INTRODUCTION

Langerhans cell histiocytosis (LCH) is a very rare condition with an incidence of 4–5 cases per million per year in children in Europe.1,2 Currently, the etiology of the disease is not completely known; there is some evidence, that it is mainly caused by unchecked proliferation of Langerhans cells, which occur predominantly in children and teenagers. The course of this disease is not uniform. In some cases, single lesions with spontaneous regression occur, resembling an abnormal reactive or inflammatory process rather than a neoplasia. On the contrary, however, multiple lesions with infiltrative growth pattern like malignant tumors can appear with a mortality rate of up to 50%.3

Clinical symptoms are variable. Some children may initially present with a rash. However, the disease involves the skeleton in up to 80% of patients with infiltration of skull bones, skull base, hip/pelvis, femur, and ribs. Other organ manifestations of LCH are involvement of lymph nodes in up to 30% of cases, pituitary gland (in up to 25%), lungs (in up to 20%), liver, spleen, bone marrow, gastrointestinal tract, and central nervous system.3,4

A clinical classification system differentiates between a single organ disease, in which only one organ system is affected, and a multiorgan or systemic disease, in which two or more organs/organ systems are involved.5 Additionally, this classification system describes risk lesions, which are defined as lesions with a higher risk of fatal courses or a higher risk of clinical complications.4 A LCH lesion in the temporal bone is classified as a risk lesion, because severe complications can occur, that is, hearing loss, balance disorders, facial nerve palsy, or intracranial extension. Single system-LCH involving exclusively the temporal bone is rare. Roughly half of children with temporal bone LCH show bitemporal involvement.6–9

Treatment strategies vary and are depending on the clinical course of the LCH and comprise: watchful waiting, medical treatments (topic vs. systemic application—corticosteroids, immunosuppressants, chemotherapeutics), irradiation, or surgery.4

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CASE REPORT

Case report: Langerhans cell histiocytosis of the temporal bone in children: Challenging diagnosis of a rare disease with some pitfalls

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Abstract
A 4-year-old girl was admitted to hospital with disturbance of balance. After being questioned, parents remembered an otitis with effusion 3 months earlier. CT-scans revealed destruction of both temporal bones. Initial biopsy showed granulomatous, necrotic inflammation, which led to comprehensive differential diagnoses. A second tissue sample confirmed Langerhans cell histiocytosis.

KEYWORDS
facial paralysis, hearing loss, Langerhans cell histiocytosis, neuroblastoma, orofacial granulomatosis, temporal bone, vestibular diseases
2 | CASE HISTORY AND EXAMINATION

A 4-year-old female child with Turkish ancestry was admitted to the emergency service of a Children’s Hospital with progressive walking disturbances and recurrent falling. The parents reported that their daughter had been able to walk and run steadily, but 5 days before admittance she staggered and fell down repeatedly, or at least she grabbed parents’ hands to avoid falling. During her first fall, she hit her head on a hard surface, which resulted in mild head injury and an open wound on the nasal bridge that had to be stitched. There were no known prior diseases, surgeries, or allergies.

The medical examination revealed a seemingly healthy child, GCS 15/15, no infection of the wound on the nasal bridge, no more obvious staggering; both ear canals were blocked by effusion/cerumen. All other clinical findings were in accordance with an “age-appropriate” child. Routine blood samples taken during admittance were without any pathological findings. According to the history of recurrent falling and balance disorders, the kid was admitted to the hospital for further differential diagnostic examinations and treatment.

3 | DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT

During ENT examination with the pediatric otorhinolaryngologist, the parents reported “problems with the ears” for 3 months. Initially, an acute otitis media with effusion and ear drum perforation had been diagnosed. By systemic and topical application of antibiotics the effusion decreased but was never receding completely. The parents further reported a slight language development delay, while being raised bilingually. Their daughter had received speech therapy.

Microscopic examination of the left ear showed no effusion, and the ear drum was intact without signs of inflammation. On the right side, the external ear canal was blocked by polyps and secretion. The tympanic membrane could not be seen—even after intensive cleaning of the external ear canal—due to polyp formation. A nystagmus was not evident by examination with Frenzel’s nystagmus glasses. The clinical facial nerve function was found to be intact on both sides. A computed tomography (CT-scans) of the head and temporal bones was recommended. It revealed evidence of osteolysis of the temporal bone on both sides, including both mastoids, both superior and lateral semicircular canals, and the external auditory canal on the right side. Additionally, there was a third osteolytic lesion in the dorsal parts of the left-sided parietal bone adjacent to the occipital bone (Figure 1). CT-scans further showed hypertrophic adenoids and slightly swollen mucosa of the paranasal sinuses. CT-morphologically the bone lesions were regarded as highly suspicious for Langerhans cell histiocytosis or metastatic neuroblastoma.

Audiologically, a profound hearing loss close to deafness on the right side with almost normal hearing on the left ear was found (Figure 2). Clinical and video-assisted head impulse tests could not be performed reliably due to the young age of the child and non-cooperative behavior.

In the next step, a biopsy from the right external auditory canal was taken in general anesthesia. Here, a very soft, pinkish, in parts grayish, polypoid tissue with some inclusions of cartilage like fragments was found. Tissue samples were taken for both histopathological and microbiological examinations. Another sample was taken from the underlaying bone of the eroded external ear canal wall.

Histopathologically, granulomatous inflammation and necrosis were described without any pathologic Langerhans cells, eosinophilia (which may occur in LCH), or any malignant cells.

According to these findings, further differential diagnoses of subacute to chronic osteolytic mastoiditis/osteolytic temporal bone lesions were considered (Table 1). Further blood and urine samples were taken, a chest radiograph and skin test for tuberculosis were performed. The girl's immune competence was checked roughly to evaluate whether it would be necessary to consider opportunistic infections or grave courses of usually mild diseases. This included full blood count, vaccine titers, serum electrophoreses, serum immunoglobulin status, and HIV status (Table 2).

However, none of these examinations led to a final diagnosis. After re-evaluation of the histopathologic findings and discussion with the pathologists, a second biopsy of the affected bone was taken along with a biopsy of the hypertrophic adenoids. Intraoperatively, the bone of the skull (including the peristium) was found to be completely osteolytic, and the resulting osteolytic bone defect was filled with soft, necrotic tissue (Figure 3). Initially suspected Langerhans cell histiocytosis could now be confirmed morphologically and immunohistochemically by positive staining for CD1a and CD 207 (Langerin) (Figure 4).

The patient was referred to the Dept. of Pediatric Oncology, where staging was completed by whole-body MRI. Fortunately, no further lesions were detected, and a chemotherapy with vinblastine and corticosteroids was started immediately. Treatment was continued for 6 months, after which almost full remission was achieved. Magnetic resonance imaging (MRI) scans showed only minimal residual enhancement in both temporal bones.
However, to date re-mineralization of the bone was not evident. Audiologic controls still showed a hearing loss, which will be monitored closely in the future.

4 | DISCUSSION

Langerhans cell histiocytosis (LCH) is a rare disease probably caused by unregulated proliferation of dendritic Langerhans cells. It affects mostly young children with an age maximum (91% of cases) of 1–4 years9 (mean age of 3 years).7,19 The course and prognosis of this condition is not uniform and the symptoms are often unspecific.3 In this case report, we present the case of a girl with a craniofacial LCH, in whom both temporal bones and the dorsal parietal bone were affected. Due to the variety of symptoms of craniofacial LCH, diagnosis is challenging and misdiagnosis and mistreatment are common.9,20 Furthermore, clinical symptoms of temporal bone LCH are very unspecific with scalp/postauricular lesions, otalgia, persistent ear infections/granulation of the external auditory canal, or hearing loss (mostly conductive hearing loss).19,21–23 In very rare cases, disturbance of balance and uncertain gait are the only noteworthy symptoms for the parents indicating labyrinthine involvement and bringing them to the emergency room.

However, temporal bone or ear affection are regarded to be not very common in LCH. Some authors report that only 30% of pediatric patients present with involvement of temporal bone or ears.24,25 Nevertheless, the rates vary from 4% through 61%, and among children with craniofacial LCH, the temporal bone is the most common localization (82%).8,28 Up to half of the patients show a binaural involvement (30%–55%), like the patient in this case report.7–9 In CT-scans, the osteolytic lesions of the temporal bone display a typical “punched out” aspect 5 (Figure 1) and are usually located in the mastoid, squamous temporal bone, external auditory canal, middle ear, or petrous apex.19

Langerhans cell histiocytosis diagnosis is based on histological and immunohistological examinations4 and may sometimes be challenging. The case reported here shows how many potential differential diagnoses have to be considered (Table 1), particularly, if an initial tissue sample shows neither typical histological characteristics of LCH nor CD1a or CD207 (Langerin) staining in immunohistochemistry, which are regarded to be pathognomonic for LCH.4

Our patient suffered from single system (SS) multifocal bone involvement, which usually remains limited to the skeleton. The treatment options vary from watchful waiting to systemic chemotherapy, depending on the duration and extension of disease, symptoms, and risk of permanent sequelae.4 Our patient received the most commonly applied systemic chemotherapy for single system-LCH (multifocal bone affection) with vinblastine and corticosteroids. She is now in remission at her 6-month follow-up. When contemplating the therapeutic options, the
decision for this therapy was made due to the vast labyrin-thine destruction, risk of permanent balance disturbances and risk of deafness.

In general, craniofacial single- or multifocal bone LCH has a favorable prognosis. However, mortality rates of 4.5% through 18% have been reported. Recurrences occur frequently, particularly in cases with multifocal bone involvement (16.7%–57%). Additionally, there is a high rate of long-term morbidity and sequelae when the temporal bone is involved by the disease:

- Hearing is often lost when the otic capsule is destroyed. Otic capsule involvement is reported in 20%–29% of cases, resulting in sensineuronal hearing loss in 75% of these cases. After treatment, there may be bone re-mineralization and re-constitution of the labyrinth, but the sensineuronal hearing loss usually persists except for extremely rare cases. However, there are reports of successful cochlear implantation to restore hearing ability.

- Even if there is no initial hearing loss, several authors report progressive bilateral hearing loss following treatment. Nandurie et al. observed hearing loss in 38% of their patients 5 years post-therapeutically. The following reasons for this progressive hearing loss should be discussed: ototoxic chemotherapy, acquired cytomegalovirus infection in the immunocompromised situation, or an initially undiscovered congenital inner ear anomaly. Finally, a bias during audiologic examination should also be considered, particularly when subjective hearing tests are performed: in young patients (i.e., at initial diagnosis) hearing assessment is not as easy as in older patients. It is time-consuming and requires a lot of empathy and experience by the examiner. Therefore, younger patients may present with apparently better hearing results. In doubtful cases, objective hearing tests should be performed, even if a sedation is required.

- Chronic otitis externa and chronic otitis media with or without effusion occur often even after successful treatment and remission. However, children with LCH do not develop middle-ear cholesteatoma more often than their healthy counterparts.

- The temporal bone location of LCH may also affect the facial nerve, leading to persistent facial nerve palsy.

- Intracranial extension of temporal bone LCH occurs quite often (up to 35%) and may lead to persistent damage of parts of the central nervous system with severe neurological sequelae.

To highlight the learning points of this case:

- Otitis with effusion that does not improve despite adequate therapy should always be a warning sign. It should lead to further radiologic evaluation, particularly if associated with hearing loss, balance disorders, or facial nerve palsy—even if the symptoms occur time-delayed.

- When working with children who cannot yet articulate their symptoms accurately, the medical staff has to look for indirect signs of temporal bone involvement such as “falling” for vestibular disturbances or “delay of language development” for hearing loss. The parents cannot connect different symptoms into one clinical picture, so medical staff has to be diligent in questioning the parents and meticulous in examining the sometimes non-cooperative child.

- The differential diagnoses of temporal bone lesions are extensive; histology should always be acquired to rule out malignancy. If there is a discrepancy between clinically malignant signs and benign histology, a second biopsy is required, and an interdisciplinary discussion with the pathologist might be advantageous.
| Disease                                      | Characteristics                                                                 | Typical diagnostic findings                                                                                                                                 |
|---------------------------------------------|---------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Tuberculous mastoiditis                    | • Primary TB: usually lung, specific intrapulmonary lesion (Ghon focus), possibly with lymph node affection (Ghon complex) | • Blood count: leukocytosis, blood sedimentation ↑, serum: CRP ↑  
• Chest X-ray/CT-scan: very variable, discrete infiltrates, through Ghon focus/complex, through mediastinal enlargement (with calcification), to characteristic (apical) cavities (even bi-pulmonary)  
• Indirect pathogen detection: tuberculin skin test or blood interferon-γ release assay  
• Direct pathogen detection: from body fluids or tissue biopsies |
| Syphilitic osteomyelitis                    | • Stage I: chancre, lymphadenitis  
• Stage II: exanthema, condylomata lata  
• St. III: cardiovascular syphilis, gummata (granulomas of skin, mucous membranes, and bone) | • Blood count: leukocytosis  
• Blood sedimentation ↑  
• Serum: CRP ↑, TPPA/TPHA pos. |
| Granulomatosis with polyangitis (Wegener’s granulomatosis) | • Immune-vasculitis with granulomatous inflammation  
• Early symptoms: chronic rhinitis/sinusitis, chronic otitis/mastoiditis  
• Affected organs later: lung > kidney, eyes > skin > heart, nervous system | • Blood count: anemia, leukocytosis, thrombocytosis  
• Blood sedimentation ↑, serum: CRP ↑, cANCA ↑ > RF ↑ > ANA ↑  
• Urinanalysis: hematuria, proteinuria  
• Chest X-ray: single or bi-pulmonary round foci  
• Chest CT-scan: single or bi-pulmonary round foci, ground-glass opacification |
| Sarcoidosis                                 | • Disease with formation of granulomata, etiology unknown  
• Main focus: lung, but any other organ can be affected | • Blood count: leukocytosis, blood sedimentation ↑  
• Serum: Ca2+ ↑, CRP ↑, ACE↑, sIL-2R ↑, IgG ↑  
• Chest X-ray: bilateral lymphadenopathy, reticulonodular infiltrates, cystic and bullous changes |
| Cholesteatoma                                | • Accumulation of exfoliated keratin produced from stratified squamous epithelium  
• Genuine vs. secondary | • Ear microscopy: discharge, polyps, epitympanal crust, flaky-white debris  
• Valsalva maneuver often negative, positive fistula sign  
• Audiology testing: conductive hearing loss, reduced vestibular function |
| Malignant otitis externa                    | • External otitis that progresses into an osteomyelitis of the temporal bone  
• Weakened immune system is a prerequisite | • Ear microscopy: discharge, swollen external ear canal, exposed bone  
• Audiology: conductive or combined hearing loss, vestibular function ↓  
• Facial palsy, meningitis, sinus thrombosis, palsy of N. IX, X, XI  
• Blood count: leukocytosis, blood sedimentation ↑  
• Serum: CRP↑, Glc/HbA1c (diabetes) ↑, HIV pos. (immunosuppression) |
| Ewing sarcoma                               | • Predominant site: long bones of extremities and pelvis | • Blood sedimentation ↑, serum: CRP ↑, alkaline phosphatase ↑, NSE ↑  
• MRI (primary lesion), local lymph node sonography  
• Staging: chest CT-scan (20% pulmonary metastasis), skeletal scintigraphy |
| Osteosarcoma                                | • Predominant site: metaphyseal of long bones, esp. prox. tibia and distal femur | • Serum: CRP ↑, alkaline phosphatase ↑  
• MRI (primary lesion), local lymph node sonography  
• Staging: chest CT (10%–20% pulmonary metastasis), skeletal scintigraphy |

(Continues)
**Disease**

**Characteristics**

**Typical diagnostic findings**

**Chondrosarcoma**
- Predominant site: pelvis, prox. Femur
- Mostly adults

- Serum: CRP ↑, alkaline phosphatase ↑

**Non-Hodgkin lymphoma (NHL), e.g.**
- Plasmacytoma/multiple myeloma
- Diffuse large B-cell NHL
- Diffuse large T-cell NHL

- Blood count, blood sedimentation ↑, serum: CRP ↑, Ca²⁺ ↑, alkaline phosphatase ↑/↓, monoclonal gammapathy

**Metastatic neuroblastoma**
- Neuroblastomas arise from the sympathetic nervous system, esp. adrenal glands, retroperitoneum, and posterior mediastinum

- Urine: VMA and HVA in 24-h urine collection↑

**Giant cell tumor**
- Originates from undifferentiated mesenchymal cells of the bone marrow
- Predominant site: epiphysis of long bones

- Urine: parathyroid hormone (to rule out “brown tumor” caused by hyperparathyroidism)

**Benign/semi-benign lesions**
- Osteoblastoma, chondroblastoma, central giant cell lesion/granuloma, non-ossifying fibroma, fibrous dysplasia, aneurysmal bone cyst, vestibular schwannoma, meningioma, glioma, neurona, chordoma, epidermoid, jugulotympanic paraganglioma, ACI aneurysm

Note: This table gives an overview of considerations on differential diagnoses of subacute to chronic mastoiditis with osteolysis in tomographic imaging. Histopathologically, some of the described lesions present themselves primarily as granulomatous chronic inflammation, such as tuberculous mastoiditis, syphilitic ostemyelitis, granulomatosis with polyangiitis (Wegener’s granulomatosis), and sarcoidosis. In other lesions, necrosis is predominant (e.g., malignant otitis externa). And, the last group is predominated by its osteolytic aspect in tomography: Ewing sarcoma, osteosarcoma, chondrosarcoma, non-Hodgkin lymphoma (NHL), neuroblastoma, giant cell tumor.10–18

Abbreviations: ACE, acetylcholinesterase; ANA, antinuclear antibody; cANCA, cytoplasmic antineutrophil cytoplasmic antibodies; CRP, C-reactive protein; CT, computed tomography; Glc, glucose; HVA, homovanillic acid; IgG, immunoglobin G; LDH, lactate dehydrogenase; MIBG, metaiodobenzylguanidine; NSE, neuron specific enolase; pANCA, perinuclear antineutrophil cytoplasmic antibodies; RF, rheumatoid factor; sIL-2R, soluble interleukin-2 receptor; TB, tuberculosis; TPHA, Treponema pallidum hemagglutination assay; TPPA, Treponema pallidum particle agglutination assay; VMA, vanillylmandelic acid.
Audiologic examinations should always be performed in patients with mastoid/temporal bone affections to detect a significant hearing loss early and to consider adequate hearing rehabilitation for speech development. In doubtful cases with uncertain results of subjective hearing tests or when the children are too young for subjective hearing tests, objective assessment of hearing is mandatory.

**AUTHOR CONTRIBUTION**
Dr. med. Anja Pähler vor der Holte took care of the child on the ward and wrote the manuscript. Prof. Dr. Dr. Hans-Jürgen Welkoborsky provided intellectual inputs and supervised the work.

**ACKNOWLEDGEMENTS**
We would like to thank Dr. Juergen Weidemann (head of the department of pediatric radiology, children’s and youth hospital "Auf der Bult", Hanover, Germany) for...
supplying the CT images and Prof. Dr. Ludwig Wilkens (head of the Department of Pathology, Nordstadt Clinic, Hanover, Germany) for providing the histopathological images.

CONFLICTS OF INTEREST
The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT
The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

CONSENT
Written informed consent was obtained from both parents of the patient to publish this report in accordance with the journal’s patient consent policy.

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REFERENCES
1. Salotti JA, Nanduri V, Pearce MS, Parker L, Lynn R, Windebank KP. Incidence and clinical features of Langerhans cell histiocytosis in the UK and Ireland. Arch Dis Child. 2009;94:376-380.
2. Kürten T, Angerstein W, Ganzer U. Häufigkeit von Hörstörungen bei Langerhans-Zell-Histiozytose im Kindes- und Jugendalter. Dissertation. Medical Faculty of the Heinrich-Heine-University; 2005.
3. Tillotson C V., Anjum F., and B. C. Patel. 2021. Langerhans Cell Histiocytosis StatPearls [Internet], last update Feb 25, 2021.
4. Haupt R, Minkov M, Astigarraga I, et al. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer. 2013;60:175-184.
5. Leung AKC, Lam JM, Leong KF. Childhood Langerhans cell histiocytosis: a disease with many faces. World J Pediatr. 2019;15:536-545.
6. Nanduri VR, Pritchard J, Chong WK, Phelps PD, Sirimanna K, Bailey CM. Labyrinthine involvement in Langerhans’ cell histiocytosis. Int J Pediatr Otorhinolaryngol. 1998;46:109-115.
7. Saliba I, Sidani K. Prognostic indicators for sensorineural hearing loss in temporal bone histiocytosis. Int J Pediatr Otorhinolaryngol. 2009;73:1616-1620.
8. modest MC, Garcia JJ, Arndt CS, Carlson ML. Langerhans cell histiocytosis of the temporal bone: a review of 29 cases at a single center. Laryngoscope. 2016;126:1899-1904.
9. Yang S, Yao H. Analysis of clinical features of Craniofacial Langerhans cell histiocytosis in children. (In Chinese). Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2020;34:999-1001.
10. Amboss. 2021. https://www.amboss.com/. Accessed on January, 2021.
11. Radiopaedia. 2021. https://radiopaedia.org/. Accessed on January, 2021.
12. Alqunae M, Aldaihani A, AlHajery M. Non-Hodgkin’s Lymphoma of the Middle Ear Presenting as Mastoiditis. Case Rep Otolaryngol. 2018;2018:7639784.
13. Duerr HR. “Gutartige”Knochenentumoren - nicht immer ungefährlich. (In German). Ortopädie Rheuma. 2005;2:46-52.
14. Kaya I, Benzer M, Turhal G, Sercan G, Bilgen C, Kirazli T. Giant cell tumor of the temporal bone and skull base: a case report. J Int Adv Otol. 2018;14:151-154.
15. Isaacson B, Berryhill W, Arts HA. Giant-cell tumors of the temporal bone: management strategies. Skull Base. 2009;19:291-301.
16. Vaid S, Jadhav J, Chandorkar A, Vaid N. Bilateral Non-Hodgkin’s Lymphoma of the temporal bone: a rare and unusual presentation. Case Rep Otorhinolaryngol. 2016;2016:2641876.
17. Alvi SA, Flynn JP, Gener M, Cullen R. Burkitt Lymphoma of the temporal bone. Otol Neurotol. 2018;39:e410-e412.
18. Uy JAG, Zugo J, Acuin J. Primary Lymphoma of the temporal bone: the first locally reported case. Philipp J Otolaryngol Head Neck Surg. 2008;23:23-27.
19. Majumder A, Wick CC, Collins R, Booth TN, Isaacson B, Kutz JW. Pediatric Langerhans cell histiocytosis of the lateral skull base. Int J Pediatr Otorhinolaryngol. 2017;99:135-140.
20. Chen L, Wang WQ, Xu H, Chi FL. Langerhans cell histiocytosis of the temporal bone: 22 cases analysis. (In Chinese). Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2010;45:212-216.
21. Wang X, Liu W, Xie S, Peng A, Ren J. Langerhans cell histiocytosis of the temporal bone in children: 7 cases analysis. (In Chinese). Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2013;29:1898-1902.
22. Abdel-Aziz M, Rashed M, Khalifa B, Talaat A, Nassar A. Eosinophilic granuloma of the temporal bone in children. J Craniofac Surg. 2014;25:1076-1078.
23. Kleinjung T, Woenckhaus M, Bachthaler M, Wolff JF, Wulf SR. Langerhans’ cell histiocytosis with bilateral temporal bone involvement. Am J Otolaryngol. 2003;24:265-270.
24. Braier J, Chantada G, Rosso D, et al. Langerhans cell histiocytosis: retrospective evaluation of 123 patients at a single institution. Pediatr Hematol Oncol. 1999;16:377-385.
25. Cunningham MJ, Curtin HD, Jaffe R, Stool SE. Otologic manifestations of Langerhans’ cell histiocytosis. Arch Otolaryngol Head Neck Surg. 1989;115:807-813.
26. Nelson BL. Langerhans cell histiocytosis of the temporal bone. Head Neck Pathol. 2008;2:97-98.
27. Kürten T, Groeger M, Angerstein W. Frequency of hearing disorders in children with langerhans’ cell histiocytosis. (In German). Laryngorhinootologie. 2008;87:96-99.
28. Haupt R, Nanduri V, Calevo MG, et al. Permanent consequences in Langerhans cell histiocytosis patients: a pilot study from the Histioyte Society-Late Effects Study Group. Pediatr Blood Cancer. 2004;42:438-444.
29. Willis B, Ablin A, Weinberg V, Zoger S, Wara WM, Matthay KK. Disease course and late sequelae of Langerhans’ cell histiocytosis: 25-year experience at the University of California, San Francisco. J Clin Oncol. 1996;14:2073-2082.
30. Förster U, Klingebiel R, Schulte OU, Sarioglu N, Lehmann R. Langerhans cell histiocytosis: petrosal remodelling after chemotherapy--Case report and review of the literature. (In German). Laryngorhinootologie. 2003;82:166-170.
31. Kusumakumary P, James FV. Permanent disabilities in childhood survivors of Langerhans cell histiocytosis. Pediatr Hematol Oncol. 2000;17:375-381.
32. Losie JA, Yong M, Kozak FK, Chadha NK. Unique case of hearing recovery after otic capsule destruction and complete sensorineural hearing loss caused by Langerhans cell histiocytosis. *Otol Neurotol*. 2017;38:1129-1132.

33. Zlodre JK, Rennie AT, Ramsden JD. Langerhans’ cell histiocytosis of the temporal bone: successful treatment of sensorineural hearing loss with low-dose radiotherapy. *J Laryngol Otol*. 2009;123:676-679.

34. Gupta G, Jain A, Grover M. Successful cochlear implantation in Langerhans cell histiocytosis: a rare case. *Cochlear Implants Int*. 2018;19:115-118.

35. Segel JM, McKinnon BJ. Bilateral cochlear implantation in bilateral Langerhans cell histiocytosis. *Cochlear Implants Int*. 2013;14:178-180.

36. Torkos A, Czigner J, Kiss JG, Tóth F, Szamosközi A, Jóri J. Cochlear implantation for treatment-induced ototoxic deafness in Langerhans cell histiocytosis. A case report. *Eur Arch Otorhinolaryngol*. 2005;262:496-500.

37. Nanduri V, Tatevossian R, Sirimanna T. High incidence of hearing loss in long-term survivors of multisystem Langerhans cell histiocytosis. *Pediatr Blood Cancer*. 2010;54:449-453.

38. Kato K, Otake H, Tagaya M, et al. Progressive hearing loss following acquired cytomegalovirus infection in an immunocompromised child. *Am J Otolaryngol*. 2013;34:89-92.

39. Kitazawa K, Matsumoto M, Senda M, et al. Mondini dysplasia and recurrent bacterial meningitis in a girl with relapsing Langerhans cell histiocytosis. *Pediatr Blood Cancer*. 2004;43:85-87.

40. Simmonds JC, Vecchiotti M. Cholesteatoma as a complication of Langerhans cell histiocytosis of the temporal bone: a nationwide cross-sectional analysis. *Int J Pediatr Otorhinolaryngol*. 2017;100:66-70.

**How to cite this article:** Pähler vor der Holte A, Welkoborsky H-J. Case report: Langerhans cell histiocytosis of the temporal bone in children: Challenging diagnosis of a rare disease with some pitfalls. *Clin Case Rep*. 2022;10:e06057. doi: 10.1002/ccr3.6057