CASE REPORT

Multifocal Langerhans' Cell Histiocytosis Involving Bilateral Temporal Bones, Lungs, and Hypothalamus in an Adult

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ABSTRACT—Multifocal Langerhans' cell histiocytosis is a rare condition in adults. We present the case of a 31-year-old female who initially presented with a clinical appearance of acute mastoiditis. The patient ultimately had involvement of bilateral temporal bones, lungs, and hypothalamus. Treatment with methotrexate, steroids, and desmopressin acetate (DDAVP) resulted in initial clinical improvement though not resolution. The patient also underwent radiation therapy to the hypothalamic lesion. The literature is reviewed, focusing on the diagnostic challenge of this disease process in adults. The additional morbidity of surgery, specifically mastoidectomy, in the setting of Langerhan's cell histiocytosis is discussed.

Histiocytosis X is a term coined by Lichtenstein in 1953 to describe a group of disorders characterized by a proliferation of histiocytes. Descriptions of the disease date back to 1893 when Hand published a case report of a 3-year-old boy with exophthalmos, polyuria, and a skull lesion.¹ He later felt, in comparing this case with several other cases in the literature (also characterized by membranous bony defects, diabetes insipidus, and exophthalmos), that these cases represented a new syndrome.² Letterer, in 1924, presented a case of a 6-month-old boy with a history of fever, suppurative otitis media, generalized purpura, diffuse lymphadenopathy, hepatosplenomegaly, and purulent postauricular drainage who died after a 10-week illness. Siwe, in 1933, described a similar case and reviewed five other cases from the literature suggesting they represented the same disease.³ Finally, eosinophilic granuloma of the bone was first described in 1940 by Lichtenstein and Jaffe.⁴ It was not until 1953 and Lichtenstein, however, that

eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease were grouped together as part of the same spectrum of diseases.⁵

This unified classification, however, is controversial as these disease entities have different clinical courses and prognoses. Letterer-Siwe disease is characterized by disseminated organ involvement and a rapidly progressive clinical course that is usually fatal. It is generally seen only in children less than 2 to 3 years of age. Hand-Schuller-Christian disease is also characterized by disseminated disease involving multiple bony lesions or involving other organ systems, but its clinical course is much more benign. It typically affects children between the ages of 1 and 5, but can present in young adulthood. The classic triad of diabetes insipidus, exophthalmos, and osteolytic skull lesions only occurs in 10% of patients. Eosinophilic granuloma is the most benign form and may involve single or multiple bony lesions.⁶⁻⁸

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Lieberman et al⁹ proposed in 1969 that the term histiocytosis X be eliminated entirely and the disease entities reclassified as unifocal eosinophilic granuloma, multifocal eosinophilic granuloma, and a type of lymphomatous disorder. Other classification systems have also been suggested in the attempt to better understand and appropriately define these disorders. The term histiocytosis X, however, remained widely accepted until 1987 when the Histiocyte Society proposed discarding this term and its associated eponyms and reclassifying these disorders as Langerhans' cell histiocytosis (LCH).¹⁰

Pathologically, LCH is characterized by the proliferation of atypical Langerhans' cells that form granulomas in conjunction with lymphocytes, polymorphonuclear cells, and eosinophils. The sine qua non of the Langerhans' cell are penitilaminar cytoplasmic inclusion bodies known as Birbeck granules by ultrastructural analysis.¹¹ Diagnostic criteria based on pathological examination have been defined by the Histiocyte Society.¹⁰ A "definitive diagnosis" requires electron microscopic identification of Birbeck granules, or demonstration of CD1 antigenic determinants on lesional cells by immunohistochemistry, both of which require fresh not formalin-fixed tissue. The diagnosis is considered "presumptive" if it is based on histologic features only. The level of diagnostic confidence is increased, however, if two or more of the following are seen: positive immunostain for adenosine triphosphatase (ATPase), S-100 protein, α -D-mannosidase, or characteristic binding of peanut lectin.

Clinically, LCH can involve several organ systems, but most frequently manifests as osseous lesions, characteristically involving the flat bones of the skull, ribs, pelvis, and scapula. Lesions are typically sharply marginated lucencies frequently with scalloped edges on plane films. Other areas that can be involved in this disorder include skin, oral mucosa, lymph nodes, lungs, gastrointestinal tract (notably liver and spleen), and hematologic involvement. Neuroendocrine involvement can include diabetes insipidus, panhypopituitarism, and hypogonadism.⁶⁻⁸

Overall head and neck manifestations are seen with an incidence as high as 62 to 90%¹²⁻¹⁶ and are the presenting symptoms in 34 to 40% of these cases.¹⁴ Temporal bone involvement occurs in 8 to 61% of cases depending on the series,^{12,14-29} with bilateral involvement in 31% of the cases.¹⁶ Otologic symptoms were the initial presentation in 25% of these cases, specifically otorrhea, granulation tissue/polyps in the external auditory canal (EAC), fistula in the posterior EAC, postauricular soft tissue swelling, hearing loss, otitis externa, vestibular manifestations, and facial nerve palsy.^{16,20,26,30} Otologic involvement can present as otitis externa, otitis media, or even mastoiditis.

We present a case of LCH in an adult with involvement of multiple organ systems who initially presented with a clinical picture of acute mastoiditis.

The patient is a 31-year-old white female with a past medical history notable only for hypothyroidism diagnosed 2 years prior who initially presented 13 months ago to another institution with complaints of right otalgia and vertigo. Computed tomography (CT) findings were consistent with osteomyelitis of the right mastoid for which the patient underwent a right mastoidectomy during which the vertical portion of the facial nerve was injured. The facial nerve was intraoperatively decompressed and primarily reanastomosed.

The day following the initial surgery, the patient was referred to our institution with a complete peripheral facial nerve paralysis for revision mastoidectomy and exploration of the facial nerve repair. Reexploration was notable for extensive granulation tissue, an intact facial nerve reanastomosis, and a fistula of the posterior semicircular canal (Fig. 1), which was then patched with fascia. Pathology at that time revealed inflamed granulation tissue. Postoperatively, the patient underwent right tarsorrhaphy and was treated with antibiotics.

Over the following 5 months, facial nerve function returned to the present House-Brackman grade III and the symptoms of dizziness gradually resolved. However, 7 months after her original surgery, the patient presented with conductive hearing loss in the opposite, left ear secondary to serous otitis media and was treated with antibiotics and steroids. Within 5 days, her left otalgia worsened and she developed postauricular soft tissue swelling and erythema along with external auditory canal edema. Left myringotomy was performed and

Figure 1. Axial CT scan, right ear. Note the mastoidectomy defect and the fistula at the posterior semicircular canal. There is a dehiscence of the posterior wall of the facial nerve recess.

serous fluid obtained, which was sent for culture. Patient was started on intravenous (I.V.) Rocephin and CT scan obtained, which revealed opacification of the left mastoid with coalescence and bony destruction of the lateral aspect of the mastoid bone and sigmoid plate with thinning of the tegmen tympani and fluid filling the middle ear (Figs. 2 and 3). Two days later, the patient underwent a left complete mastoidectomy and placement of a tympanostomy tube with findings of extensive granulation tissue as well as dehiscences of the mastoid cortex, area of the sigmoid plate with no evidence of sigmoid thrombosis, and the tegmen of the antrum with granulation tissue adherent to the dura. Intraoperative cultures for bacteria and fungus were negative. Routine microscopic examination as well as immunohistochemistry was consistent with the diagnosis of LCH (Figs. 4 and 5). This prompted review of the pathology from the previous right revision mastoidectomy 8 months prior. Microscopic examination revealed a fibroinflammatory reaction with rare histiocytes positive for S100 immunohistochemical staining, consistent with, but not definitive for LCH.

Further questioning revealed a 2-year history of amenorrhea, polydipsia, and polyuria in addition to her known symptoms of hypothyroidism. Further radiographic evaluation included skull films and skeletal survey, which were negative. Chest Xray (CXR) and chest CT, however, revealed asymmetric cystic and interstitial lesions in the upper and mid lung fields bilaterally consistent with eosinophilic granuloma (Fig. 6A and 6B). Magnetic resonance imaging (MRI) revealed a suprasellar mass within the hypothalamus and optic

Figure 2. Axial CT scan, left ear. There are lytic changes in the mastoid, with dehiscence of the sigmoid plate and overlying mastoid cortex.

Figure 3. Coronal CT scan, left ear. Note dehiscence of mastoid tegmen tympani.

tract (Fig. 7). Laboratory studies revealed low gonadotropins and mildly elevated prolactin consistent with panhypopituitarism. Her thyroid-stimulating hormone (TSH) and serum sodium levels were normal. Urine osmolality and specific gravity, while somewhat, dilute were within normal limits. Patient was started on estrogen replacement with Premarin and Provera. Her diabetes insipidus symptoms were treated with desmopressin acetate (DDAVP) nasal spray with significant improvement. Therapy with Methotrexate and Hydrocortisone was initiated for treatment of multifocal LCH for a period of 6 months. Patient initially did well but follow-up MRI 6 months later revealed increase in size of the hypothalamic lesion. The patient subsequently underwent radiation treatment to the hypothalamic lesion (10 Gray) with a good response. She continues to do well with the exception of continued weight gain felt

Figure 4. Histiocytes showing diffuse positive staining with S-100 protein (arrow). (magnification $\times 200$)

Figure 5. Electron photomicrograph of a Langerhans histiocyte showing the nucleus (n), endoplasmic reticulum (er), and numerous Birbeck granules (b). (magnification $\times 18,800$.) Inset: Birbeck granules. The presence of Birbeck granules differentiates LCH from other histiocyte disorders. (magnification $\times 68,000$)

to be secondary to her hypothalamic involvement. Her other systemic symptoms are well-controlled.

DISCUSSION

LCH, while extensively reported in the pediatric population, is rare in adults. The incidence of LCH in adults in the literature, especially with multisystem disease, is difficult to ascertain, as case reports are few^{17,31-37} and adults have frequently been included in predominantly pediatric series rather than being defined as a separate group.^{13,19-21,25,26,29,38-41} The literature on LCH, therefore, generally reflects what is known about the disease in the pediatric population.^{11,39,42-51}

Adults comprised 1 to 4% of the patients diagnosed with LCH in the reviewed pediatric series that distinguished the adult from the pediatric patients.^{23,52} Wester et al⁵³ reported the only adult series found on review of the literature in 1982. Adult patients were defined as being 16 years old or older. All 61 patients had bony involvement with rib, skull, and pelvis the most frequent sites. Forty-six patients had solitary skeletal lesions and the remainder had multiple bony lesions. In addition,

Figure 6. (A) (B) Axial CT scan, chest; lung windows. Shows upper lobe cystic changes compatible with LCH. The lower lobes are less involved, typical for LCH.

Figure 7. Coronal postgadolinium T1-weighted image showing an enhancing hypothalamic and infundibular mass.

eight patients also had extraskelatal disease which consisted of pulmonary (5%), mucocutaneous, or hypothalamic-pituitary involvement (5%). Only one of these patients had pulmonary and hypothalamic-pituitary axis involvement. However, all three patients with hypothalamic-pituitary axis involvement had another site of extraskelatal disease. The authors note that approximately 75% of their patients had solitary lesions compared to the quoted 25 to 60% of patients in the pediatric literature,^{14,20,24} indicating that multifocal disease is much less common in an adult population. Importantly, they also note that age of presentation does not appear to influence or predict the course or severity of the disease, as has been noted in the pediatric population. Rather, the presence of extraskelatal disease is the only predictor of a more protracted course of the disease process in the adult population.⁵³

Overall LCH most frequently manifests as osseous lesions, characteristically involving the flat bones of the skull, ribs, pelvis, and scapula. Involvement of the temporal bone occurs in 8 to 61% of cases,^{12,14-29} depending on the series and is frequently bilateral. Otologic symptoms are the initial presentation in 4 to 25% of these cases.^{16,20,26,30} Despite the extensive bony destruction of the temporal bone characteristic of this disease, facial nerve palsy as a result of the disease process is rare occurring in less than 2.8% of patients.⁵⁴ The facial nerve appears resistant to invasion and facial nerve palsy is thought to result from compromise of the blood supply to the nerve from destruction of the osseous canal. The cochlea and labyrinth are also generally resistant to invasion.

Temporal bone surgery in the setting of LCH carries a high rate of morbidity, which includes postaural

fistula, sensorineural and conductive hearing loss, and facial nerve injury. Facial nerve palsy has been reported as a postoperative complication of mastoidectomy in LCH in several series with an incidence as high as 12%.^{14,54} Tos stated in reference to his series, "The risk of operative damage to the facial nerve is considerable because of the completely imperspicuous approach. . . ." Irving et al¹⁴ also noted that the morbidity of mastoid surgery in the setting of LCH is high. In their series, 25 out of 131 children had temporal bone involvement and 11 mastoidectomies were performed on 8 children. Complications included one transient facial nerve paralysis, two cases of sensorineural hearing loss, two postaural fistulas, and one case of bilateral conductive hearing loss. Moreover, the disease remained active in all but one of these patients and required further treatment. Temporal bone surgery was felt to have an unacceptably high rate of complication in the setting of LCH and more conservative treatment was recommended. Irving states "destruction of the mastoid by LCH can place the facial nerve at risk owing to the loss of normal anatomic landmarks. . . ." Levy et al⁵⁵ reported a similar case of transient postoperative facial palsy where the facial nerve was lying free in the auditory cavity with no bony covering at operation "resembling a piece of string. . . ." Facial nerve transection has also been reported as a complication of transcanal biopsy of granulation tissue in a patient with LCH.²⁰

Other organ systems involved in LCH include skin, mucosa, lymph nodes, lungs, liver, spleen, and bone marrow involvement. Neuroendocrine involvement includes diabetes insipidus, panhypopituitarism, and hypogonadism. The most frequent neuroendocrinopathy is diabetes insipidus, which has incidence quoted as high as 25 to 50% with half of these cases actually manifest at initial presentation.⁶⁻⁸ Pulmonary involvement in LCH is uncommon with an incidence of 5 to 10% in patients with multifocal disease.^{39,53} Pulmonary involvement carries a high morbidity due to progressive fibrosis. Chest radiograph frequently shows a reticulonodular pattern with sparing of the costophrenic angles. Findings on high-resolution CT include thin-walled cysts ("honeycombing") usually <1 cm in size or nodules of <5 mm in diameter. There is a characteristic upper lobe predominance of these lesions with sparing of the costophrenic angles.⁵⁶

Treatment of LCH varies depending on the extent of involvement. Patients with isolated osseous lesions have been treated with observation, curettage, excision, and radiation therapy. Patients with multiple osseous lesions may also be treated with chemotherapeutic agents. Radiation therapy is generally utilized for painful lesions or those that are progressive or difficult to access surgically. Patients with isolated lesions generally have an excellent prognosis with recovery rates of 80 to 100%.^{18,23,52} Recurrences overall are rare, but patients can later develop multifocal disease.

Patients with multifocal disease, however, have a more protracted course of disease with high morbidity. Patients with multifocal LCH with onset after the age of two frequently suffer chronic or recurrent osseous or visceral lesions, but rarely die of their disease. This is in contrast to patients with acute disseminated LCH (Letterer-Siwe disease), where prolonged survival is unusual despite aggressive therapy. In these patients with a more chronic course of multifocal LCH, systemic therapy is felt to be essential to prevent prolonging the disease and to lessen the associated morbidity. Treatment generally consists of methotrexate, prednisone, and occasionally other chemotherapeutic agents such as vincristine, vinblastine, or cyclophosphamide.^{1,11,14,23,27,39,57} Patients with symptoms of diabetes insipidus or panhypopituitarism also require treatment with vasopressin and hormonal therapy, respectively. However, despite therapy the majority of these patients will have persistent disease with a significant associated morbidity.⁵²

CONCLUSIONS

This case of multifocal LCH involving bilateral temporal bones, lungs, and hypothalamus highlights the difficulty in diagnosis of LCH in adults. The rarity of this lesion in adults can result in delayed diagnosis and treatment of the disease as an infectious process. The surgical treatment of acute mastoiditis carries additional morbidity in the setting of LCH, making delay in diagnosis potentially hazardous. The extensive lytic bony destruction typical of LCH renders structures including the facial nerve, tegmen, sigmoid sinus, and less commonly the labyrinth, vulnerable to injury and caution should be exercised during surgery and the facial nerve monitor utilized. When performing mastoidectomy for acute mastoiditis, tissue should not only be sent for culture but also for pathology. A detailed history is essential to elicit the presence of systemic symptoms and multifocal disease. LCH should be considered in any immunocompetent adult with an atypical presentation of mastoiditis.

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