ACTA SCIENTIFIC OTOLARYNGOLOGY

Volume 1 Issue 1 September 2019

Case Report

Langerhan's Cell Histiocytosis Mimicking as Atticoantral disease

Munish Saroch[1], Amit Saini[2]* and Gaveshna Gargi[3]

[1]*Professor, Department of otolaryngology, Dr RP Govt. Medical College, Tanda, Kangra, Himachal Pradesh, India*

[2]*Medical Officer, Department of otolaryngology, Dr RP Govt. Medical College, Tanda, Kangra, Himachal Pradesh, India*

[3]*Assistant Professor, Department of Medicine, Dr RP Govt. Medical College, Tanda, Kangra, Himachal Pradesh, India*

***Corresponding Author:** Amit Saini, Medical Officer, Department of otolaryngology, Dr RP Govt. Medical College, Tanda, Kangra, Himachal Pradesh, India.

Received: August 26, 2019; **Published:** September 06, 2019

Abstract

Eosinophilic granuloma (EG) is the localised benign form of Langerhan's Cell Histiocytosis (LCH). Very rarely EG can involve temporal bone and mimic common otologic disorders like otitis externa, otitis media, aural polyps, acute mastoiditis etc. We report a case of 6 year old child who presented with aural polyp in right ear and was misdiagnosed as CSOM atticoantral type, HRCT Temporal bone showed soft tissue mass in mastoid tip cells with erosion of posterior canal wall on right side. Patient underwent modified radical mastoidectomy right ear and was subsequently found to have Langerhan's Cell Histiocytosis on histopathology. Child was put on systemic steroids and showed no recurrence. Eosinophilic granuloma can mimic as chronic suppurative otitis media which can delay the diagnosis and treatment of this condition.

Keywords: Langerhan's Cell Histiocytosis; Eosinophilic Granuloma; Histiocytosis X; CSOM Atticoantral

Introduction

Langerhan's Cell Histiocytosis (LCH) is a rare disease involving multiple organs with an incidence of approximately 5.4 per million. LCH is found more in males and is common in children of 1 to 4 years of age [1]. LCH was formerly named as Histiocytosis X by Liechtenstein in 1953 as the cause of disease was unknown [2]. In 1985 during the meeting of Histiocytosis Society it was decided that term "Langerhan's Cell Histiocytosis" would replace all the previous nomenclature describing this disease including Histiocytosis X [3]. LCH involves the reticuloendothelial system and has three different variants Eosinophilic granuloma (EG), Hand–Schuller–Christian (HSC) and Letterer–Siewe (LS) diseases.

The two extremes of this disease are clinically identified. The most common form is solitary eosinophilic granuloma of bone, which has excellent prognosis [4]. At the other end of the spectrum, the disease affects multiple organs and tissues including bones, skin, pituitary, lung, liver, and the hematopoietic system [5]. This involvement may cause multiple organ dysfunctions and have a fatal outcome. A high index of suspicion is required to recognize the otologic manifestations of histiocytosis X for two reasons: the systemic manifestation of the disease is often so dramatic that the ear findings are overlooked, and the otologic findings of LCH can mimic more common diseases, including simple otitis externa, otitis media, aural polyps, acute mastoiditis, chronic otitis media,

and metastatic lesions. In the present report, a child with EG is presented, who was initially misdiagnosed and treated for chronic suppurative otitis media (CSOM) atticoantral type. The diagnosis and the management of this relatively rare disease are discussed.

Case Report

We report a case of 6 year old male child who presented to the outpatient department with history of discharge from right ear since six months and decreased hearing from right ear since last three months. Otoscopic examination of ear revealed intact tympanic membrane on left side and polypoidal mass completely filling the right external auditory canal. The pure tone air conduction average was 60dB on right side with normal hearing on left side. High Resolution Computed Tomography (HRCT) of temporal bone was performed and it revealed soft tissue density present in right mastoid antrum tip cell area with erosion of posterior canal wall and sinus plate (Figure 1). Diagnosis of chronic supurrative otitis media atticoantral type right ear was kept. Patient was planned for mastoid exploration.

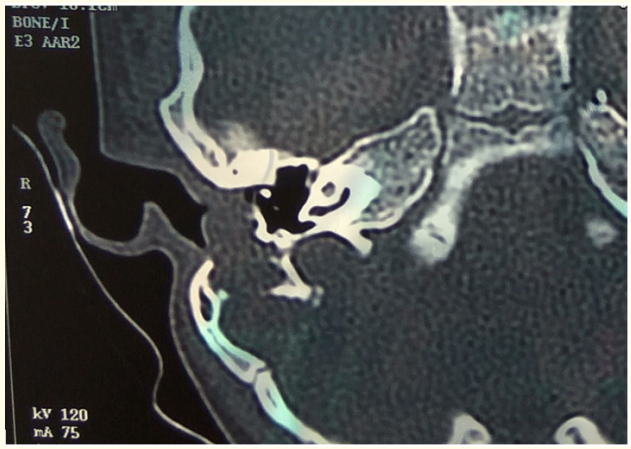

Figure 1: HRCT image showing soft tissue in right mastoid tip cells with posterior canal wall dehiscence.

During surgery it was found that there was soft tissue mass in the mastoid tip cell region which had eroded the posterior canal wall entered middle ear and caused necrosis of right incudostapedial joint (Figure 2).

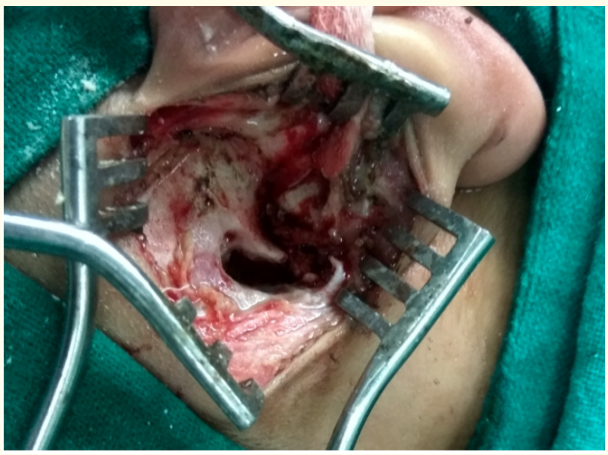

Figure 2: Intraoperative image with posterior canal wall dehiscence and soft tissue mass.

Modified Radical Mastoidectomy was performed on right side with type 3 tympanoplasty and malleostapedopexy (Figure 3). Wide meatoplasty was performed and wound was closed in layers. The soft tissue was sent for histopathological examination and it revealed sheets of atypical histiocytes having grooved nuclei and moderate ill defined eosinophillic cytoplasm along with presence of eosinophils in background and possibility of EG was kept.

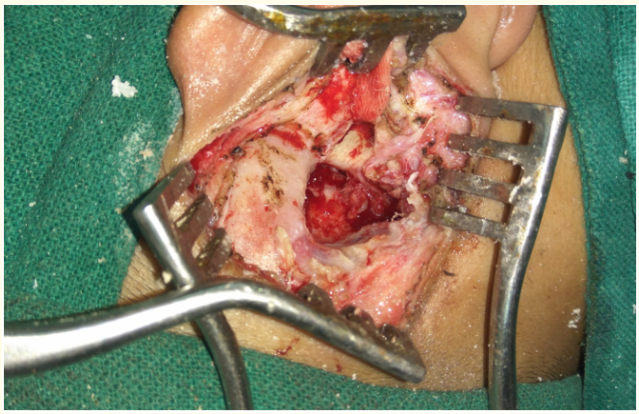

Figure 3: Modified radical mastoidectomy completed.

Complete body survey to consider metastasis with skeletal survey was normal and post operative pure tone air conduction average improved to 40 dB. The patient was put on oral steroids for two months and has been on follow up since eighteen months without any local or systemic recurrence (Figure 4).

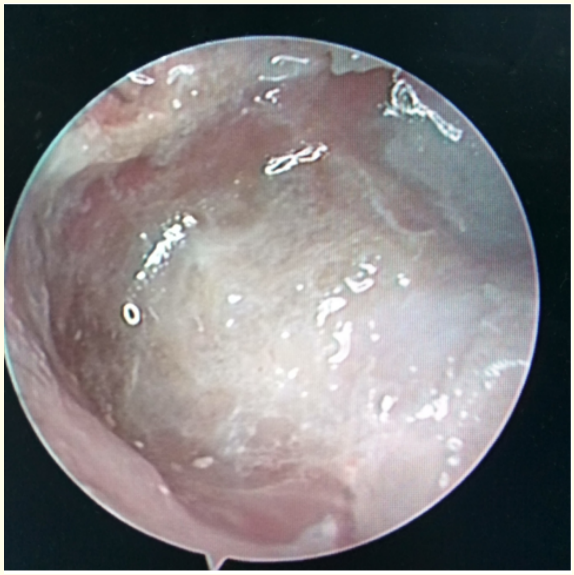

Figure 4: Endoscopic view of mastoid cavity at six months postoperatively without any recurrence.

Discussion

Though LCH is an benign disease it is locally destructive and can involve multiple organs. The histologic hallmark of this disease is a proliferation of the Langerhans dendritic cell.

Langerhans cells are antigen presenting cells that have been found in peripheral tissues, such as skin, vaginal mucosa, buccal mucosa, trachea, thymus, blood vessels and lymph nodes. LCH is characterized by abnormal proliferation of Langerhans cells with a granular infiltration. Although epidermal Langerhans cell has been presumed to be the cell of origin in LCH, recent studies showed that other cell groups including dermal langerin$^+$ cells, lymphoid tissue-resident langerin$^+$ dendritic cells and monocytes could have been induced by local environmental stimuli to acquire LCH phenotype. EG most commonly occurs in the long bones of the skeleton such as femur, ribs, vertebrae and EG of the craniomaxillofacial bones are quietly rare. Temporal bone is the most common site for EG that followed by mandible in this region [6,7].

Eosinophilic granuloma (EG) is the benign form of Langerhans Cell Histiocytosis. EG accounts for up to 55-60% of all cases of LCH. The prognosis of EG is better than the LS and HSC diseases with 90% survival rate in which spontaneous resolution of solitary or multiple lytic bone lesions may also be seen in EG. About 50~ 80% of paediatric LCH is found in the head and neck regions. The temporal bone is involved approximately 15 to 60% of cases in this region. Otologic presentations include mastoid swelling or temporal bone mass, otalgia, and otorrhea. The otologic findings are very similar to the otitis media, otitis externa, cholesteatoma, and the other conditions; for this reason, the diagnosis is usually delayed and it causes management dilemma [8].

In the present case also the condition was mimicking as CSOM atticoantral variety and definite diagnosis was only made after histopathology. But fortunately in our patient disease was solitary and complete resolution was achieved and no harm was done to the child.

The patients who are diagnosed as EG should undergo complete work up to know the extent of the disease. The battery of tests which should be performed include hematologic evaluation, erythrocyte sedimentation rate, liver function tests, urinalysis, chest X ray, bone scintigraphy, and metastatic series [2].

The treatment of disease depends upon the extent at time of diagnosis. The localised form of LCH with solitary bone lesion requires local curettage. In an asymptomatic patient wait and watch policy can also be used. The systemic version of the disease requires extensive treatment, particularly in young children. The drugs used in this form of disease are corticosteroids, vinca alkaloids, mercaptopurine, methotrexate, and etoposide (VP16). Recent reports of successful treatment of recurrent or refractory disease with 2 chlorodeoxyadenosine, a purine analogue, and cyclosporine, an immune modulator, offer a welcome addition to management of LCH [9]. These cytotoxic agents should be used with caution in children after carefully weighing the risks versus benefits.

Radiotherapy has also been found to be beneficial in these cases. But in head and neck region radiotherapy used with caution as it can cause serious damage to vital organs. In addition, there are reports of secondary malignancy resulting from radiation treatment [10].

Conclusion

LCH is a multifocal disease which can present with ear symptoms and can cause dilemma in its management. High index of suspicion should be made when extensive bone erosion is seen on

HRCT temporal bone. Whenever diagnosis of LCH is confirmed histopatologically extensive search should be made for systemic involvement and close follow up should be obtained.

Funding

There are no funding source for this study.

Conflict of Interest

Authors declare that there are no conflict of interest.

Consent of Ethics

We confirm that this work is original and has not been published elsewhere nor is it currently under consideration for publication elsewhere.

Informed Consent

Informed consent was obtained from parents of patient.

Bibliography

1. Saliba I., *et al*. "Langerhans cell histiocytosis of the temporal bone in children". *International Journal of Pediatric Otorhinolaryngoly* 72 (2008): 775Y86.

2. Appling D., *et al*. "Eosinophilic granuloma in the temporal bone and skull". *Otolaryngoly Head Neck Surgery* 91 (1983): 358-365.

3. Dimentberg RA and Brown KL. "Diagnostic evaluation of patients with histiocytosis X". *Journal of Pediatric Orthopaedics* 10 (1990): 733-741.

4. Smith RJ and Evans JN. "Head and neck manifestations of histiocytosis X". *Laryngoscope* 94 (1984): 395-399.

5. Jones RO and Pillsbury HC. "Histiocytosis X of the head and neck". *Laryngoscope* 94 (1984): 1031-1035.

6. Merad M., *et al*. "Origin, homeostasis and function of Langerhans cells and other langerin-expressing dendritic cells". *Nature Reviews Immunology* 8 (2008): 935-947.

7. Egeler RM., *et al*. "Langerhans cell histiocytosis: fascinating dynamics of the dendritic cell-macrophage lineage". *Immunological Reviews* 234 (2010): 213-232.

8. Hui Zheng., *et al*. "Pediatric Langerhans cell histiocytosis of the temporal bone: clinical and imaging studies of 27 case". *World Journal of Surgical Oncology* 16 (2018): 72.

9. Stine KC., *et al*. "Efficacy of continuous infusion 2-CDA (cladribine) in pediatric patients with Langerhans cell histiocytosis". *Pediatric Blood Cancer* 43 (2004): 81-84.

10. Gold DG., *et al*. "Second neoplasms after megavoltage radiation for pediatric tumors". *Cancer* 97 (2003): 2588-2596.

Volume 1 Issue 1 September 2019