Case Report

Langerhans cell histiocytosis involving bilateral temporal bones in infant patient: a case report

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Abstract

Langerhans' cell histiocytosis (LCH) is a rare pediatric disease of unknown etiology. The mastoid and skull base are common sites of involvement in LDH. We present a case of bilateral langerhans histiocytosis in 9-month-old infant presented with bilateral post auricular swelling for 2 month mimic the picture of acute mastoiditis. Patient diagnosed after CT scan and open biopsy from mastoid cavity.

KEY WORDS: Langerhans cell histiocytosis, temporal bone, children

Introduction

Langerhans cell histiocytosis (formerly called histiocytosis X by Lichtenstein in 1953) refers to a group of disorders that are characterized by proliferation of cytologically benign histiocytes.[1] Langerhans' cell histiocytosis (LCH) is a rare pediatric disease of unknown etiology affecting 1-5 children per 1 million each year.[2] The clinical spectrum of disease is quite varied, ranging from a solitary eosinophilic granuloma to diffuse multisystem involvement. The head and neck is the most common site of involvement, occurring in approximately 60% LCH patients. Head and neck manifestations are diverse and include skull and temporal bone lesions, cervical lymphadenopathy, and skin rash. Diagnosis can be difficult as these lesions mimic other common conditions seen by the otolaryngologist, including otitis externa, acute mastoiditis, and gingivitis. In this study, infant with bilateral temporal bone LCH who was misdiagnosed as otitis media with mastioditis.

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Case Report

In March 2015, a nine months Saudi girl, presented to the outpatient clinic in Armed Forces Hospital, Southern Region in Khamis Mushayt, Saudi Arabia, with history of bilateral post auricular swellings and protruding of left auricle. Her complaint started before 3 months of presentation with periauricular edema and swelling of right ear followed by protrusion of pinna.

Patient seen by general physician and stared her on antibiotic and analgesia as case of otitis media, patient improved regarding pain and edema subsided but she developed lump over the mastoid in right side. After 4 weeks she developed the same problem in left ear but more severe than that of right side. Patient started to have severe pain and periauricular edema with significant protrusion of pinna associated with low grade fever. Parents were observing her for 2 months waiting for spontaneous remission.

Patient came to our clinic with left ear protrusion, severe tenderness, palpable lump over the mastoid of right ear, with history of bilateral ear discharge. There were no history of facial palsy, convulsions, or vomiting.

Clinical examination show sagging of posterior and superior walls of external auditory canal in left side with significant postauricular swelling and palpable lump on right mastoid. Facial nerve intact and normal function for other cranial nerves. Multiple palpable cervical lymph nodes in both sides of the neck. Normal eye movement is found without exophthalmos.

So, patient admitted as case of acute mastoiditis and started on intravenous antibiotics. CT scanning of temporal bone done, revealed ill-defined soft tissue lesion in left mastoid and

filling the middle ear cavity with extensive destruction of mastoid septations, left squamous temporal bone, lateral mastoid wall extended to subcutaneous tissues of post auricular region with small erosion of lateral semicircular canal. While in right side there was erosion in the lateral part of temporal bone and features of oto-mastoiditis. Multiple significant lymph nodes in the neck bilaterally [Figures 1-4].

Auditory assessment done for the patient inform of auditory brain stem response (ABR) which revealed normal hearing in right ear and moderately severe hearing loss in left ear. Decision made to take the patient to theatre for exploration and biopsy under general anesthesia for left ear with postauricular incision. We found soft tissue lesion filling the mastoid region and erosion of lateral wall and roof of mastoid, exposing the dura [Figure 5]. Biopsy taken and sent for histopathology result came as Langerhans cell histocytosis [Figure 6]. Patient referred to pediatric oncology team, no other lesions identified. They started her on corticosteroid and chemotherapy and initially she responds to treatment.

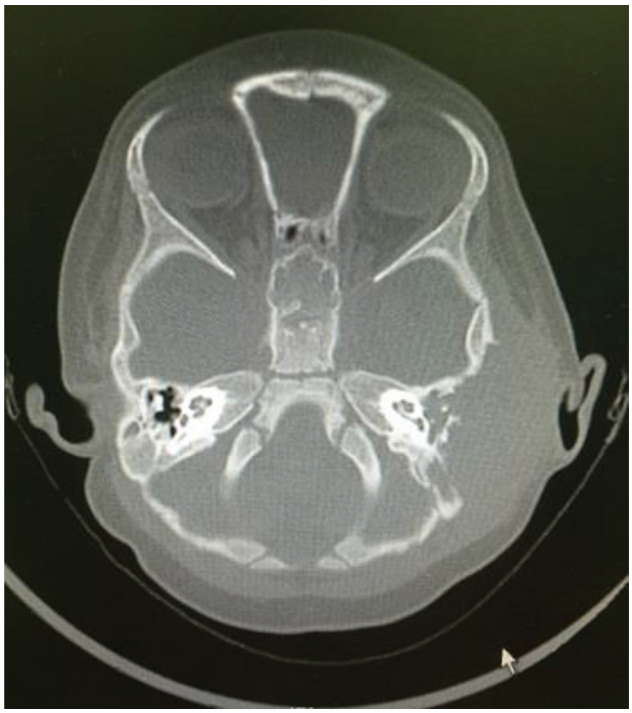

Figure 1: Axial ct scan of temporal bones show ill-defined soft tissue lesions involving both temporal bones.

Discussion

Langerhans histiocytosis (previously called histiocytosis X) describes a group of idiopathic disease caused by the abnormal proliferation of histiocytes. Histiocytes, normally benign cells found in the dermis or epidermis, accumulate in the skin, bone, lymph nodes, and visceral organs. This group is comprised of three diseases, eosinophilic granuloma, Hand-Schillller-Christian disease, and Letterer-Siwe disease, which describe progressively more aggressive and widespread manifestations of the same underlying pathology.[3]

Patients with eosinophilic granuloma develop solitary osteolytic lesions without systemic manifestations. The course

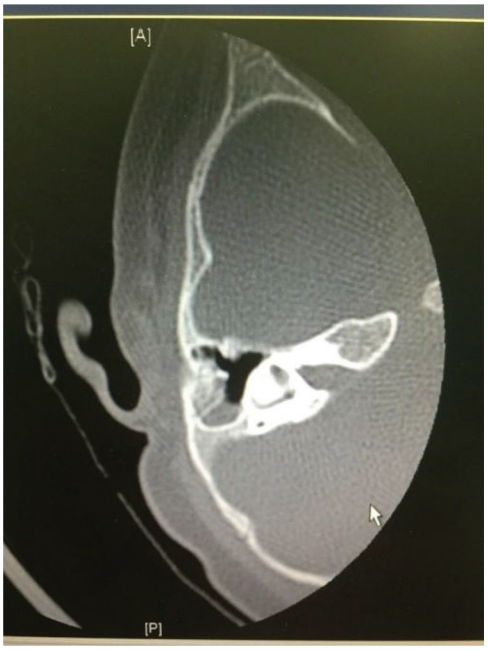

Figure 2: High resolution CT scan of right temporal bone show oto-mastoiditis.

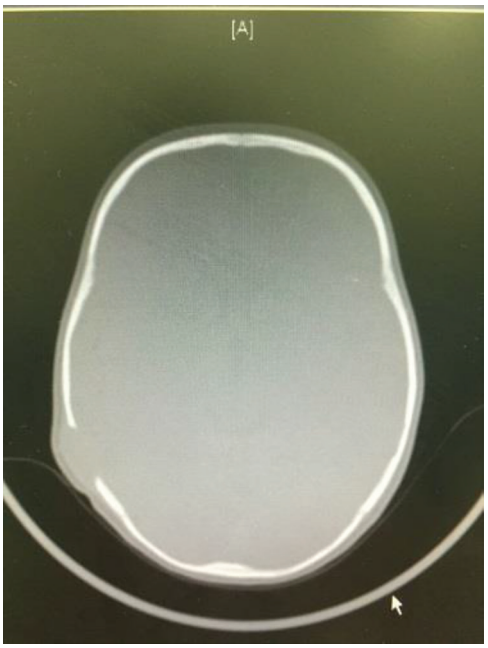

Figure 3: Erosion of right temporal bone and subgaleal soft tissue mass.

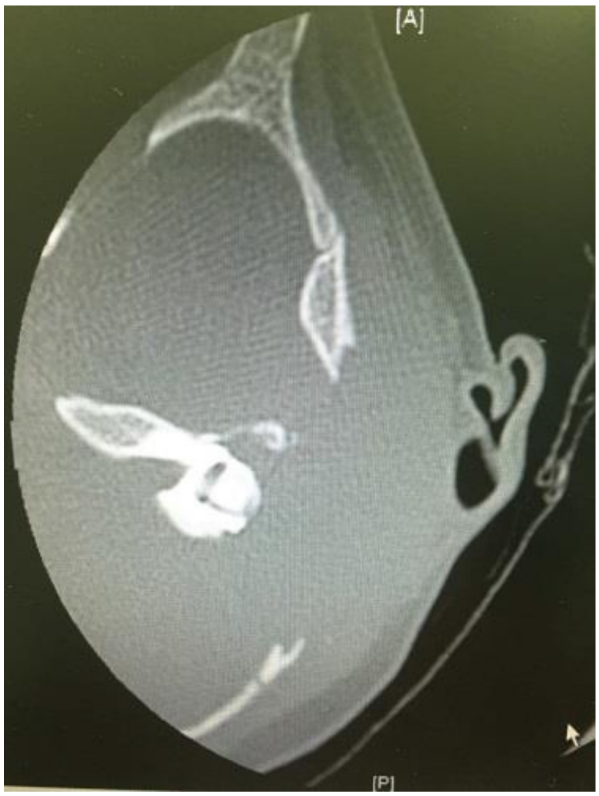

Figure 4: High-resolusion CT scan of left temporal bone show erosion of lateral semicurculaer canal.

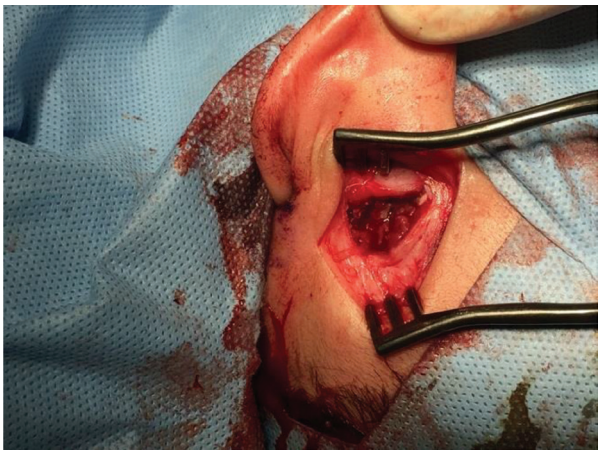

Figure 5: Soft tissue mass in left mastoid with erosion of all mastoid bone and exposure of dura.

of this disease is typically benign and patients often undergo spontaneous regression of their lesions. For those who do not, local excision of the granuloma or intralesional steroids can be performed. Hand-Schiiller-Christian disease typically affects patients less than 5 years old, who develop multifocal osteolytic lesions with rare extraskeletal involvement up to 25% patients present with the triad of osteolytic skull lesions,

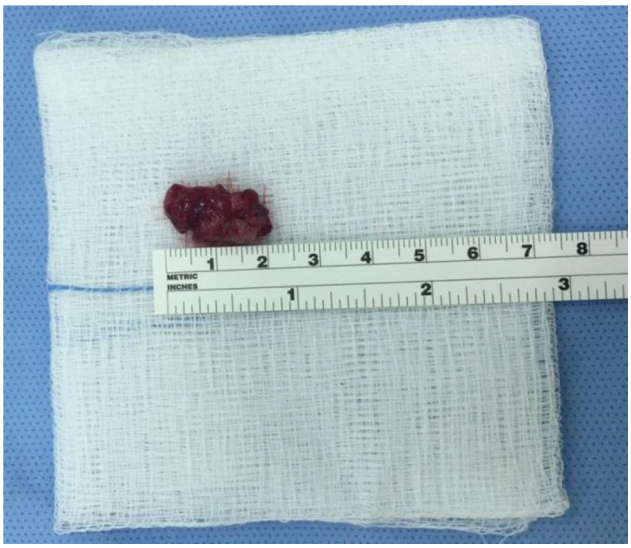

Figure 6: Mass taken for biopsy.

exophthalmos, and diabetes insipidus.[4]

Letter-Siwe is the most aggressive of this group, presenting most commonly in children less than 3 years old, who develop disseminated disease with diffuse involvement of multiple organs. Prognosis is poor with most patients succumbing to the disease in childhood.[5] Lesional biopsy for each of these diseases demonstrates characteristic tennis racket-shaped Birbeck granule on histology, the result of cytoplasmic inclusions bodies. In addition, diagnosis can be made by demonstrating the presence of the CD1 antigen with immunohistochemistry.

The otologic manifestations of these diseases result from the development of a granuloma within the temporal bone.[6] As the granuloma expands, it can cause conductive hearing loss, otorrhea, facial nerve paralysis, vertigo, and SNHL depending on which anatomic structures the granuloma violates.[7] The majority of patients suffering from this disorder first present (63%) or later develop head and neck manifestations.[8] The skull is frequently involved (42%); the temporal bone can be affected either as a solitary lesion or as part of a multisystem involvement in up to 61% patients.[9] When the temporal bone is involved, 30% of affected patients demonstrate bilateral disease.[10] Manifestations of LCH may be confused with more common disorders such as chronic suppurative otitis media and acute mastoiditis, which are not responsive to medical therapy,[11] thereby delaying the diagnosis, as observed in our case.

The cases presented demonstrate the classic postauricular swelling, which is seen in 10-30% of all patients with LCH. The ear often protrudes anteriorly, since the mass is subgaleal and lifts the entire auricle away from the temporal bone. There is frequently erosion of the bony posterior external auditory canal with sagging of the canal wall skin in such a way that the visualization of the tympanic membrane is difficult. Interestingly, although the facial nerve is exposed in the middle ear in 50% of all individuals, there have been only 14 reported cases of facial nerve paralysis in patients with LCH in the English literature.[12] Other areas of erosion include the tegmen tympani,

the bony plate covering the sigmoid sinus, the squama of the temporal bone and rarely the vestibular labyrinth. The mastoid air cell system is usually destroyed when this disease involves the temporal bone. Palpation of the mass reveals soft pseudofluctuation with associated cutaneous erythema but no tenderness. Because of the propensity of this condition to erode the posterior canal wall, a secondary external otitis and stenosis may develop. Perforation of the tympanic membrane is uncommon and vestibular abnormalities are rare.[11] Exophthalmos results from lesions involving the orbital walls and the surrounding soft tissue reaction. The skin is a common site of involvement in more advanced LCH and the scalp is particularly apparent in this condition. A seborrheic rash with almost dandruff-like condition can result, although much more profound greasy, weeping dermatitis is also seen at times.[9] The flat bones of the skull, ribs, pelvis, and scapula are most commonly involved. In the cranium, the frontal areas are most commonly involved and lesions may be single or multiple. The most commonly involved sites are the mandibles, especially posteriorly and along the alveolar ridges. The oral cavity can also be the site of mucosal ulcerations and stomatitis, which histologically reveal the typical histiocytic and eosinophilic infiltration. The dermatic lesions due to seborrheic dermatitis and the recurrent ear infections will raise the suspicion of LCH, whereas anemia, splenomegaly, hepatomegaly, and the transaminase increase indicate systemic manifestation of the disease.

The incidence of LCH is estimated to be three cases per 1,000,000 children per year and more than 50% of all cases are seen in patients under age of 10 years. Recent studies have shown that approximately 30% of LCH cases occur in adults, with an incidence rate of 1.8/1,000,000.[13] The average age of onset is 1-3 years, and the disease occurs more commonly in males.[14] The natural history of this disorder is interesting because it has the potential for spontaneous resolution when the child enters adulthood.[15]

For patients with systemic disease and a temporal bone lesion, systemic steroids are the first line of therapy with etoposide, vincristine, and vinblastine reserved for refractory patients. For patients with disease localized to the temporal bone, surgical debulking can be performed with topical and intralesional injection of steroids.

Radiation therapy has also been successful in treating temporal bone lesions refractory to resection,[16] Although radiation therapy has been a traditionally viable and even popular option, it is not recommended in the head and neck secondary to significant potential side effects and does not offer control in a primary or adjuvant fashion that is acceptable. In addition, there are reports of secondary malignancy resulting from radiation treatment.[17]

Conclusion

The role of otolaryngologist in LCH is limited to diagnostic biopsy and to the treatment of localized disease (curettage, aural polypectomy). In the case of multifocal LCH, as in our patient, an invasive surgical approach is not suitable and chemotherapy should be administered. A multidisciplinary approach to LCH patients and in particular of the otolaryngologist is required to recognize LCH early, and to plan the best management of such a delicate site of disease.

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