International Journal of Pediatric Otorhinolaryngology 100 (2017) 66–70

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Cholesteatoma as a complication of Langerhans Cell Histiocytosis of the temporal bone: A nationwide cross-sectional analysis

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ARTICLE INFO

Article history:
Received 28 March 2017
Received in revised form
19 June 2017
Accepted 20 June 2017
Available online 22 June 2017

Keywords:
Langerhans cell histiocytosis
Langerhans cell histiocytosis of temporal bone
Temporal bone malignancy
External auditory canal cholesteatoma
Cholesteatoma
National inpatient sample
Kids inpatient database

ABSTRACT

Objective: To determine if patients with Langerhans Cell Histiocytosis (LCH) of the temporal bone have a higher risk of developing cholesteatoma.
Methods: Review of literature and cross-sectional weighted analysis of patients under 19 with a diagnosis of LCH from the National Inpatient Sample (NIS) and Kids' Inpatient Database (KID) from 2000 to 2013. ICD-9 codes and demographics were analyzed; pairwise comparisons and multivariate analyses were performed.
Results: Only seven cases of cholesteatoma after the treatment for LCH of the temporal bone have been documented in the literature. No significant association between cholesteatoma and LCH was seen (OR 0.747 [0.149–3.751]). Patients with LCH did have a higher incidence of chronic otitis media, chronic otitis externa, chronic sinusitis, hearing loss, and otitis media with effusion.
Conclusion: Our results show that patients with Langerhans Cell Histiocytosis do not appear to have a higher risk of developing cholesteatoma. However they are more likely to be diagnosed with chronic otitis externa which should be differentiated from cholesteatoma or recurrence of LCH.

1. Introduction

When Alfred Hand Jr., a 25-year old resident at the Children's Hospital of Philadelphia, reported on the death of a 3-year old who had presented with exophthalmia, hepatosplenomegaly and polyuria, he suspected that the was dealing with tuberculosis. It was only 28 years later when he noted the similarities between his case and those described by Schuller and Christian in Germany that he realized that he was dealing with a distinct entity – a disease today known as Langerhans Cell Histiocytosis (LCH) [1].

LCH is thought to be a myeloproliferative process typically seen in children that has a variable presentation. It is characterized by abnormal accumulation of histiocytes which leads to secretion of cytokines leading to an inflammatory response that causes lytic lesions in bone, destruction of the pituitary, and infiltration of the liver, spleen, and skin [2].

It is a rare condition with an estimated incidence of 5.4–8.9 per 1,000,000 but important for otolaryngologists since a reported 69–73% of patients have head and neck manifestation with up to 50% of cases having a head and neck finding as their sole presenting symptom [3]. Just under 20% of patients with LCH have temporal bone involvement, most typically in patients with the multisystem form. Otorrhea is seen in 18% of patients with LCH and in 60% of patients with LCH of the temporal bone [4].

Cholesteatomas are epidermal inclusion cysts of the middle ear or mastoid that contain desquamated keratinizing debris. They are often a consequence of chronic otitis media and, left untreated, they can cause hearing loss, vertigo, facial nerve paralysis, and meningitis [5].

Here, we present a case report of a 6-year-old female with a history of treated Langerhans Cell Histiocytosis in remission who presented to our clinic four years later with bilateral cholesteatomas. We perform a review of the literature to search for similar cases and compare our case to the few other documented occurrences of cholesteatoma after treatment of Langerhans cell Histiocytosis of the temporal bone [6,7].

To test our hypothesis that lytic destruction of the temporal bone from LCH could create an environment that promotes cholesteatoma formation, we performed a cross-sectional database analysis to see whether patients with LCH had a higher than average incidence of cholesteatoma.

The Agency for Healthcare Research and Quality is a division of the United States Department of Health and Human Services which

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http://dx.doi.org/10.1016/j.ijporl.2017.06.022
0165-5876/

has been collecting deidentified inpatient patient data since 1988. The annual release of this data, the National Inpatient Sample (NIS), and the pediatric-only version, the Kids' inpatient database (KID) which is released every three years, provides a large sample of patients to study which is ideal for analyses of relatively rare conditions such as LCH. For these reasons, we queried the National Inpatient Sample (NIS) and Kids' inpatient database (KID) to evaluation the association between LCH and cholesteatoma.

2. Case report

A 6-year-old female with a history of multi-system Langerhans cell Histiocytosis that involved bilateral temporal bones status post treatment three years' prior with chemotherapy (vinblastine, 6-mp, prednisone) presented to our office with a 5-day history of right blood-tinged otorrhea. On exam, a polypoid mass of the posterior right bony external auditory canal (EAC) was seen. A CT temporal bone showed findings concerning for recurrence of Langerhans Cell Histiocytosis versus cholesteatoma so she was taken to the operating room for a biopsy of the mass; pathology was consistent with cholesteatoma. The patient underwent a canal wall up mastoidectomy, removing cholesteatoma from the antrum and reconstructing the posterior EAC with conchal cartilage. 1 year post-operatively with she had no ipsilateral hearing loss with grossly normal Pure Tone Average (PTA) and a Word Recognition Score (WRS) of 96%. However, at her one-year exam, the patient had dark otorrhea and a foreign body sensation in her left ear. Endoscopic office exam showed a posterior left external auditory canal defect communicating with the mastoid. MRI confirmed a left-sided mastoid cholesteatoma (Fig. 1).

She underwent a left sided canal wall up mastoidectomy, reconstructing the posterior EAC with conchal cartilage. Similar to the right side, her cholesteatoma only involved the mastoid antrum. She did well post-operatively with an unchanged audiogram and no signs of recurrent cholesteatoma at her 6-month follow up (Fig. 2).

3. Methods

3.1. Literature review

A systematic search in OVID medline was conducted. Relevant synonyms for the MESH terms “Langerhans Cell Histiocytosis” and “Cholesteatoma” were used to compile an initial list of articles. Relevant articles were isolated after reviewing abstracts. Only studies that reported development of cholesteatoma after a diagnosis of LCH were evaluated. Related publications that were not identified by the initial literature search were found through Google Scholar. Selected articles and related reviews were hand searched for relevant cross-references.

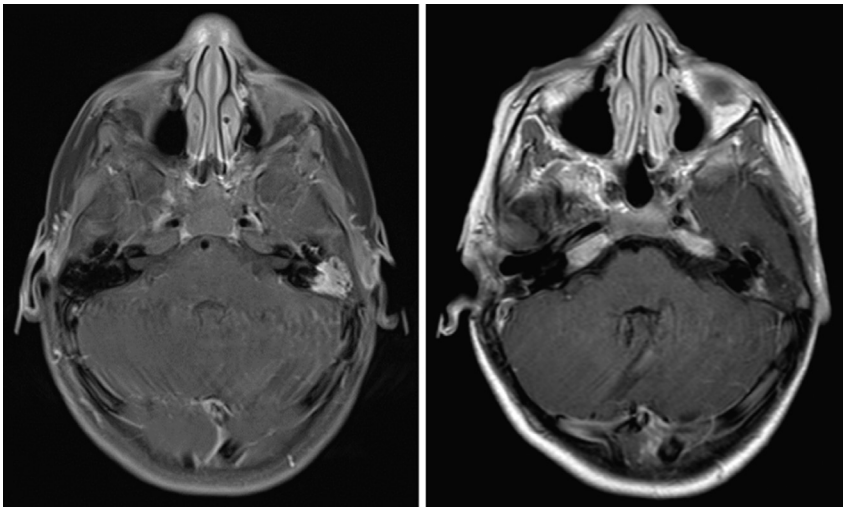

Fig. 1. (Left) MRI T1 post contrast axial cut of our patient at her initial diagnosis of LCH. Note the hyperintesnity in her left temporal bone. Both cholesteatoma and LCH are iso/hyperintense on T2 and hypointense on T1.8 However, only LCH enhances with contrast. (Right) MRI T1 post contrast axial cut of our patient 10 months after her right-sided mastoidectomy: cholesteatoma is now seen as the hypointense mass in the left EAC.

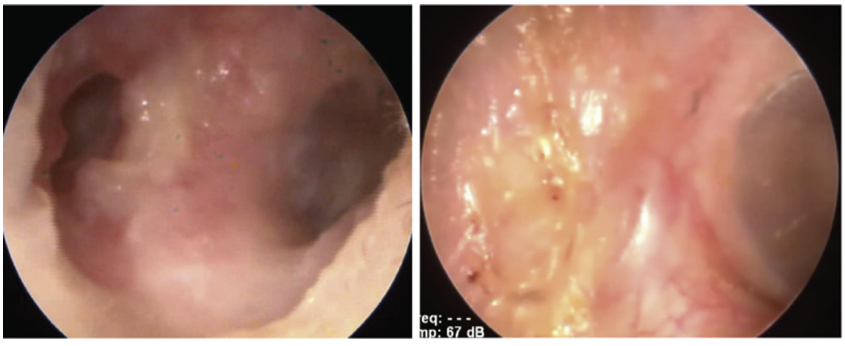

Fig. 2. (left): Cholesteatoma formation in the defect in the posterior EAC of the right ear. (Right) The same ear three month after a canal wall up mastoidectomy and EAC reconstruction.