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Case Report

Langerhans Cell Histiocytosis of Bilateral Mastoid Cavity

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Langerhans cell histiocytosis (LCH) is a rare disease, which may involve various organ systems; therefore, it has multiple clinical manifestations. Case report: We present the case of a 56-year-old woman admitted to Amerikan Hospital Ear-Nose and Throat outpatient clinic with a complaint of progressive hearing loss in both ears, which had started 10 years ago. She was treated with corticosteroids for 10 years until last year, 2017. Surgical exploration was performed and histologic evaluation revealed LCH. Conclusions: LCH has clinical manifestations depending on the site of infiltration. In adults, isolated bilateral mastoid infiltration, as an initial symptom, is a very rare condition. With corticosteroid uptake, the period of initial symptom was 10 years in our patient, which is, as per our knowledge, the longest reported in literature. This infiltration may mimic acute or chronic infections of the ear. Therefore, LCH should be considered in the differential diagnose of patients who present with bilateral mastoid cavity disease.

KEYWORDS: Langerhans cell, histiocytosis, mastoid

INTRODUCTION

Langerhans cell histiocytosis (LCH) is an uncommon disease of unknown pathogenesis. LCH originates from bone marrow-derived histiocytosis called Langerhans cells. The Langerhans cell's clonal myoproliferation leads to the infiltration of different organs. The clinical presentations of LCH vary depending on the site of involvement [1].

Here, we report a very rare case of LCH in an adult, which manifested with only ear complaints, was influenced by patient's corticosteroid uptake, and was ongoing for 10 years. This case is important for considering LCH in the differential diagnose of bilateral mastoid cavity diseases and to understand the clinical aspect of LCH.

CASE PRESENTATION

A 57-year-old woman presented with hearing loss in both ears for 10 years, which worsened in the last 6 months. She had a diagnosis of rheumatoid arthritis for 11 years, which was treated with corticosteroids for 10 years. She self-stopped the usage of oral corticosteroid tablets 9 months ago. She had a history of Hashimoto's thyroiditis and regular levothyroxine tablet intake. She was a non-smoker, and her family history was unremarkable.

Upon examination, the external ear canals and tympanic membranes were edematous. There was no pain in the mastoid areas with palpation. In an audiological evaluation, the tympanograms of both ears were of type B and pure tone averages were 25 dB and 23 dB for the left and right sides, respectively, with a mild conductive component on both sides.

Computed tomography (CT) scan of the temporal bone revealed soft tissue lesions with mastoid cortex erosion in both ears (Figure 1).

Complete blood count results were in normal ranges. Exploration for the right mastoid cavity was performed. During the operation, the external ear canal was observed as partially eroded, the mastoid cortex was eroded, and the cavity was invaded with soft tissue mass; the bone overlying the sigmoid sinus and dura was also partially defective because of the same soft tissue (Figure 2). Frozen sections of this hemorrhagic soft tissue were analyzed and LCH was reported. Near-total excision of the soft tissue mass was performed without causing any injury to the anatomic structures in the mastoid cavity.

After confirmation of the persistent diagnosis, staging examinations were performed.

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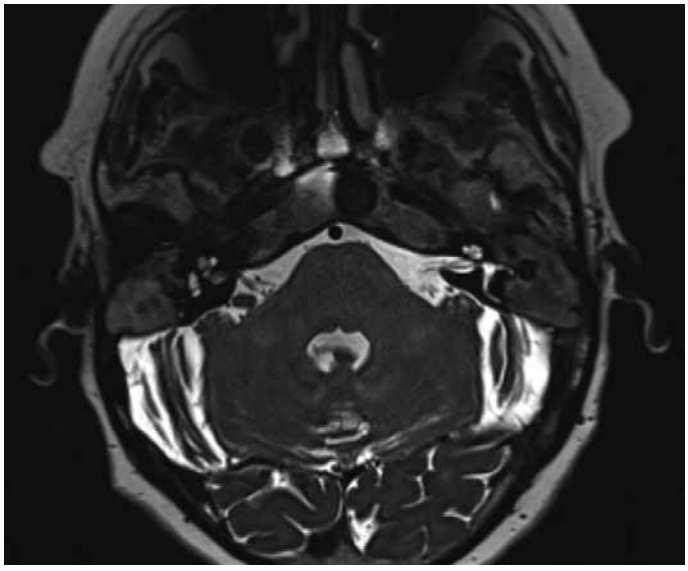

Figure 1. Axial CT section showing LCH infiltration of bilateral mastoid cavities.

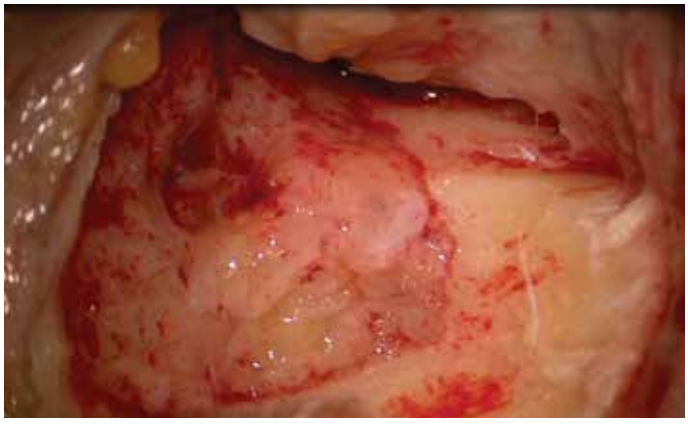

Figure 2. Intraoperative view of LCH infiltration in the mastoid cavity.

A positron emission tomography scan revealed a nodular 1 × 1-cm mass in the right lung and high avidity of flourodeoxyglucose in the second cervical vertebra, both reported as LCH infiltrations.

The case was diagnosed as "multifocal LCH disease" by the medical oncology department, and systemic treatment including prednisone and vinblastine was initiated.

The patient returned to her home country after the first session of chemotherapy. The communication with the patient and her local physicians continued for 1 month and was lost thereafter.

In addition, because of the lack of communication, no informed written consent was obtained from the patient. This case report does not contain any specific identity information of the patient.

DISCUSSION

LCH is an idiopathic disease characterized by abnormal proliferation of histiocytes. LCH usually affects children aged 1–4 years. The male:female ratio is 2:1. LCH can involve every organ, but the most commonly involved areas are bones, skin and lymph nodes. The most common presenting symptoms are skin rash, dyspnea, polydipsia and polyuria [2, 3].

LCH is divided into three groups: acute disseminated form, chronic multifocal form, and chronic focal form. Acute disseminated form is known as the Letterer-Siwe syndrome and chronic multifocal form is known as the Hand-Schuller Christian syndrome. The age of onset is important in the prognosis, whereas the prognosis is better in adults. Localized LCH has a good prognosis than the multiple-organ-involvement form [4].

Our patient was an adult with the disease in both ears. Clinical manifestation and audiological findings were first interpreted as infectious diseases of the bilateral middle ear. However, the history of the aggravation of the symptoms after discontinuing corticosteroids lead us to anticipate a different pathology than an ordinary otitis media. In literature, most reported cases with temporal involvement experienced ear symptoms for shorter periods than did our patient [5, 6]. The regular use of corticosteroid may be the reason for the patient to have mild problems of the ears and lengthen the period of the disease to become evident.

Granuloma formation in different organ systems causes different clinical manifestations; consequently, the misdiagnosis rates are very high. Definitive diagnosis can be achieved only with the detection of CD1a and S-100 proteins and Birbeck granules adhering to the cytoplasmic membrane.

Generally, we do not use frozen section examination during acute or chronic otitis media surgeries. However, in our patient, the history, bilateral involvement of mastoid bones, patient's hearing level changes after corticosteroid discontinuation, and different appearance of the soft tissue under operating microscope compared to inflammation or cholesteatoma led to perform a frozen section evaluation with the suspicion of LCH. Thereafter, with the diagnosis of the LCH, the soft tissue excision was performed with extreme caution without injuring the normal anatomical structures. In addition, triamcinolone-soaked gel foam pieces were applied to the cavity at the end of the operation.

In mastoid operations planned for LCH infiltration, a conservative surgery is recommended. Modugno et al. also underlined that the ossicular chain may remain uninvolved, which makes conservative surgical treatment possible [6].

After the definitive diagnosis, staging examinations revealed lung and rib involvements in our patient; however, there are also some cases with only temporal bone involvement reported in literature. Kleinjung et al. [7] reported an LCH case in a 2-year-old child with only bilateral mastoid infiltration. Bone involvements are seen mostly in skull bones; femur, pelvis, or vertebra involvements are rare. Temporal bone involvement leads to symptoms such as hearing loss, otalgia, retroauricular swelling, otorrhea, and ear discharge, which may mimic the symptoms of otitis media. In magnetic resonance imaging and CT, osteolytic lesions and soft tissue masses are usually seen.

Chemotherapy, radiotherapy, and surgery are treatment options for LCH. Localization of the tumor is an important factor for choosing the treatment regime. Chemotherapy is the treatment option for a multifocal disease, radiotherapy for lesions that cause organ dysfunction, and corticosteroids for local lesions [8].

In conclusion, LCH is a rare, idiopathic disease involving all systems. Therefore, otolaryngologists should consider granulomatous diseases and LCH in the differential diagnose of bilateral mastoid diseases.

CONCLUSION

LCH with only ear complaints in adults is a rare condition. In contrast, particularly in bilateral mastoid diseases without an onset period of the complaints, LCH should be kept in mind to avoid misdiagnoses and over treatment.

Informed Consent: Written informed consent could not be taken from the patient, because of the lack of communication. This case report does not contain any identity information about the patient.

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Conflict of Interest: The authors have no conflict of interest to declare.

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