

LANGERHANS CELL HISTIOCYTOSIS WITH BILATERAL TEMPORAL BONE INVOLVEMENT IN AN ADULT

Otolology

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Özet

ERİŞKİN BİR HASTADA BİLATERAL TEMPORAL KEMİK TUTULUMLU LANGERHANS HÜCRELİ HİSTİYOSİTOZ

Langerhans hücreli histiyositoz, histiyositlerin anormal proliferasyonuna bağlı olarak ortaya çıkan, lokal ve sistemik tutulum gösterebilen miyeloid neoplazidir. Çocukluk çağının nadir görülen bir hastalığı olmakla birlikte erişkinlerde de görülebilmektedir. Hastalığın erkek-kadın oranı 2:1'dir. Ana tutulum yerleri deri, kemik ve yumuşak doku olmakla birlikte multifokal sistemik tutulum da olabilir. Temporal kemik tutulumu hastalığın belirtilerinden biridir. Temporal kemik tutulumunun en yaygın iki semptomu otore ve sübjektif işitme kaybıdır. Otalji ve postauriküler şişlik/hassasiyet eşlik edebilir. Bilateral temporal kemik tutulumu nadirdir, ancak literatürde daha önce bildirilen vaka raporları vardır. Bildirilen vakaların çoğu pediatrik yaş grubundadır; ilk şikayetler genellikle kulak akıntısı ve işitme kaybıdır. Bu yazıda sol kulakta kulak ağrısı ve postaurikuler hassasiyet şikayeti ile başvuran ve muayenesinde bilateral temporal kemik tutulumu saptanan Langerhans hücreli histiyositoz olgusunu sunuyoruz.

Anahtar kelimeler: Temporal kemik, Langerhans hücreli histiyosiyoz, Otalji

Abstract

LANGERHANS CELL HISTIOCYTOSIS WITH BILATERAL TEMPORAL BONE INVOLVEMENT IN AN ADULT

Langerhans cell histiocytosis is myeloid neoplasia that occurs due to abnormal proliferation of histiocytes and can have local and systemic involvement. It is a rare disorder of childhood but also can occur in adults. The male-female ratio of the disease is 2:1. Although the main sites of involvement are skin, bone, and soft tissue, multifocal systemic involvement can occur. Temporal bone involvement is one of the manifestations of the disease. The two most common symptoms of temporal bone involvement are otorrhea and subjective hearing loss. Otalgia and postauricular swelling/tenderness may accompany. Bilateral temporal bone involvement is rare, but there are previously reported case reports in the literature. Most reported cases are in the pediatric age group; the first complaints are usually otorrhea and hearing loss. We present a case of Langerhans cell histiocytosis, who applied to us with the complaint of otalgia and postauricular tenderness in the left ear, and who had bilateral temporal bone involvement on examination.

Keywords: Temporal bone, Langerhans cell histiocytosis, Otalgia

Introduction

Langerhans cell histiocytosis is an idiopathic disease involving the skin, bone, lung, liver, lymph nodes, mucocutaneous tissues, and endocrine organs locally or widely, with the cloning of abnormal histiocytes. Although the disease is usually seen in children, it can also occur in adulthood. Although it primarily affects children aged 1-4, its incidence in children has been determined as 5-6 per million in the United States. Langerhans cell histiocytosis has historically been divided into 3 classes. First is eosinophilic granuloma, a variant with single or multiple bone involvement without visceral involvement. The tumor may resolve spontaneously. It is considered benign. The second is Letterer Siwe's disease, a variant that progresses to multiple granulomas. It can invade the

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liver, spleen, lymph nodes, and central nervous system and have a poor course.

Hand Christian Schuller disease is the last variant. It presents with the triad of multiple bone lesions, exophthalmos, and diabetes insipidus. Schüller firstly described otologic manifestations of Langerhans cell histiocytosis in 1915 [1]. Also, Letterer described a case with a fever, purulent otitis media, lymphadenopathy, and hepatosplenomegaly [1].

Among the main symptoms of otological involvement are ear discharge resistance to medical treatment, hearing loss, swelling, and pain in the mastoid region. The posterior wall of the external auditory canal may erode [2]. The symptoms can be confused with conditions such as cholesteatoma and acute suppurative otitis in different periods of the disease.

We present a case of Langerhans cell histiocytosis with bilateral temporal bone involvement, who came to us with the complaint of pain in the left ear.

Case Report

A 50-year-old male patient with no known comorbidity and no previous otolaryngological operation was admitted to our clinic complaining of pain in the left ear. The patient indicated that the pain had been going on for a long time and was increasing gradually. There were no complaints of hearing loss or ear discharge. The patient did not have facial paralysis before. In the otoscopic examination, bilateral tympanic membranes were intact, and the external auditory canal was normal. Tenderness was detected on palpation of the left mastoid process. In further evaluation, audiometry and temporal CT examinations were performed. The patient had a symmetric bilateral mild sensorineural loss in pure tone audiometry. (The patient could not undergo a speech discrimination test (SDT) due to their inability to speak the same language as the audiologist and lack of proficiency in English.). Local destructive lesions in bilateral mastoid cells and middle ear were observed in temporal CT (Figure 1). An incisional biopsy was planned. The lesion area was reached with the left postauricular incision. After the skin and subcutaneous tissues were passed, it was observed that the bone tissue expected to be seen was eroded, and a granulomatous lesion protruding beyond it was detected. Biopsy material was obtained, and the result of the histopathological examination was compatible with Langerhans Cell Histiocytosis. A position emission tomography-computer tomography (PET-CT) scan was performed with the oncology opinion. There were lytic sclerotic lesions that involved both temporal bones, with higher FDG uptake in the left temporal bone (SUVMAX: 15.28) compared to the right (SUVMAX: 10.53). (Figure 1).

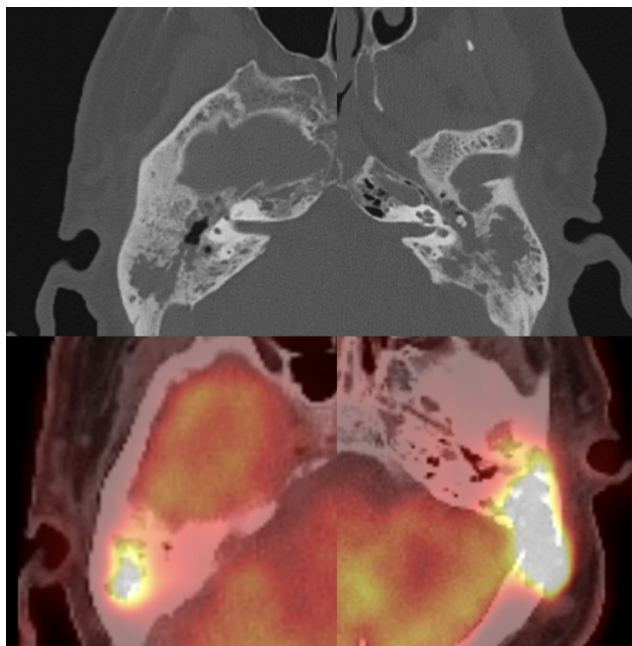


Figure 1

CT and PET-CT image of bilateral temporal bone before treatment

In addition, there was a slightly increased FDG uptake (SUVmax: 3.79) in the transverse process of the T9 vertebral body anterior corpus, which was considered suspicious in terms of involvement of the primary disease. No pathological FDG uptake was detected in the remaining areas of the body. The oncology council decided to administer low-dose radiotherapy to the left temporal bone, and follow-up was recommended for the lesion in the right temporal bone. As the tumoral structure in the right temporal region demonstrated lytic sclerotic features on imaging and the diagnosis of LGH was established in the opposite temporal bone, no biopsy was considered for the right side. Follow-up was recommended as there were no clinical complaints or symptoms. During follow-up, the patient was examined monthly and underwent audiometric testing; temporal CT scans were performed every six months. The patient did not report any complaints in their right ear, no changes in their audiogram, and no increase in tumor size in the CT scan. After 30 courses of radiotherapy for the lesion in the left temporal bone, control temporal CT scans revealed that the lesion had regressed. (Figure 2) The patient's left otalgia symptom had disappeared entirely after treatment.

There was no change observed in the monthly evaluations of the patient's right ear and in the 6-month CT evaluation. The patient presented again complaint in the right ear during the 9-month follow-up and with otalgia was evaluated in the oncology council. As a result of the evaluation, due to the patient's ongoing complaint, 30 sessions of low-dose radiotherapy were recommended. After treatment, the patient's otalgia complaint decreased but did not completely disappear. The post-treatment appearance of the lesion in the right temporal bone can be seen in Figure 2.

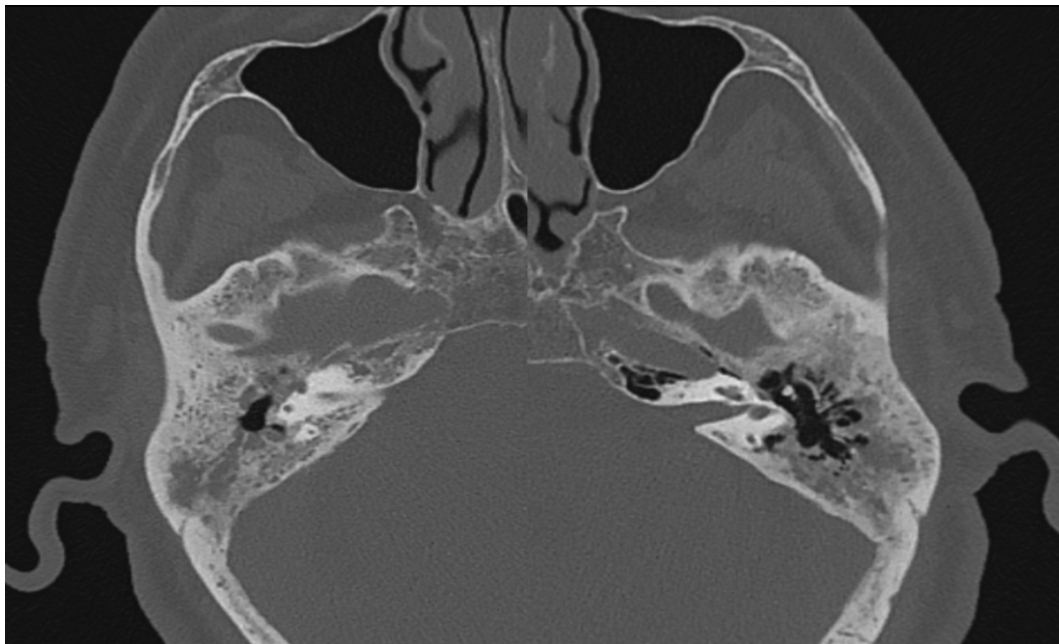


Figure 2

CT image of bilateral temporal bone after treatment

There was no significant difference in the patient's post-treatment audiometry compared to pre-treatment audiometry. (Figure 4)

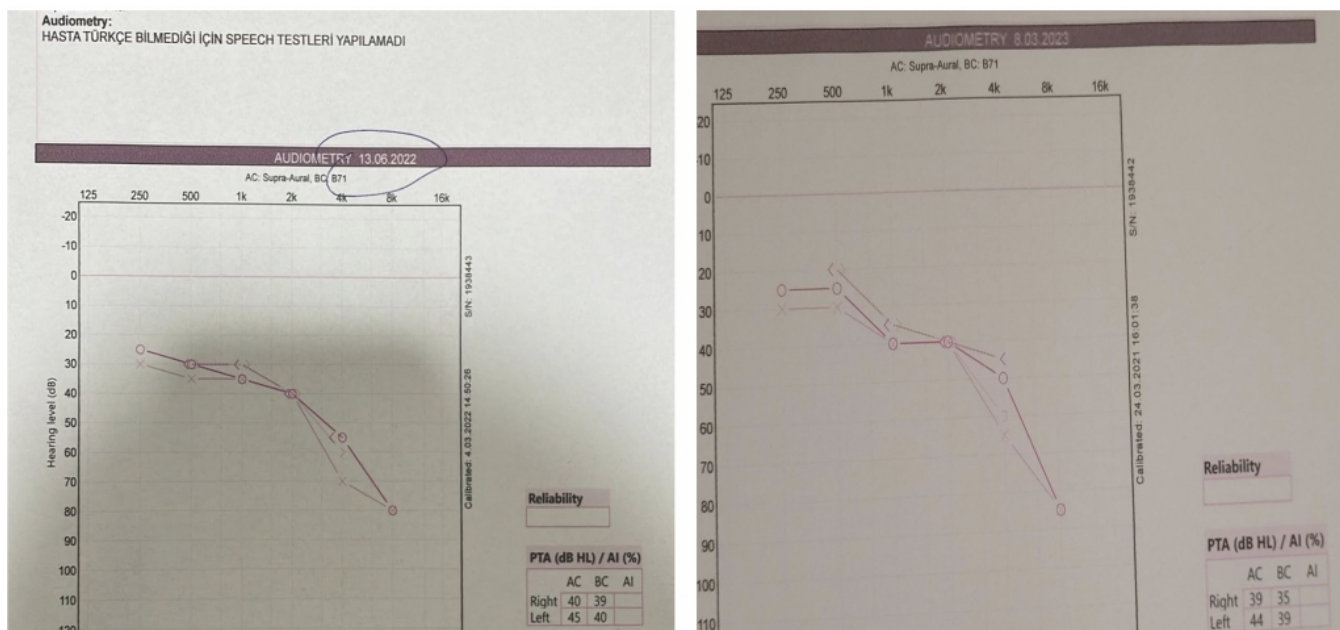


Figure 3

Audiograms of the patient before and after treatment

Discussion

Langerhans cell histiocytosis is a disease caused by abnormal proliferation of histiocytes, progresses with granuloma formation, and can cause invasion and local destruction. At the same time, it may have a multifocal course and may lead to conditions such as systemic involvement and organ failure [3]. It is seen twice as often in

men than in women, and its incidence in adults is rarer than in children [4].

LCH frequently affects the external and middle ear. Still, accurate diagnosis can be challenging due to its clinical presentation. It can be mistaken for more prevalent conditions such as mastoiditis, chronic otitis media, recurrent otitis externa or otitis media, stenosis, or polyps in the external auditory canal. Other less common symptoms include peri-auricular or middle ear masses, conductive hearing loss, otalgia, otorrhagia [9].

Some tumoral formations can also cause destruction in the middle ear. Cholesteatoma is the most commonly seen among them and is considered in the differential diagnosis before LGH. It can be differentiated by diffusion restriction on MRI examination. Glomus tumors (jugular and tympanic) can sometimes be aggressive and cause destruction the middle ear, antrum, and mastoid when they reach advanced stages. The tumor's expansion of the jugular foramen on the CT scan can be a clue. CT angiography is important in making the differential diagnosis. Rarely, primary or secondary neoplasms can cause destructive lesions in the middle ear. Fibrous dysplasia is a benign but locally destructive bone tumor seen in the mastoid bone and middle ear. Malignant tumors and metastases are much less commonly observed.

In a case series of 20 patients presented by Modest et al., the most common symptoms were otorrhea, subjective hearing loss, otalgia, postauricular swelling, and soft tissue swelling, respectively. In the same study, the most common finding was osteolytic lesions in the temporal bone [5]. Treatment-resistant chronic otitis with the symptoms above, unilateral disease becoming bilateral over time, or bilateral onset of the illness should bring Langerhans Cell Histiocytosis to mind in the diagnosis. A definitive diagnosis is made by detecting CD1a and S100 proteins and Birbeck granules attached to the cytoplasmic membrane in the biopsy material [4].

Chemotherapy, radiotherapy, surgery, or their combinations can be used in the treatment. In cases with multifocal involvement, the first choice is chemotherapy containing vinblastine and prednisone [6]. Multiagent chemotherapy can be considered in relapses [5].

Radiotherapy is usually given as external beam radiation in the dose range of 1000 - 1,800 cGy in locally destructive progressive diseases [4]. In local and non-large lesions, the treatment is the surgical removal of the mass, and if the surgical margin is positive, it is supported with low-dose radiotherapy [7].

The case we presented was a male patient with no complaints other than unilateral ear pain and postauricular tenderness. Although he did not have any complaints in the other ear, a lesion filling the mastoid cavity was incidentally observed in the other ear in the CT scan. Although cases of Langerhans cell histiocytosis with temporal bone involvement are common, it is rare that the patient has bilateral participation and is an adult. It should be considered in the differential diagnosis of bilateral diseases. Systemic involvement should be determined and followed up because it has been reported that systemic symptoms may begin a while after the illness that starts with temporal bone involvement [5]. In the presented case, systemic chemotherapy was not considered since the mass was limited to the temporal bone, had no involvement in another region, and had no systemic symptoms. Surgery was not considered because of extensive involvement in the temporal bone. The risk of total hearing loss and facial paralysis is high in surgery to be performed on a patient with a mass with this prevalence. At the same time, CSF leakage due to possible dural defects may be encountered. For this reason, it was decided that the patient should receive radiotherapy with the decision of the head and neck oncology council.

Close follow-up of the patient after treatment is essential because the risk of relapse of the disease is high. The lesion should be followed up with 1 MRI or PET every 6 months [8].

Informed Consent: From the patient

References

1. McCaffrey, T. V. & McDonald, T. J. Histiocytosis X of the ear and temporal bone: review of 22 cases. *The Laryngoscope* 89, 1735–1742 (1979).
2. Marioni, G., De Filippis, C., Stramare, R., Carli, M. & Staffieri, A. Langerhans' cell histiocytosis: temporal bone involvement. *J. Laryngol. Otol.* 115, 839–841 (2001).
3. Alhaidri, N. E., Alim, B., Alrushaid, N. R., Fatani, H. & Binnasser, A. S. Temporal Bone Langerhans Cell Histiocytosis: An Uncommon Bilateral Presentation. *Cureus* (2021) doi:10.7759/cureus.12732.
4. Clinic of Ear Nose and Throat, Vehbi Koc Foundation American Hospital, Istanbul, Turkey, Bahar, S., Dal, T., & Clinic of Ear Nose and Throat, Vehbi Koc Foundation American Hospital, Istanbul, Turkey. Langerhans Cell Histiocytosis of Bilateral Mastoid Cavity. *J. Int. Adv. Otol.* 14, 341–343 (2018).
5. Modest, M. C., Garcia, J. J., Arndt, C. S. & Carlson, M. L. Langerhans cell histiocytosis of the temporal bone: A review of 29 cases at a single center: LCH of the Temporal Bone. *The Laryngoscope* 126, 1899–1904 (2016).
6. Allen, C. E., Merad, M. & McClain, K. L. Langerhans-Cell Histiocytosis. *N. Engl. J. Med.* 379, 856–868 (2018).
7. Chevallier, K. M., Wiggins, R. H., Quinn, N. A. & Gurgel, R. K. Differentiating Pediatric Rhabdomyosarcoma and Langerhans Cell Histiocytosis of the Temporal Bone by Imaging Appearance. *Am. J. Neuroradiol.* 37, 1185–1189 (2016).
8. Erdevicki L, Belic B, Lukić A, Marković D. Langerhans cell histiocytosis of the temporal bone. *Ser J Exp Clin Res.* 2009;10(4):147-149.
9. Saliba I, Sidani K, El Fata F, Arcand P, Quintal MC, Abela A. Langerhans' cell histiocytosis of the temporal bone in children. *Int J Pediatr Otorhinolaryngol.* 2008 Jun;72(6):775-86. doi: 10.1016/j.ijporl.2008.02.001. Epub 2008 Mar 19. PMID: 18355926.

Information About Previous Presentations

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