of mastoid antrum as well as tegmen tympani were observed (Fig. 2B). Magnetic resonance imaging (MRI) demonstrated a regular-shaped soft-tissue mass in the right mastoid region with a relative defined boundary. The mass presented as a mix of isointensity and hyperintensity on T1-weighted images, hypointensity on T2-weighted images, and was moderately enhanced with a size of 5.0 × 4.5 × 2.0 cm. The right temporal lobe was compressed. Additionally, a lesion in the left mastoid cavity was observed with an isointensity on both T1 and T2-weighted images, and it was significantly enhanced (Fig. 2C–E).

Fig. 1 – The postauricular swelling in the right ear of a 3-year-old child.

The child was scheduled for resection of the lesion in the right pars temporalis under general anesthesia. Through microotoscopy, a large amount of granulation tissue in the right EAC was observed, which was crisp and easily bleeding with abundant necrotic tissue. Histopathological study of the material obtained confirmed the diagnosis of LCH (Fig. 3). The mass was infiltrated by large histiocytes which had oval nucleus with longitudinal furrow, increased lymphocytes and eosinophils. Necrotic lesions were also observed. Immunohistochemistry showed positive staining of the cells with CD1a (Fig. 4A) and S-100 (Fig. 4B), confirming the diagnosis of LCH.

As soon as the diagnosis of LCH has been confirmed, the patient was referred to the pediatric oncology department for therapy where he was treated with a 12-month course of vinblastine chemotherapy with prednisolone. After 3 years of follow-up, the patient showed no clinical evidence of recurrence, with a slightly improved hearing level (trichotomy) of 20 dB for the left ear and 35 dB for the right ear.

3. Discussion

Since 1987, LCH has become the accepted name to describe the collection of conditions previously individually known as EG, HSC and LS diseases [1]. The clinical presentation of LCH varies greatly, ranging from a single-system involvement to a multisystem life-threatening disease. EG is a benign tumor-like condition of the disease, characterized by single or multiple benign osteolytic lesions [3]. HSC disease is a chronic and multifocal diffuse disease consisting of multiple osteolytic and soft tissue lesions, with a classic 'Christian triad' of diabetes insipidus, cranial bony lesions and exophthalmos [12,13]. LS disease is an acute, fulminant and rapidly fatal condition, characterized by extensive skin eruptions, erosive osteolytic lesions, pulmonary infiltrations, anemia, fever, thrombocytopenia, lymphadenopathy and hepatosplenomegaly [12].

The etiology of LCH remains unknown but recently there have been some assumptions. Over the years, LCH has been