A Case of External Auditory Canal Stenosis in Langerhans Cell Histiocytosis

Inkyung Sohn, Han-Bin Lee, Sung-Yeul Kim and Yun-Hoon Choung
Department of Otolaryngology, Ajou University School of Medicine, Suwon, Korea

Langerhans cell histiocytosis (LCH) refers to a group of disorders of the reticuloendothelial system characterized by a proliferation of histiocytes, which includes eosinophilic granuloma, Letterer-Siwe diseases, and Hand-Schüler Christian disease. The clinical presentation of LCH depends on the site of involvement. It can range from multifocal lesions to a solitary lesion. Tissuescharacteristically involved in LCH are bone, skin, lung, liver, spleen, bone marrow, lymph nodes and the hypothalamo-pituitary region, although the involvement of other organs such as the bowel can occur. We experienced a case of external auditory canal stenosis in LCH of multiple organ involvement in a 28-year-old male, and report it with a review of the relevant literature.

KEY WORDS: Langerhans cell histiocytosis · Temporal bone · External auditory canal · Stenosis.

Introduction

Histiocytosis is a group of proliferative disorders characterized by the infiltration and accumulation of histiocytes and other immune effector cells within various tissues.1) The term histiocyte includes monocytes and macrophages, dermal/interstitial dendritic cells, and Langerhans cells. Langerhans cell histiocytosis (LCH) is characterized by the aberrant proliferation of Langerhans cells normally found in the dermis.2) It frequently affects skull base areas, mainly temporal bones and frontal bones. In cases involving temporal bones, LCH is frequently located in the mastoid process and can infiltrate into the middle and external ears. It displays diverse symptoms depending on the site of involvement.3) But, specific symptoms and treatments have not been reported.

We experienced a case of external auditory canal stenosis in LCH with multiple organ involvement in a 28-year-old male, and report it with the review of the relevant literature.

Case Report

A 28-year-old male who had suffered from hearing impairment in the right side for 3 days was first observed in our outpatient clinic. The patient took medications for bilateral chronic otitis media about 10 years ago, and, for the past 2 years, had been undergoing chemotherapy and radiotherapy due to multifocal LCH involving the scalp, the axilla, and the lung. There was no symptom except right side hearing impairment. Both external auditory canals were nearly obstructed and eardrums were not observed. Bilateral otorrhea and granulomas in the external auditory canal were observed, but swelling of the posterior auricular area was not observed. Initial hearing test showed normal hearing thresholds in both ears (Fig. 1A). A computed tomography scan revealed obstructed bilateral external auditory canals with soft tissue density lesions, but the mastoid and the middle ear were clear (Fig. 2). The diagnosis of LCH was confirmed by skin biopsy in the outpatient clinic. The pathologic findings showed proliferative Langerhans cells in the dermis and positive staining with S-100 and CD1a in immunohistochemistry (Fig. 3A, B and C). T-tubes were inserted in both external auditory canals (Fig. 3D) and radiotherapy was commenced. However, both external auditory canals were totally obstructed due to progression of the disease 6 months after starting the treatment. Two years after the first visit, bilateral conductive hearing loss was discovered (air-bone gaps, 45 dBHL)(Fig. 1B). Therefore we planned surgical treatment to maintain patency of the external auditory canal. First, a right
The left side tumor excision was done; the left side was performed 4 months later. A circumferential incision was made on the site about 5 mm medial from the external auditory canal opening, and granulation tissue and skin adhering to the tumor were completely removed. A part of the bony external auditory canal was exposed, but drills were not used. The exposed bone was covered with gelfoam and packed with ear Merocels. Merocels were replaced with Nelaton catheters 1 week after the surgery to prevent growth of granulation tissue (Fig. 3E). Three months after the surgery, obstruction re-occurred due to recurrence of the disease, and Nelaton catheters were frequently changed because of discharge and granulation tissue. Finally, 3 years and 6 months after the surgery, both external auditory canals were totally obstructed again (Fig. 3F, G). The last hearing test showed conductive hearing loss (air-bone gaps, 35 dBHL), but the patient is not undergoing any treatment for obstruction of the external auditory canal and hearing impairment at present. Treatment with Talidomide (50 mg qd) was recently tried in the Department of Oncology because of a new exacerbation of LHC with skin and oral involvement.

Discussion

LCH is an abnormal proliferative disease of histiocytes including eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease. In 1953, Lichtenstein described them together as ‘Histiocytosis X-related manifestations of a single nosologic entity’. The incidence of LCH is around 5.4 per million and it is predominantly found in males. Its peak incidence period is noted in children 1–4-years-of-age, but it can affect patients of any age. Its cause has not been defined yet. It can range from multifocal lesions to a solitary lesion. In 60% of cases, it affects the skull base as a solitary lesion. Cases involving temporal bone account for 19–25% of cases, and about one-third display bilateral involvement. In cases involving temporal bone, granulomas cause obstruction of the bony external auditory canal, intractable otorrhea and pain. It is often misdiagnosed because the clinical presentation may mimic more common conditions like mastoiditis, chronic otitis media, or recurrent otitis externa. Hearing loss can range from mild conductive hearing loss to severe sensorineural hearing loss according to the extent of the disease.
Although conductive hearing loss is the most common problem, permanent sensorineural hearing loss may result from damage to the inner ear and bony labyrinth. LCH has an unpredictable natural history varying from a rapid progressing and fatal disease to spontaneous resolution. Poor prognostic factors are age at presentation (younger than 2-years-of-age), multisystem involvement and vital organ dysfunction.

Diagnosis of LCH is confirmed by the characteristic morphology and the presence of CD1a or CD207 (langerin) positive histiocytic cells. Nowadays, electron microscopic confirmation of the presence of Birbeck granules-immature Langerhans cell with characteristic pentagonal organelles is rarely used.

In this case, the patient had a history of multifocal histiocytosis and revealed bilateral otorrhea and stenosis of the external auditory canal. A computed tomography scan showed a soft tissue density lesion in both external auditory canals without bony destruction. Therefore, LCH was suspected, and was confirmed by skin biopsy. Comparatively young age of presentation and multifocal lesions predict poor prognosis.

There are several treatment modalities for temporal bone LCH, including surgery, radiotherapy, chemotherapy and injection of steroids. These treatments can be used alone or in combination depending upon the extent and severity of the disease. The preferred treatment is surgical removal when possible. Chemotherapy is helpful in a multifocal disease or when lesions are inaccessible to local treatment. Solitary radiotherapy is no longer recommended, but it is used as an additional treatment for residual lesions after surgical excision.

The major symptom of this case was hearing impairment due to stenosis of the external auditory canal. Surgical excision and insertion of T-tubes and Nelaton catheters were tried, but the effect was transient. At the present time, both external auditory canals are completely obstructed, and there is no interval change in conductive hearing loss. Although the preferred treatment for a localized lesion is surgical excision, it is a limited treatment unless preventive measures of obstruction are not developed. New techniques or methods are needed to prevent restenosis of the external auditory canal due to postoperative radiotherapy or tumor expansion.

REFERENCES