Langerhan Cell Histiocytosis of the Temporal Bone with Otic Capsule Involvement: A Case Series

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ABSTRACT

Langerhan cell histiocytosis (LCH) is a rare disease that can affect any organ system in the body. Bony lesions are common within the temporal bone and are involved in 15 – 41% of cases. In fact, the ear is often the presenting symptom. Nevertheless, lesions involving the otic capsule are exceedingly rare with only 1.5 reported cases in the literature. Here, we report a case series of eight patients with LCH of the mastoid, three of which involve the otic capsule.

With IRB approval, the cases of collaborating otolaryngologists in conjunction with our institutional radiographic and pathologic databases were searched for examples of LCH involving the temporal bone. Eight cases of LCH of the temporal bone with three demonstrating otic capsule involvement, both clinically and radiographically were identified. Review of post-treatment imaging revealed all three patients had restoration of the bony labyrinthine architecture. Furthermore, all of the patients for which an audiogram could be obtained demonstrated near or complete restoration of their hearing.

Though LCH of the temporal bone is a common site within the spectrum of the disease, involvement of the otic capsule remains rare. Here, we report, to our knowledge, the largest series of otic capsule involvement by LCH and demonstrate both architecture and hearing is recovered with appropriate treatment. Lastly, restoration of the bony architecture of the labyrinth suggests the radiographic changes seen with LCH may be caused by demineralization and not destruction.

INTRODUCTION

Langerhan cell histiocytosis (LCH) is a rare clonal proliferation of pathologic cells with characteristics of Langerhan cells (LCs). Physiologically, LCs are antigen presenting dendritic cells normally found in the epidermis and respiratory and genital epithelia. These cells have a unique ability to translocate from their origin tissue to lymph nodes to activate the immune system. In contrast, LCH is an abnormal proliferation of langerhan cells in a mixed stroma of varying degrees of macrophages, T lymphocytes, eosinophils and multinucleated giant cells (Fig 1A) (1). The proliferation of LCs in LCH is self-sustaining with pathologic LCs secreting inflammatory cytokines that both recruit and sustain new LCs. Of historical interest, in 1987 LCH was designated the name to encompass the following entities: histiocytosis X (previously used as an umbrella term described by Lichtenstein in 1938), Hand-Schuller-Christian, lipoid granulomatosis, Abt-Letterer-Siwe and eosinophilic granulomatosis (2).

LCH is a rare disease with an incidence of 8.9 cases per million per year (1). It has a slight male predominance, 1.7:1, and can affect any age group, though the peak incidence is in children ages one to four years old. The disease can affect any organ system either singularly or systemically. Bony lesions are common either alone or in combination with other organ systems, and, interestingly, LCH involves the temporal bone in 15 – 61% of cases (2). In fact, ear symptoms such as otitis media resistant to medical treatment, mastoid swelling, pre or post-auricular mass, polyps, periantricular edema or erosion of the posterior canal are the presenting symptoms in 5 – 25% of cases (3). The mastoid and squamous portion of the temporal bone are most often affected. The petrous portion is rarely involved and if so, the otic capsule is often left unscathed (4). As a result, conductive hearing loss is common but a sensorineural component has only been reported in approximately 15 cases.

Here we report our institutional findings of eight patients with LCH involving the mastoid, three of which demonstrate radiographic and clinical evidence of otic capsule involvement. Of those three, two demonstrated mixed or SNHL and one presented with conductive hearing loss. After treatment for LCH all three cases demonstrated hearing improvement and restoration of the bony architecture of the otic capsule.

RESULTS

Eight patients with LCH of the mastoid either as unilocal, multifocal or recurrent disease were identified. Table 1 illustrates the important characteristics of each subject. Specifically, three of the eight subjects had radiographic and/or clinical evidence of otic capsule involvement. Most illustrative of this was Subject 63 who demonstrated mixed SNHL at presentation and subsequently fully regained his hearing post treatment. Figure 2 (A – C) demonstrates pre-treatment CT scans (bone window) and audiograms pre and post-treatment demonstrating restoration of bony architecture and function.

Table 1: Characteristics of the Study Subjects

<table>
<thead>
<tr>
<th>Subject</th>
<th>Age at Dx</th>
<th>Sex</th>
<th>Presenting Symptom</th>
<th>HL</th>
<th>Primary Location</th>
<th>Treatment</th>
<th>Inner Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3</td>
<td>M</td>
<td>R ear pain</td>
<td>CHL</td>
<td>R mastoid/mastoid</td>
<td>S, V&amp;P</td>
<td>Yes</td>
</tr>
<tr>
<td>2</td>
<td>2.5</td>
<td>M</td>
<td>R ear drainage, Partial DI</td>
<td>Mixed</td>
<td>R Tbone</td>
<td>S, V&amp;P</td>
<td>Yes</td>
</tr>
<tr>
<td>3</td>
<td>5</td>
<td>M</td>
<td>HL</td>
<td>SNHL</td>
<td>B/L mastoids</td>
<td>S, V&amp;P</td>
<td>Yes</td>
</tr>
<tr>
<td>4</td>
<td>U</td>
<td>M</td>
<td>pituitary, diffuse/recurrent</td>
<td>NA</td>
<td>R mastoid</td>
<td>unknown</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>M</td>
<td>L ear pain/drainage</td>
<td>NA</td>
<td>L Tbone and calvarium - recurrent</td>
<td>S, V&amp;P</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>3</td>
<td>M</td>
<td>Chronic R ear drainage</td>
<td>NA</td>
<td>R mastoid/occiput</td>
<td>S, Chemo</td>
<td>No</td>
</tr>
<tr>
<td>7</td>
<td>4</td>
<td>F</td>
<td>R ear pain</td>
<td>NA</td>
<td>R calvarium and mastoid</td>
<td>S, V&amp;P</td>
<td>No</td>
</tr>
<tr>
<td>8</td>
<td>5.5</td>
<td>M</td>
<td>Chronic L ear infection</td>
<td>NA</td>
<td>L mastoid/EAC</td>
<td>S, V&amp;P</td>
<td>No</td>
</tr>
</tbody>
</table>

DISCUSSION

Langerhan cell histiocytosis is a complex and rare disease that can involve any organ system. Specifically, the temporal bone is involved in 19 – 27% of cases with hearing loss present in 7.4 – 28.5% of cases with 10% showing SNHL (5). Diagnosis is histologic identification of characteristic cell types and positive staining for CD1a (Fig 1B) and S100. For treatment, per the 2009 recommendations of the Histioctyosis Society, ear/temporal bone lesions are considered “CNS high risk” lesions and require systemic treatment in addition to surgery (4). Though the above statistics indicate the commonality of temporal bone disease with LCH, erosion of the otic capsule is quite rare. One study by Naduri et al reviewed all patients with LCH from their institution from 1965 – 1998. Fifty-eight of 275 cases involved the ear. Eighteen of those had mastoid involvement, but only one affected the inner ear (2). From this it appears the otic capsule, due to its high density, is resistant to erosion by the granulation tissue of LCH. Furthermore, it appears the membranous labyrinth often remains intact as demonstrated by Lopez-Rios in 1968 that upon autopsy of a 23 month old with extensive multi-system LCH with otic capsule intact had an intact membranous labyrinth (6). Our study reflects these findings, as demonstrated by subjects one through three. Resorption of osteocytic lesions in LCH after treatment has been well documented in the literature often occurring as soon as six months after starting treatment (2,5). The exact mechanism of bony erosion in LCH is poorly understood but recent success in treating LCH associated bone pain and lytic lesions (modestly in recurrent disease) with bisphosphonates (BPs) has elucidated parallels between osteoclasts and pathologic langerhan cell activity. Recent studies have shown nitrogen containing BPs work by inhibiting a key site in the mevalonate pathway involved in cholesterol synthesis in osteoclasts disrupting cell function and directing it towards apoptosis. BPs have an anti-inflammatory effect further decreasing osteoclast activity (7,8). Bisphosphonates are presumed effective in LCH because pathologic langerhan cells have elements of the mevalonate pathway and are exceedingly sensitive to inflammatory cytokines that propagate the disease (8). Thus, pathologic langerhan cells in LCH produce lytic lesions similarly to osteoclasts, through demineralization. With elimination of the offending langerhan cells the remaining healthy bone remineralizes the lesions and restores function.

CONCLUSIONS

Langerhan Cell Histiocytosis of the temporal bone is a locally destructive entity that can cause significant morbidity. Though rare, it should be suspected in patients as described above and, if diagnosed, should be treated in close conjunction with hematologists/oncologists. However, the severity of the disease suggested by pre-treatment imaging and audiograms does not necessarily predict functional outcome.

METHODS AND MATERIALS

After obtaining IRB approval, the radiographic, pathologic and clinical records of patients treated at Yale New Haven Hospital for LCH of the temporal bone were reviewed. Records of patients with a pathologic diagnosis of LCH were then scrutinized to identify the location of the lesions and characteristics of their disease process. Those not including the mastoid as a uni- or multifocal lesion were excluded. Pre and post-treatment scans were reviewed with a neuroradiologist to determine radiographic involvement of the otic capsule and resolution of identified lesions.

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REFERENCES