A Rare Cause of Otalgia - Langerhans Cell Histiocytosis Case Report.

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Abstract

BACKGROUND: Langerhans Cell Histiocytosis (LCH) is a myeloid neoplastic disorder with a rare prevalence of one case per million adults. LCH has a complicated etiology that presents as lesions composed of proliferative 'LCH cells'. These lesions can affect different organ systems including the lungs, skin, liver, and bone. This study reports the steps toward LCH diagnosis in an adult, the current state of literature in regards to LCH of the temporal bone, and important takeaways for ENT physicians.

CASE REPORT: We describe the case of a 52 year old woman with a two year history of seemingly idiopathic Central Diabetes Insipidus (CDI) complaining of right otalgia for two weeks unresponsive to antibiotic therapy. The patient has a normal otoscopy and audiogram, and an elevated CRP test (22 mg/dl). Further investigations with CT and MRI confirmed destruction of the right mastoid air cells and enhancing tissue respectively. Cholesteatoma was ruled out with diffusion weighted imaging on MRI. Cortical mastoidectomy was performed and histopathological examination of biopsy confirmed the diagnosis of LCH. The patient has a final multisystem (Temporal bone and Pituitary gland) low-risk LCH diagnosis with a great prognosis through chemotherapy and corticosteroid treatment.

CONCLUSION: Temporal bone and craniofacial manifestations of LCH are often managed by ENT physicians. A plethora of pathologies present with otalgia in ENT practice, but symptoms such as CDI and otalgia with lack of findings on otoscopy should raise suspicion for LCH.