Case Report

Langerhans' cell histiocytosis presenting as chronic otitis media a diagnostic dilemma

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ABSTRACT

Langerhans' cell histiocytosis (LCH) is a rare condition that can sometimes affect the temporal bone. When it involves the temporal bone, LCH may present with non-specific symptoms such as middle ear inflammation, ear pain, hard of hearing, external auditory canal polyps, or mastoid mucosal inflammation. In cases where patients show signs of chronic otitis media, LCH should be considered a possible differential diagnosis. This case report highlights a 38-year-old male patient who initially presented with features suggestive of chronic otitis media, was diagnosed to have LCH through histopathological analysis, and was treated according to medical oncology protocols.

Key words: Chronic otitis media, Langerhans cell, Langerhans' cell histiocytosis, Temporal bone Langerhans cell histiocytosis

Langerhans' cell histiocytosis (LCH) is a rare disorder characterized by the abnormal proliferation of Langerhans cells. Initially known as histiocytosis X, the disease was renamed in 1985 after Paul Langerhans, the German pathologist who first identified these cells [1]. Historically, LCH was categorized into three subtypes: Eosinophilic granuloma (EG), Hand–Schüller–Christian disease, and Letterer–Siwe disease [2]. EG involves a single osteolytic lesion without systemic symptoms. Hand–Schüller–Christian disease typically affects children under 5, presenting with multiple bone lesions and sometimes the triad of exophthalmos, diabetes insipidus, and a skull lesion, though this occurs in only one-third of cases. Letterer–Siwe disease, seen in children under 3, involves multiple organ systems and carries a poor prognosis. The more recent classification by the Histiocytic Society divides LCH into three categories: Class I (LCH), Class II (non-Langerhans' cell histiocytosis), and Class III (Malignant LCH), with disease presentation either localized or widespread [3]. When LCH affects the temporal bone, diagnosis is often delayed due to vague symptoms such as middle ear inflammation, ear discharge, otalgia, hard of hearing, external ear canal polyps, or mastoid mucosal inflammation. Imaging techniques such as magnetic resonance imaging (MRI) and computed tomography (CT) scans can aid diagnosis, but a high degree of clinical suspicion is required since otologic symptoms may be overshadowed by systemic manifestations and ear-related findings often mimic more common ear conditions [4]. The prognosis of LCH is closely related to age of onset, number of involved organs, and degree of functional lesion. The prognosis of single-organ involvement is better than multiplied-organ involvement, and the latter has a higher case-fatality rate. Adult LCH and a single site involvement normally have a good prognosis. Hence, maintaining a high index of suspicion for the likelihood of LCH in cases as elaborated below can benefit the perplexed surgeon in making a diagnosis, to aid the suffering patient.

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CASE REPORT

A male patient aged 38 years old, developed ear discharge 6 months prior, from the right ear which was scanty, mucopurulent, and non-foul smelling, it was occasionally blood-stained. He consulted at a local hospital for the same and was managed conservatively with topical antibiotics. The patient had improvement in symptoms after treatment but suffered from repeated episodes over 1 month. He also noticed a decrease in hearing in the right ear, which was insidious in onset and gradually progressive with the inability to hear soft sounds, not affecting his daily activities.

He developed sudden-onset giddiness a few weeks later which was associated with a feeling of imbalance, non-positional, non-rotatory, each episode lasting for a few seconds, tinnitus in the right ear, which was insidious in onset and gradually progressive not associated with blurring of vision, palpitations, sweating, or loss of consciousness. He consulted with a neurologist for the same.

He was advised a MRI of the brain which showed an ill-defined extra-axial mass lesion in the supratentorial region along

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the left posterior parietal convexity in the parasagittal region. Another similar aggressive lytic lesion was seen in the right middle ear eroding the posterior wall of the external auditory canal and mastoid air cells.

He presented to our outpatient department with the above details. He was vitally stable, with a pulse rate of 88 beats/minute, blood pressure of 120 by 80 mm of mercury, and a respirator rate of 16 cycles/min. He was moderately built and nourished, with no significant findings on general examination. Local examination revealed a proliferative lesion in the external auditory canal in the posteroinferior and posterosuperior aspect about 1–2 mm from the tympanic membrane, with an intact tympanic membrane.

The patient was further evaluated with pure tone audiometry which showed moderate conductive hearing loss in the right ear and FDG-positron emission tomography (FDG-PETCT). The FDG-PETCT scan revealed FDG uptake in the extra-axial enhancing lesion in the left parietal convexity in the parasagittal region with erosion of the inner table of adjacent parietal bone (size 1.3 × 2.3 cm SUV max 14.8) FDG uptake was seen in the lytic lesion with soft-tissue component involving right petrous temporal bone and mastoid air cells (size 2.3 × 2.4 cm SUV max 9). FDG uptake in right upper paratracheal, lower paratracheal nodes (largest measuring 1.6 × 1 cm, SUV max 8.8), and sub-centimetric subcarinal lymph node (SUV max 5.3). Mild diffuse FDG uptake was seen in the marrow of multiple bones of the axial and appendicular skeleton, likely due to marrow activation, the visualized bones were otherwise normal with no lytic or sclerotic lesion or focal abnormal FDG uptake, as per the report. On discussion with the radiologist, histopathological analysis was advised.

After a pre-anesthesia checkup and obtaining informed consent from the patient, he underwent cortical mastoidectomy and biopsy under general anesthesia. Intraoperatively it was found that the periosteum overlying the mastoid antrum and posterior canal wall was eroded by the proliferative mass filling the mastoid cavity extending, as mentioned into the external auditory canal, the limits could not be clearly defined. The excised tissue was sent for histopathological studies which revealed fragments of tissue with a dense lymphohistiocytic infiltrate admixed with foamy macrophages, occasional fragments showing many eosinophils admixed with histiocytes and lymphocytes (Fig. 1).

Immunohistochemistry studies showed occasional focal aggregates of CD1a and S100 positive histiocytes, CD68 highlighting the numerous macrophages in the background, and CD20, and CD3 highlighting in the background suggestive of LCH (Fig. 2).

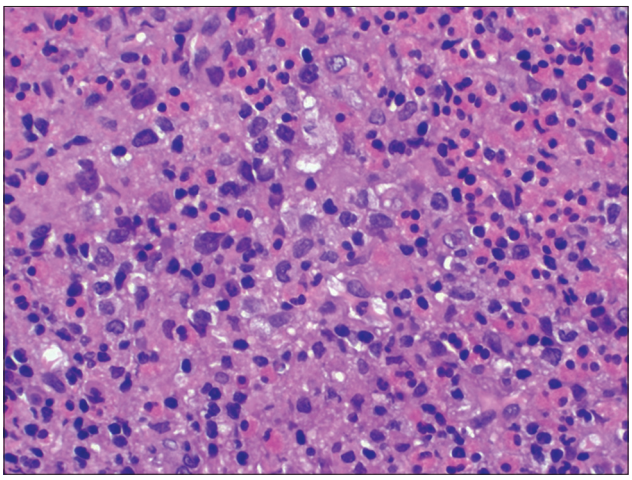

Figure 1: Hematoxylin and Eosin (H&E) staining under low-power microscopic examination (×40) showing dense lymphohistiocytic infiltrate admixed with foamy macrophages and eosinophils admixed with histiocytes and lymphocytes

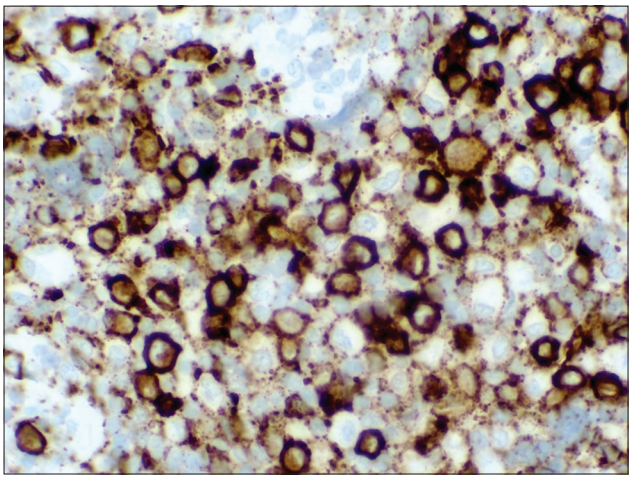

Figure 2: Immunohistochemistry by polymer technique under low-power microscopic examination (×40) showing positive CD1a histiocytes. CD68 highlights the numerous macrophages in the background. CD20 and CD3 highlight the background mixed population of B and T lymphocytes

DISCUSSION

LCH is a rare disorder marked by osteolytic bone lesions containing histiocytes derived from mononuclear phagocytic and dendritic cells. It often affects the head and neck, with 50–80% of cases involving these regions [5]. The estimated incidence of LCH is 2–5 cases/million people/year [6], with children aged 1–3 being the most affected age group (4–5 cases/million) [7]. LCH typically presents as either unifocal or multifocal, with unifocal involvement accounting for 70% of cases. Temporal bone involvement occurs in 15–60% of cases, primarily affecting the mastoid, middle ear, or external auditory canal.

LCH can affect different organ systems, leading to a wide range of clinical symptoms depending on the tissues involved, such as the skin, bones, lymph nodes, or internal organs such as the lungs, liver, and spleen. The way it is presented can also differ by age. For example, skin rashes are a common early symptom in children and are sometimes mistaken for seborrheic or atopic dermatitis that does not respond well to typical topical treatments. The rash associated with LCH can vary from a single lesion to widespread coverage and may include scaly papules, nodules, or plaques, often resembling seborrheic dermatitis. However, some cases can present with a normal physical examination and unifocal diseases as seen in patients presenting to the otorhinolaryngology outpatient department with temporal bone localization.

Qurashi *et al* state that the otorhinolaryngologist needs to be familiar with this temporal bone localization as, in their study, 50% of the patients with LCH had aural symptoms at presentation. The symptomatology is indistinguishable from otitis media or otitis externa, as it stimulates mastoiditis, cholesteatoma, or even metastatic disease of the temporal bone. The presence of granulation tissue in the middle ear without suppuration is suggestive of LCH [8].

Hence, misdiagnosis of temporal bone LCH is common due to non-specific symptoms such as purulent ear discharge, ear pain, hearing loss, and sometimes vertigo, which are seen in 5–25% of cases [9]. The prognosis largely depends on the number of organs involved, organ dysfunction, patient age, and whether high-risk organs (spleen, liver, hematopoietic system, and possibly lungs) are affected [10].

For diagnosis, CT is recommended as the first-line imaging, revealing bone destruction and soft-tissue changes. Typical findings are usually described as lytic, punched-out, destructive, or erosive lesions, leading to a wide differential diagnosis. MRI is used to assess disease spread or exclude other conditions. A definitive diagnosis is made through histopathological analysis, which reveals multinucleated Langerhans cells along with eosinophils, neutrophils, and lymphocytes. Electron microscopy shows Birbeck granules, and immunohistochemistry confirms positivity for CD1a and/or CD207 (Langerin), markers of immature dendritic cells [11].

In the past low dose irradiation was the treatment of bony lesions with risk of necrosis and pain. At present, chemotherapy is the first line of management with radiation being preserved for specific situations. Vinblastine is the first-line chemotherapeutic agent used along with systemic steroids of which prednisolone is preferred.

Isolated otologic lesions with a low risk of causing serious complications can often be managed with conservative surgical methods, including curettage, excision, or mastoidectomy, depending on the disease's spread. For single-system, single-location bone involvement, complete removal of a bone lesion is generally an option if it is smaller than 2 cm, whereas curettage or partial excision may be used for lesions measuring between 2 and 5 cm as per Haupt *et al*. [12].

Unifocal LCH has an excellent prognosis, with up to 100% survival at 5 years, treated through surgery or chemotherapy. In cases of multisystem disease, the 5-year survival rate is 98%. Mortality differs based on the site of involvement 0.9% for unifocal bone lesions and 30–84% for multisystem involvement. Temporal bone involvement carries a higher risk of central nervous system complications, making systemic therapy the standard of care [13].

CONCLUSION

LCH is a rare disorder characterized by the abnormal proliferation of Langerhans cells. The prognosis of LCH is closely related to age of onset, number of involved organs, and degree of functional lesion. Adult LCH and a single site involvement normally have a good prognosis.

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