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Beyond Ear Discharge: Multisystem Langerhans Cell Histiocytosis with Temporal and Calvarial Involvement in a Child

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Abstract

Introduction: Langerhans cell histiocytosis (LCH) is a rare clonal myeloid neoplasm that commonly affects the bone and skin in children. Temporal bone involvement is uncommon and often mimics chronic otitis media, which may lead to delayed diagnosis.

Case Presentation: We report a 4-year-5-month-old girl who presented with persistent right ear discharge and a visible mass in the external auditory canal for four months. Physical examination revealed diffuse papular and crusted lesions on the scalp and trunk, along with frequent nocturnal urination. The patient had a previous history of right ear tumor excision and parietal craniotomy. Histopathological analysis confirmed Langerhans cell histiocytosis, supported by positive S-100 immunohistochemical staining. Imaging demonstrated a soft-tissue mass obliterating the external ear canal and a parieto-occipital lesion with osseous destruction but no intracranial extension. The patient received vincristine and prednisone chemotherapy following the LCH-IV protocol, resulting in significant clinical improvement.

Conclusion: Temporal bone LCH may clinically resemble chronic ear disease. Early recognition through biopsy and immunohistochemistry is crucial for establishing the diagnosis and initiating timely systemic therapy, thereby improving long-term outcomes.

Keywords: Langerhans Cell Histiocytosis, Temporal Bone, Pediatric Oncology, Calvarial Lesion

Introduction

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by the clonal proliferation of pathologic dendritic cells exhibiting phenotypic features of Langerhans cells.[1] Once considered an inflammatory condition, LCH is now classified as a neoplastic disease of myeloid origin due to the identification of recurrent activating mutations in the MAPK pathway, particularly BRAF V600E and MAP2K1 mutations.[2] The disease displays a highly variable clinical spectrum, ranging from isolated bone lesions to disseminated multisystem involvement with potentially life-threatening organ dysfunction.[3]

The annual incidence of LCH is estimated to be 2–9 cases per million children, with a peak occurrence between 1 and 4 years of age, and a slight male predominance.[4] The most frequently affected organs are the bones (approximately 80%),[5] followed by the skin, pituitary gland, and liver. Involvement of the temporal bone occurs in about 15–30% of pediatric cases,[6] often presenting with otorrhea, postauricular swelling, or hearing loss, symptoms that can mimic chronic otitis media or mastoiditis, leading to delayed or missed diagnosis.[7]

Because of its nonspecific presentation, LCH involving the temporal bone poses a diagnostic challenge, particularly when systemic manifestations are subtle or absent.[8] Accurate diagnosis requires histopathological confirmation, typically demonstrating Langerhans-type cells with grooved nuclei, along with positive immunoreactivity for CD1a, Langerin (CD207), and S-100 proteins.[7,8] Early recognition and initiation of systemic therapy—most commonly with vincristine and corticosteroids under standardized protocols such as LCH-III or LCH-IV—are crucial for improving prognosis and preventing disease progression.[9,10]

Here, we report a case of a 4-year-5-month-old girl with temporal bone and calvarial LCH, who initially presented with persistent ear discharge and scalp lesions, illustrating the importance of clinical suspicion and timely biopsy in atypical ear disease presentations in children.

Case Presentation

A 4-year-5-month-old girl was referred to the pediatric oncology clinic with a four-month history of persistent right ear discharge and the appearance of a visible mass in the right external auditory canal. The symptoms had not improved with multiple courses of antibiotics previously prescribed for presumed chronic otitis media. There was no history of fever or otalgia, and no apparent hearing loss was reported based on parental observation and clinical assessment. Objective hearing evaluation, such as Pure Tone Audiometry, could not be performed due to complete obstruction of the external auditory canal by the mass, which represents a limitation of this case. Her mother noted frequent nocturnal urination and the appearance of crusted papular lesions on the scalp and trunk.

On physical examination, the child appeared well-nourished but had multiple erythematous papules and crusted lesions scattered over the scalp, forehead, and upper trunk. Otoscopic examination revealed a friable, reddish mass filling the right external auditory canal with serosanguinous discharge. The mass completely occluded the external auditory canal, precluding visualization of the tympanic membrane; therefore, the tympanic membrane status could not be assessed at the time of examination. No facial nerve palsy, and other cranial nerves were intact. Systemic examination revealed no hepatosplenomegaly or lymphadenopathy.

Laboratory evaluation revealed a normal complete blood count, liver, and renal function tests. Urinalysis was unremarkable except for mild polyuria suggestive of early diabetes insipidus, consistent with pituitary involvement. Magnetic resonance imaging (MRI) of the head showed a soft-tissue mass obliterating the right external auditory canal, extending into the mastoid air cells with erosion of the temporal bone, but without intracranial extension. High-resolution computed tomography (CT) of the temporal bone performed in February 2023, prior to referral, demonstrated a soft-tissue mass involving the right external auditory meatus with extensive bony destruction, including erosion of the tympanic membrane, zygomatic process of the temporal bone, and adjacent temporal bone structures; these findings were initially suggestive of cholesteatoma. A second lesion was identified in the right parieto-occipital bone, showing lytic destruction of the calvarial table and adjacent soft-tissue enhancement. (Figure 1)

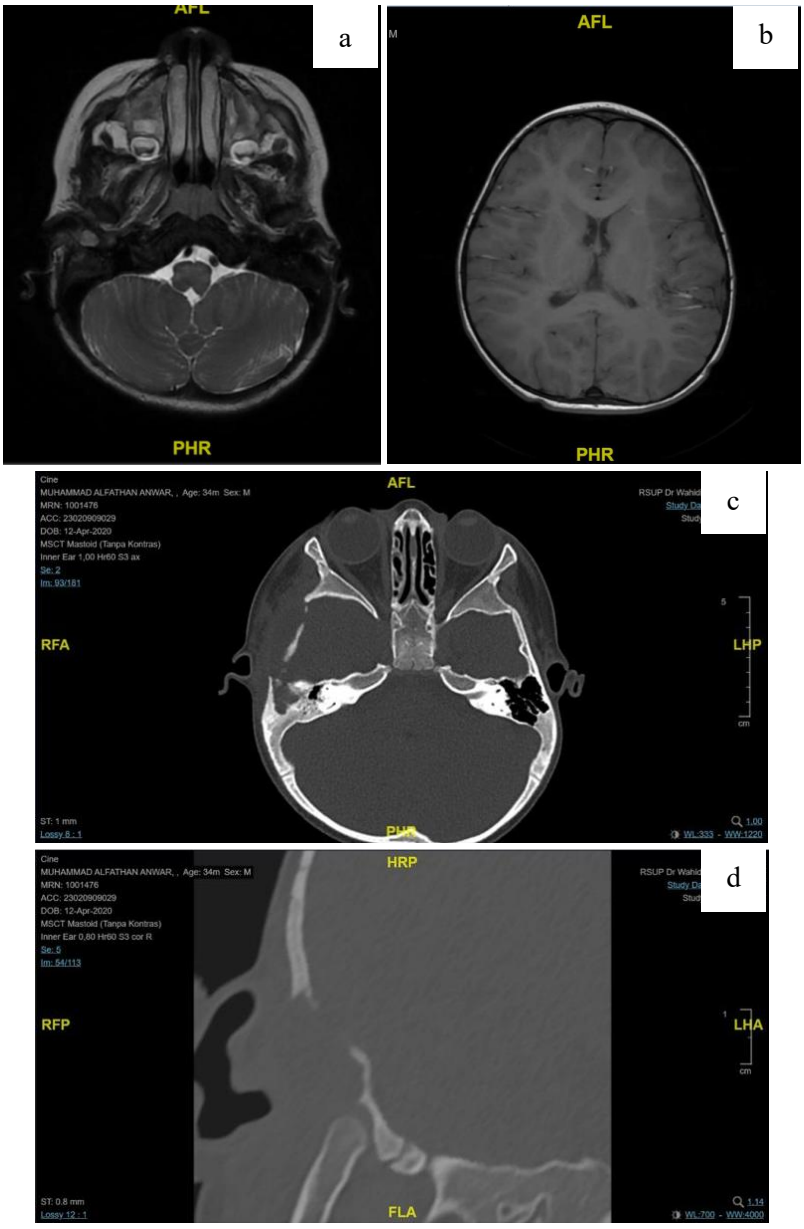




Figure 1. (A) A Soft-Tissue Mass Obliterating the Right External Auditory Canal, Extending into the Mastoid Air Cells with Erosion of the Temporal Bone, But Without Intracranial Extension. (B) A Second Lesion Was Identified in the Right Parieto-Occipital Bone, Showing Lytic Destruction of the Calvarial Table and Adjacent Soft-Tissue Enhancement. (C-E) High-Resolution CT Scan of the Temporal Bone (Bone Window) Demonstrating Extensive Bony Erosion of the External Auditory Canal and Surrounding Temporal Bone Structures.

The patient had a prior history of surgical excision of a right ear canal mass and parietal craniotomy performed at another hospital, but the histopathological diagnosis at that time was inconclusive. The first symptoms appeared in August 2022, and the patient subsequently underwent three surgical procedures, with the most recent performed approximately one month prior to the current admission, reflecting a prolonged and recurrent disease course. Specifically, excision of granulation tissue was performed in February 2023, followed by canal wall up tympanomastoidectomy in March 2023 due to persistent otorrhea, and a third procedure consisting of craniotomy and revision mastoidectomy five months later, after which histopathological examination confirmed Langerhans cell histiocytosis.

Histopathological examination revealed proliferation of large histiocytic cells with oval to reniform nuclei, nuclear grooves, and moderate eosinophilic cytoplasm, accompanied by an eosinophil-rich inflammatory infiltrate. Immunohistochemical staining showed positivity for S-100, confirming the diagnosis of Langerhans cell histiocytosis (LCH). (Figure 2)

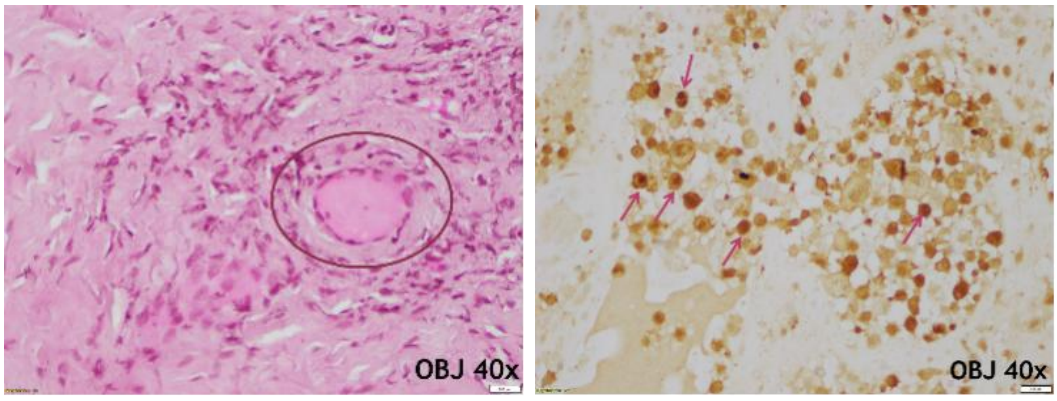


Figure 2. (A) A Proliferation of Large Histiocytic Cells with Oval to Reniform Nuclei, Nuclear Grooves, and Moderate Eosinophilic Cytoplasm, Accompanied by A Mixed Inflammatory Infiltrate Rich in Eosinophils. (B) Immunohistochemical Staining Showed the Lesional Cells Were Positive For S-100

Given the presence of multifocal bone involvement with possible endocrine dysfunction, the disease was classified as multisystem LCH. The patient was initiated on systemic chemotherapy according to the LCH-IV protocol, consisting of vincristine (6 mg/m^2 weekly) and prednisone (40 mg/m^2/day for 4 weeks) as the initial phase of therapy. Local otolaryngology management included regular aural toilet and topical therapy; however, definitive local surgical treatment was limited due to the extent of the mass. Patient care was coordinated through a multidisciplinary team involving otolaryngology, pediatric oncology, radiology, pathology, and neurosurgery.

After the first two cycles of chemotherapy, the patient demonstrated marked clinical improvement. The ear discharge resolved completely, the external auditory canal mass regressed, and the scalp lesions healed significantly. Follow-up imaging after three months revealed a reduction in the size of both temporal and parieto-occipital lesions, with evidence of early bone healing. Post-treatment audiometric evaluation has not yet been performed; however, subjective improvement in hearing was reported by the parents. The patient's diabetes insipidus remained clinically stable under medical management. Long-term follow-up is planned for at least five years, including clinical evaluation, endocrine monitoring, radiologic surveillance, and otolaryngologic follow-up.

Discussion

Langerhans cell histiocytosis (LCH) represents a rare clonal proliferative disorder of myeloid dendritic cells characterized by the accumulation of CD1a+, Langerin (CD207)+, and S-100+ histiocytes in one or multiple organs.[5] Although once considered an inflammatory disease, advances in molecular biology have established LCH as a neoplastic disorder driven by mutations within the mitogen-activated protein kinase (MAPK) pathway, most commonly BRAF V600E, which is detected in approximately 50–60% of cases.[10] These molecular insights have redefined the disease as a clonal myeloid neoplasm with inflammatory features, accounting for its variable clinical spectrum and unpredictable course.[11]

The estimated annual incidence of LCH is 2–9 per million children, with a peak age of onset between 1 and 4 years, and a slight male predominance.[2,5] Bone is the most commonly affected organ, involved in approximately 80% of pediatric cases.[4] Common skeletal sites include the skull, ribs, femur, and vertebrae.[12] Temporal bone involvement occurs in 15–30% of pediatric LCH cases, making it the third most frequent site of cranial involvement, after the parietal and frontal bones.[13,14] Because symptoms such as chronic otorrhea, postauricular swelling, otalgia, or hearing loss overlap with those of chronic otitis media or mastoiditis, the diagnosis is often delayed or missed.[13]

In the present case, the child initially presented with persistent otorrhea and a visible mass in the external auditory canal, leading to an initial misdiagnosis of chronic ear infection. The otorrhea persisted for approximately one year prior to definitive diagnosis, during which multiple courses of empirical antibiotic therapy were administered without sustained clinical improvement, highlighting a significant diagnostic delay. This clinical scenario is consistent with prior reports, such as those by Gendre et al. (2023) and Ocal et al. (2022), which emphasize that LCH of the temporal bone may clinically and radiographically mimic chronic suppurative otitis

media, cholesteatoma, or even malignancy.[7,15] Such overlapping features underscore the necessity of maintaining a high index of suspicion in children with atypical or refractory ear disease, particularly when accompanied by osseous destruction on imaging or cutaneous lesions elsewhere.

Radiologically, LCH lesions are typically described as punched-out osteolytic defects without sclerotic margins, sometimes with a soft-tissue component extending beyond the bone cortex.[16] In our case, MRI and CT imaging revealed a soft-tissue mass obliterating the right external auditory canal, extending into the mastoid air cells with bony erosion, and an additional parieto-occipital lesion, consistent with multifocal bone disease. High-resolution computed tomography of the temporal bone further demonstrated extensive bony destruction of the external auditory canal and adjacent temporal bone structures, emphasizing the importance of CT imaging in evaluating the extent of osseous involvement and differentiating LCH from other destructive otologic pathologies.

Histopathological and immunohistochemical confirmation remain the gold standard for diagnosis. The presence of Langerhans-type histiocytes with grooved or "coffee-bean" nuclei and positive staining for S-100, CD1a, and Langerin (CD207) is an diagnostic hallmark.[17] In our patient, histopathology demonstrated classic morphologic features of LCH, and immunohistochemical staining showed positivity for S-100. Although additional immunohistochemical staining for CD1a or Langerin could not be performed due to facility limitations at the time of evaluation, which represents a diagnostic limitation, the diagnosis was strongly supported by characteristic histopathological findings, clinicopathological correlation, and a favorable clinical response to standard LCH-directed chemotherapy.

The association between LCH and endocrine abnormalities, particularly central diabetes insipidus (DI), is well-documented, occurring in 15–25% of cases, especially with hypothalamic-pituitary axis involvement.[18] In this case, frequent nocturnal urination suggested early DI. However, formal post-treatment endocrine reassessment was not comprehensively documented, and the objective post-therapy status of DI could not be definitively established, underscoring the need for structured and long-term endocrine surveillance in patients with multisystem LCH.

Management of LCH depends on the disease extent. Single-system LCH with isolated bone involvement can often be treated with local curettage, limited radiation, or intralesional corticosteroids.[19] However, multisystem or multifocal bone disease requires systemic therapy. Optimal management requires a multidisciplinary approach involving pediatric oncologists, otolaryngologists, radiologists, pathologists, and endocrinologists. The role of the otolaryngologist is particularly important in cases with temporal bone involvement, primarily for diagnostic biopsy, management of secondary infection, and long-term monitoring of otologic complications. Surgical intervention is generally limited to diagnostic or symptomatic purposes, while extensive surgical debridement is not routinely recommended due to the systemic nature of the disease. The LCH-IV protocol, which includes vincristine and prednisone as first-line agents, has demonstrated excellent efficacy and tolerability in children. Our patient responded favorably to this regimen, with complete resolution of ear discharge and regression of bone lesions after several cycles, consistent with response rates reported in the literature.

Prognosis in LCH varies widely. Patients with multisystem disease involving "risk organs" (liver, spleen, or bone marrow) have a poorer outcome, while those without such involvement generally achieve long-term remission with standard therapy.[20,21] Based on current risk stratification, our patient was classified as having multisystem LCH without risk-organ involvement, a category generally associated with a favorable long-term prognosis when early and sustained response to first-line therapy is achieved. Given the marked clinical and radiologic improvement observed, the expected long-term outcome in this patient is favorable, although prolonged monitoring is required to detect relapse or late sequelae such as hearing impairment or endocrine dysfunction.

This case reinforces several key clinical lessons. First, persistent otorrhea or ear canal masses unresponsive to standard therapy should raise suspicion for LCH, particularly in children with additional skin or bone lesions. Second, histopathological confirmation with immunohistochemistry remains essential for accurate diagnosis. Finally, systemic therapy guided by current international protocols such as LCH-IV can result in excellent clinical outcomes, even in multifocal disease. Fourth, high-resolution computed tomography of the temporal bone should be routinely obtained in suspected cases to fully assess the extent of bony involvement and to guide surgical planning when indicated.

Limitations

This case report has several limitations. First, complete immunohistochemical profiling with CD1a and Langerin (CD207) could not be performed due to facility limitations at the time of evaluation, which may reduce absolute diagnostic certainty, although clinicopathological correlation and favorable response to LCH-directed chemotherapy strongly supported the diagnosis. Second, high-resolution computed tomography of the temporal bone was not available during the early phase of the initial diagnostic workup, which limited timely assessment of fine bony structures and ossicular chain involvement. Third, objective audiological evaluation was not systematically performed, precluding a comprehensive assessment of hearing status before and after treatment. Fourth, structured endocrine follow-up, particularly regarding diabetes insipidus, was not fully documented following systemic therapy initiation, limiting objective evaluation of endocrine outcomes. Finally, the exact timeline and details of prior surgical intervention could not be fully ascertained from available medical records. Despite these limitations, this case provides valuable insight into the diagnostic challenges, multidisciplinary management, and favorable treatment response of pediatric temporal bone Langerhans cell histiocytosis.

Conclusion

Langerhans cell histiocytosis involving the temporal bone is an uncommon manifestation that can easily be mistaken for chronic otitis media or mastoiditis, leading to delayed diagnosis and treatment. This case highlights the importance of maintaining a high index of suspicion for LCH in children presenting with persistent otorrhea or ear canal masses unresponsive to conventional therapy—particularly when associated with cutaneous or skeletal lesions. Histopathological confirmation with immunohistochemical staining for CD1a, S-100, and Langerin remains essential for accurate diagnosis. Early initiation of systemic chemotherapy

according to established protocols such as LCH-IV can result in excellent clinical and radiologic responses, significantly improving long-term outcomes and reducing the risk of permanent complications.

Conflict of Interest

The authors declare that there are no conflicts of interest related to this study.

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