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Case Report

Cholesteatoma Masquerading as Recurrent Langerhans Cell Histiocytosis

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Langerhans cell histiocytosis is a rare condition affecting the temporal bone in up to 60% of cases. Symptoms are non-specific and the differential diagnosis includes infection, benign lesions such as cholesteatoma, and malignant lesions of the skull base. Here, we report the case of a 14-year-old child referred with chronic ear discharge, and background of multifocal Langerhans cell histiocytosis 9 years prior. Recurrence of Langerhans cell histiocytosis was initially suspected and systemic treatment was considered. Further imaging workup and surgical exploration of the mastoid showed a secondary acquired cholesteatoma arising from a dehiscent posterior ear canal wall. Surgical removal of the cholesteatoma was performed with a canal wall down procedure. We review the presentation and management of temporal bone Langerhans cell histiocytosis. We recommend that cholesteatoma should be considered in case of recurrence of otological symptoms in patients with a background of Langerhans cell histiocytosis.

KEYWORDS: Children, neuro-otology, lateral skull base, temporal bone

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare condition related to the abnormal proliferation of Langerhans cells. The presentation can be unifocal or disseminated. Diagnosis is often delayed for temporal bone localization of this disease. Symptoms include otitis media, otorrhea, otalgia, hearing loss, external auditory canal polyp, and mastoiditis. Magnetic resonance imaging (MRI) and computed tomography (CT) scans are useful for diagnosis, and the clinician should have a high index of suspicion if imaging reveals osteolytic lesions with soft tissue mass. Diagnosis should be confirmed by biopsy sampling and immunohistochemistry.[1]

Differential diagnosis includes infection of the skull base, benign lesions (cholesteatoma), and malignant neoplasms (lymphoma, sarcoma, metastases).

Prognosis is good, especially in unifocal diseases. Response to chemotherapy in the first few weeks is among the most important prognosticators.

Here, we report the case of a 14-year-old child with limited multifocal LCH affecting the mandible and temporal bone with cutaneous and gastrointestinal tract involvement. She was in complete remission for more than 9 years when she presented with a recurrence of unilateral ear symptoms. An initial diagnosis of LCH relapse was considered, but further biopsies and surgical exploration showed a secondary acquired cholesteatoma.

CASE PRESENTATION

A 14-year-old patient was referred to our ENT department with a 1-year history of left-sided otorrhea. She had no other otological symptoms (no hearing loss, otalgia, tinnitus, or vertigo).

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She had a medical history of LCH diagnosed at 3 years. The initial disease was multifocal with mandible, temporal bone, skin, and gastrointestinal tract involvement. We reviewed her charts and initial imaging done at that time (Figures 1 and 2). Imaging at the time of initial LCH diagnosis with a CT scan showed extensive bony erosion of the lateral skull base (Figure 1). Magnetic resonance imaging demonstrated soft tissue mass with hyperintensity in T1 with contrast images (Figure 2).

Biopsies were obtained on colonoscopy and results were in keeping with LCH.

She was treated with induction chemotherapy (Vinblastine) and steroids followed by Cytarabine and Cladribine. She was then considered in complete remission. The only sequelae of the disease were the absence of several teeth due to mandibular involvement. She subsequently underwent dental implant insertion for rehabilitation.

On current presentation, physical examination showed a defect in the posterior ear canal wall on the left with purulent otorrhea. Tympanic membranes were normal, and she had normal hearing on both sides.

High-resolution CT scan of the temporal bone was obtained (Figure 3). It showed a posterior external ear canal defect opening into the mastoid with soft tissue density in the mastoid air cells. The middle ear and ossicular chain were reported as normal. A skull base MRI was obtained (Figure 4). It showed a partial soft tissue filling of the mastoid on that side with T2 hyperintensity. Diffusion-weighted images could not be performed due to dental restoration artifacts. A biopsy was performed in the outpatient clinic and showed inflammatory tissues. The microbiology sample was non-contributive.

An initial diagnosis of recurrent LCH of the temporal bone was made, and the case was discussed in a multidisciplinary meeting. Given the unusual timeframe of the recurrence of symptoms, we proceeded to a mastoid exploration with a postauricular approach.

At surgery, a typical aspect of cholesteatoma extending from the external ear canal into the mastoid cavity was found. The frozen section histology confirmed stratified squamous epithelium in the mastoid cavity in keeping with cholesteatoma. There was no evidence of LCH.

A canal wall down tympanoplasty with complete removal of the cholesteatoma was performed. Final histology showed no recurrence of LCH.

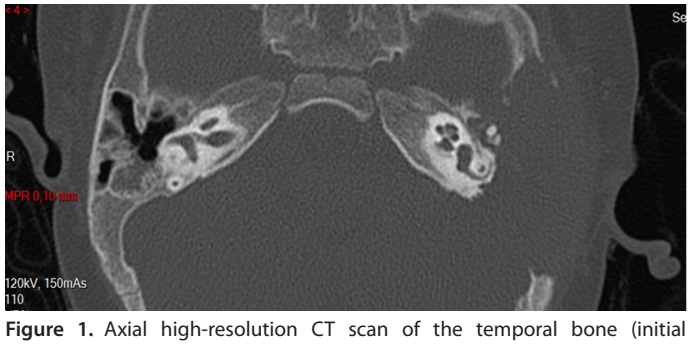

Figure 1. Axial high-resolution CT scan of the temporal bone (initial presentation). There is extensive bony destruction in the left lateral skull base. The otic capsule is intact. CT, computed tomography.

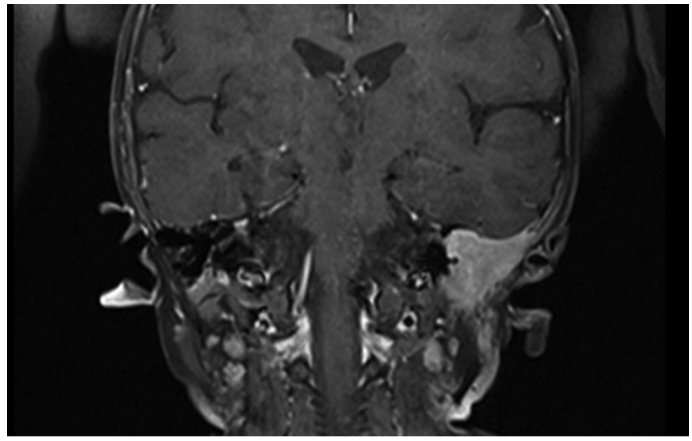

Figure 2. Coronal MRI of the skull base, T1 with contrast (gadolinium) (initial presentation). It shows a soft tissue mass in the lateral skull base of the left. It demonstrates T1 hyperintensity enhanced with contrast. MRI, magnetic resonance imaging.

Informed consent was obtained from the patient involved in this report.

DISCUSSION

Langerhans cell histiocytosis is a rare condition with a broad spectrum of clinical presentations. Langerhans cell histiocytosis can present as single organ system or multiple organ system diseases. Within an organ system, there can be one or multiple lesions. The unifocal single-system disease accounts for 70% of LCH cases.[2]

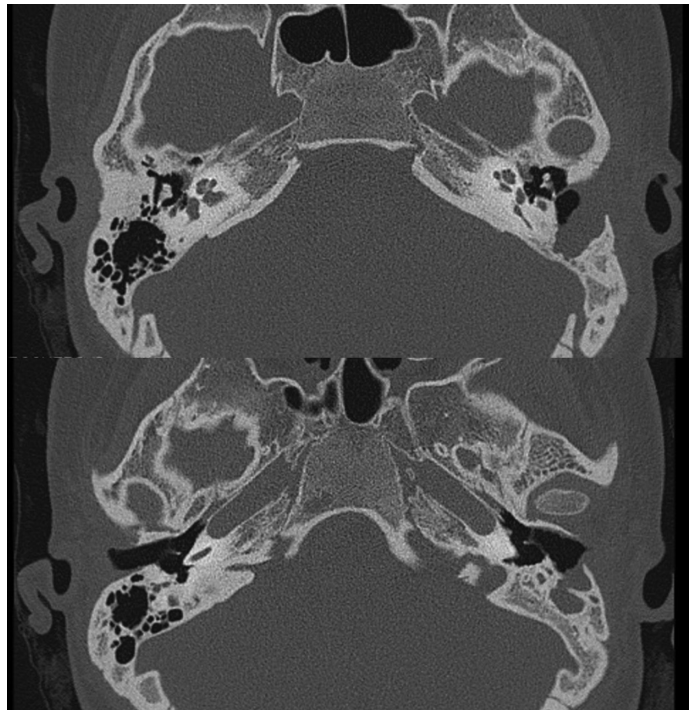

Figure 3. Axial high-resolution CT scan of the temporal bone. The middle ear and tympanic membrane appear to be intact. There is a defect in the posterior wall of the external ear canal, the mastoid is filled with a soft tissue mass. Of note, the temporal bone lateral to the otic capsule has mostly re-appeared with growth since the initial CT scan at the time of the primary LCH diagnosis (Figure 1). CT, computed tomography; LCH, Langerhans cell histiocytosis.

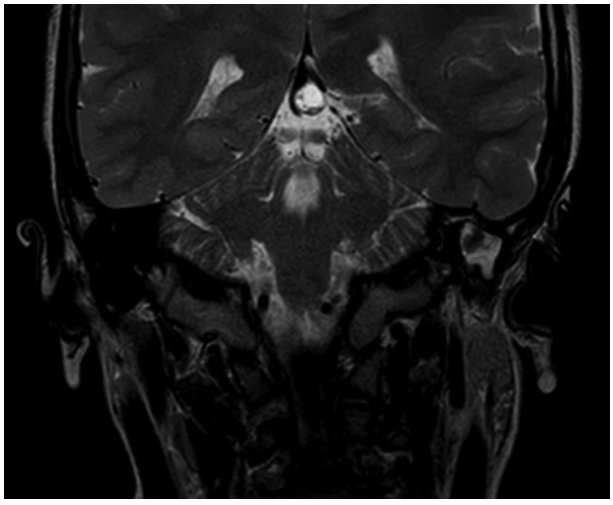

Figure 4. Coronal MRI of the skull base, T2-weighted. The soft tissue mass in the left mastoid is non-specific with heterogeneous T2 hyper- and hypointensity. MRI, magnetic resonance imaging.

Although all populations can be affected, it is most common in children with a peak incidence between 1 and 3 years of age. The incidence is estimated at 4-5 children per million subjects.[3]

Langerhans cell histiocytosis affects the temporal bone in 15% to 60% of cases, usually the mastoid, middle ear, or external ear canal. Temporal bone LCH is usually misdiagnosed at initial presentation as symptoms are non-specific: purulent otorrhea, otalgia, conductive hearing loss, and sometimes mastoiditis.

Computed tomography scan should be the first imaging investigation and usually demonstrates bony destruction and dense soft tissue. Magnetic resonance imaging is considered second-line imaging and is indicated to evaluate for further disease or rule out differential diagnosis.

Definitive diagnosis relies on biopsy results. Langerhans cell histiocytosis is defined as inflammatory myeloid neoplasia. Histologically, it consists of multinucleated Langerhans cells with assorted eosinophils, neutrophils, and lymphocytes. Electron microscopy can reveal characteristic lesions known as Birbeck granules. Immunohistochemistry shows positivity for CD1a and or CD207 (Langerin), 2 components of immature dendritic cells.[4]

Langerhans cell histiocytosis has a very good prognosis of up to 100% survival rate at 5 years in single system disease and 98% in multisystem disease. Treatment options include surgery, chemotherapy, and in some cases targeted therapy with BRAF inhibitors.[5] Unifocal disease is often treated with surgical excision sometimes followed by adjuvant radiotherapy. Multifocal disease should be treated with systemic therapy.[3] Vinblastine is the first-line chemotherapy agent. Temporal bone LCH is considered at risk of central nervous system involvement and systemic therapy is therefore considered as the standard of care.[2]

In 2009, Roger et al[6] were the first to describe 3 cases of secondary acquired cholesteatoma in patients previously treated for temporal bone LCH. Their 3 cases had a bony defect in the posterior ear canal wall secondary to LCH involvement. In all cases, patients presented with a reappearance of otorrhea and initial suspicion of recurrent LCH. In 1 case, the patient received a course of chemotherapy before the correct diagnosis was made.

Our observation correlates with this series. In case of recurrence of otologic symptoms in a patient with a background of LCH, there must be a high suspicion index for the risk of secondary acquired cholesteatoma.

In our case, we suspect that the posterior ear canal wall defect allowed for the migration of the skin into the mastoid. Pathogenesis of secondary acquired cholesteatoma after LCH could also involve metaplastic changes in the mastoid mucosa leading to cholesteatoma.

Clinical examination of the external ear canal and tympanic membrane is paramount, if required this can be performed under general anesthesia in younger patients. We also recommend early CT scan and diffusion-weighted MRI.

Histological confirmation of LCH should be obtained before systemic treatment is resumed.

In case of lytic lesions of the posterior ear canal wall following treatment of LCH, clinical follow-up in an otology-skull base clinic is advised.

CONCLUSION

Patients treated for temporal bone LCH should be followed up in a specialist neuro-otology and skull base clinic. In case of recurrence of otological symptoms, secondary acquired cholesteatoma and LCH recurrence are the 2 main differential diagnoses. Clinical examination and high-resolution CT scan of the temporal bone should be performed. Tissue samples with histology and immunohistochemistry should be obtained to guide treatment. In the case of secondary acquired cholesteatoma, systemic treatment is not needed and surgical management is indicated. Surgery allows for cholesteatoma removal with either canal wall reconstruction or canal wall down mastoidectomy procedure.

Informed Consent: Informed consent was obtained from the patient involved in this report.

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