Langerhans Cells Histiocytosis (LCH) is a disease characterized by the proliferation of Langerhans cells causing local or systemic effects. Bone involvement is the most common presentation of this disease. We report three cases of LCH in the temporal bone of pediatric patients without systemic manifestations discussing its preoperative symptoms, radiological findings and treatment strategies. The median age of our patients was 3.3 years, two cases male and 1. The preoperative symptoms were chronic ear discharge in 1 case, retroauricular acute mastoiditis in 1 case and assymptomatic temporal bone erosion in 1 case. Audiological findings showed 1 case with normal results and 2 cases with conductive involvement of hearing. The mean time of the follow up was 42 months. All of our cases were submitted to surgical diagnosis with biopsy and further resection of the disease. Postoperative corticosteroid therapy was employed in order to reduce all the possible remaining histiocytes in the operative field.

INTRODUCTION

Langerhans Cells Histiocytosis (LCH) is the current term employed in literature to describe the spectrum of diseases characterized by the proliferation of diploic Langerhans cells causing site or systemic effects (1,2). The three classical presentation is eosinophilic granuloma, in which the lesion is limited to bone in patients between 15 years old and 35 years old (isolated case of eosinophilic granuloma) and the Langerhans disease (systemic compromise). Eosinophilic granuloma (EG) is the benign form of the 3 clinical variants of Langerhans cell histiocytosis. EG is characterized by a single or multiple skeletal lesions, and it predominantly affects children, adolescents, or young adults. Solitary lesions are more common than multiple lesions. When multiple lesions occur, the new lesions appear within 1-2 years. Any bone can be involved, but the most common sites include the skull, mandible, spine, ribs, and long bones. It is esteem that 15-61% from all the patients with LCH may have otologic involvement (4,5,6). The otologic involvement is more frequent in children with multisystemic diseases. Otologic symptoms without other systemic complications can be initial presentation of the disease in 5-25% of the cases. A mastoid lesion may extend into the middle and inner ear, destroy the ossicles, and lead to deafness. In this paper, we reported three cases of LCH of the temporal bone in pediatric patients: without systemic manifestations (eosinophilic granuloma) from 2001 to October of 2006.

OBJECTIVES

The purpose of this study was to present three consecutive pediatric cases of isolated LCH of the temporal bone (eosinophilic granuloma) and to review its clinical and radiological findings. We intend to present our treatment strategies that were employed in order to conduct the cases.

CASE REPORTS

Case 1

1. T.G.P.S., 11 years old, male, student, natural from Sao Paulo, Brazil, came to the ENT service of the Professor Edmundo Vasconcelos Hospital with a complaint of retroauricular "ball" in the left side for two months. The father reported the baby had pain in the left ear, otologic discharge, hearing loss and other ear symptoms. Normal ENT examination. He was submitted to computer tomography scan (CT) of the temporal bone that showed bony erosion in the left temporal bone (Figures 3-4). Subsequently a MRI was performed due the possibility of intracranial compromise (Figures 1 and 2). We suspected of LCH and after normal pre-operative exams, surgery was performed for further diagnosis and resection of the lesion. The resected material was sent to pathologic exams that confirmed LCH. After total resection of the lesion, the patient was submitted to a corticosteroid therapy for six months showing no symptons untill the present time.

Case 2

2. J.B. 2, 8 years old, female, native from Brazil, came to the ENT service of the Professor Edmundo Vasconcelos Hospital with complaint of otologic pain and persistent discharge in the right ear since 2 months of life, resistant to clinical treatment. After treatment without success for chronic middle ear infection in her hometown, she was submitted to temporal bone CT scan (Figures 5 and 6) that showed extensive lesion in the right temporal bone with associated bony erosion. So, the biopsy of the lesion was done and the material was sent to pathologic exams to confirm LCH. The following antibiologic panels were evident: S-100 protein +, CD68 +, CD4 +, CD8 +, etc. She was also submitted to complement exams to discard diseases. X-rays of long bones - normal. The bone scan ultrasound – normal. After total resection of the lesion, the patient was submitted to corticosteroid therapy, showing no symptoms until the present time.

Case 3

3. M.M.Z. 3 years old, male, natural from Sao Paulo, Brazil came to the ENT service of Professor Edmundo Vasconcelos Hospital after evaluation with his pediatrics. The parents noticed that the child always cryed when touched in the head. The mother noticed a softness in the left retroauricular region. The child was examinated by a pediatrician that asked for X-ray of the skull when a lesion in the same region of the skull was observed. The pediatrician sent the patient to our service. The ENT exam was normal except for a well defined lesion in the left retroauricular region in the scamous part of the temporal bone. The child was investigated for satellite lesions that were not found. He was submitted to a biopsy (LCH). In the same surgical time, a total resection of the lesion was done.

DISCUSSION

The eosinophilic granuloma is the most frequent and most benign form of the histiocytosis of the Langerhans cells. The disease was first described in medical literature around the turn of the 20th century. It occurs most frequently in children and is rare beyond the age of 20. The literature states that the great majority of the patients have no or few otologic symptoms, and the diagnostic suspect is usually based on radiographic demonstration of a destructive bone lesion in the temporal bone. One of our cases didn’t show any symptoms, but in two (chronic discharge and acute mastoiditis) we could find clinical findings. In our cases, bone lesions where present but without clinical or laboratory diagnosis. CT scan is usually helpful in identify lesions in areas with complex anatomy like the temporal bone. Six lesions with destruction of the mastoid, petrous ridge, lagena tympani, and lateral sinus plate and the destruction of the inner and external ear are seen on CT scans. CT may demonstrate an isointensating and homogenously enhancing mass. On spin-echo MRIs, lesions of LCH reveal decreased signal intensity on T1-weighted and high signal intensity on T2-weighted sequences. The lesion may enhance after the administration of a gadolinium-based contrast agent.

Concerning to diagnosis it is imperative to do an examination of tissue within the temporal bone. Microscopically, it is composed of large multinucleated giant cells with a polymorphous, eosinophil-rich inflammatory infiltrate. The differential diagnosis includes temporal bone osteomyelitis, non-Hodgkin’s lymphoma, Hodgkin’s disease, infectious diseases and metastasis. Special studies, including immunohistochemistry sometimes is necessary to resolve this differential diagnosis.

The current treatment of the LCH in the literature appears to be controversial. It can be carried with surgery, systemic chemotherapy, radiation and/or combination of these treatments. Treatment depends on the individual patient. In some cases the disease will regress without any treatment at all. In others, systemic chemotherapy will be prescribed depending on the extent of disease. Treatment is planned after thorough evaluation of the patient to determine the extent of involvement. In our three cases we decided to perform surgical treatment that consists in open cavity mastoidectomy in order to remove the macroscopic tumoral mass. We had completed surgical removal of the disease with transmastoid approach and with postoperative 60 days corticotherapy. No lesion was observed in postoperative computer tomography (CT), in a mean of 42 months of post-treatment follow-up the patients show no symptoms and no satellite lesions were observed until the current moment.

CONCLUSION

Langerhans cells histiocytosis is a rare disease that can affect the temporal bone in children. The lesions in the temporal bone in infants (eosinophilic granuloma) is a clinical entity whose high suspicious is important to define the diagnosis and further treatment. Employing surgical approach of the macroscopic tumoral mass and post-operative 60 days of corticotherapy we were able to control the disease in our cases in this serie.