CASE REPORT

Bilateral Temporal Bone Langerhans Cell Histiocytosis in a Two-Year-Old Boy

Aziz Mustafa MD; Qazim Hysenaj MD; Ph.D., Lumturije Gashi-Luci MD; Ph.D.

Afrim Heta MD; Hajrije Hundozi-Hysenaj MD; Ph.D.

From the Department of ENT/Head and Neck Clinic, From University Clinical Center, Prishtina, Kosova (A. Mustafa; Q. Hysenaj) A. Heta; H. Hundozi - Hysenaj and Department of Human Pathology, University Clinical Center, Prishtina, Kosova (L. Gashi-Luci)

Corresponding author:
Aziz Mustafa M.D.
ENT/Head and Neck Surgery Clinic
University Clinical Center of Kosova
Rrethi i Spitalit p.n.
10000 Prishtina
Kosovo/UNMIK

Tel: + 37744127591
+ 381385006003410
E-mail: aziz_mustafa2000@yahoo.com

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Histiocytosis X, or Langerhans cell histiocytosis (LCH), is a rare disease that consists of three less distinct, overlapping states: eosinophilic granuloma (EG), Hand-Schüller-Christian (HSC) disease, and Letterer-Siwe (LS) disease. Of the three, EG is the least severe and most localized form, and has the best prognosis. Temporal bone presentation is clinically similar to acute otomastoiditis. We diagnosed and treated without success a two-year-old patient with bilateral temporal bone LCH.

Histiocytosis X, or Langerhans cell histiocytosis (LCH), is a disease that consists of three less distinct, overlapping states: eosinophilic granuloma (EG), Hand-Schüller-Christian (HSC) disease, and Letterer-Siwe (LS) disease. Of the three, EG is the least severe and most localized form, and has the best prognosis. A high index of suspicion is required to diagnose EG, especially when an ear disease is refractory to medical treatment. Early detection is important for managing EG properly and minimizing the complications or squeal of treatment. The definitive diagnosis of histiocytosis is made histopathologically, with the immunohistochemical detection of S-100 and CD1 antigens in tissue samples [1].

Langerhans cell histiocytosis is characterized by monocytic-macrophagic system cell proliferation. The peak incidence period is between 1 and 4. The annual incidence is 3-4 per million, and the disease is twice as common in the male gender [2].

We diagnosed and treated a two-year-old patient with bilateral temporal bone LCH. This case is unique for our institution as the very first case properly diagnosed and treated. Despite appropriate treatment, patient died because of generalisation of the lesions in other bones.

CASE REPORT

A two-year-old boy was admitted to the Pediatric-ENT Department of the ENT Clinic of Prishtina University Clinical Center for bilateral retroauricular swelling. He had a history of continuous ear discharge for the previous 3 months, and was treated as outpatient with parenteral and topical antibiotics chosen empirically. Physical examinations revealed bilateral non-erythematous pre- and retroauricular swelling, with tender fluctuation resembling a subperiosteal abscess. An otomicroscopy examination revealed swelling and narrowing of the right external auditory canal, which was filled with granulation tissue, and an extremely narrow left external auditory canal in which the tympanic membrane could not be seen. Swabs for microbiology examination and a piece of granulation tissue for pathology examination were taken. The laboratory findings on admission were ESR 19/first hour, 24/second hour, RBC 3.48(1012/L, and hemoglobin 61 g/L. The WBC and platelets were normal. Computed tomography (CT) showed bilateral, relatively symmetric destruction of the temporal bone. A soft-tissue mass involving the ossicular chain was observed in the right tympanic cleft (Figure 1.). For his severe anemia, the patient was transfused with 150 ml of type O (zero) Rh + blood. After his RBC improved, the patient underwent bilateral incision and biopsy from both mastoids under general anesthesia. The planned mastoidectomy was not performed because of the lack of

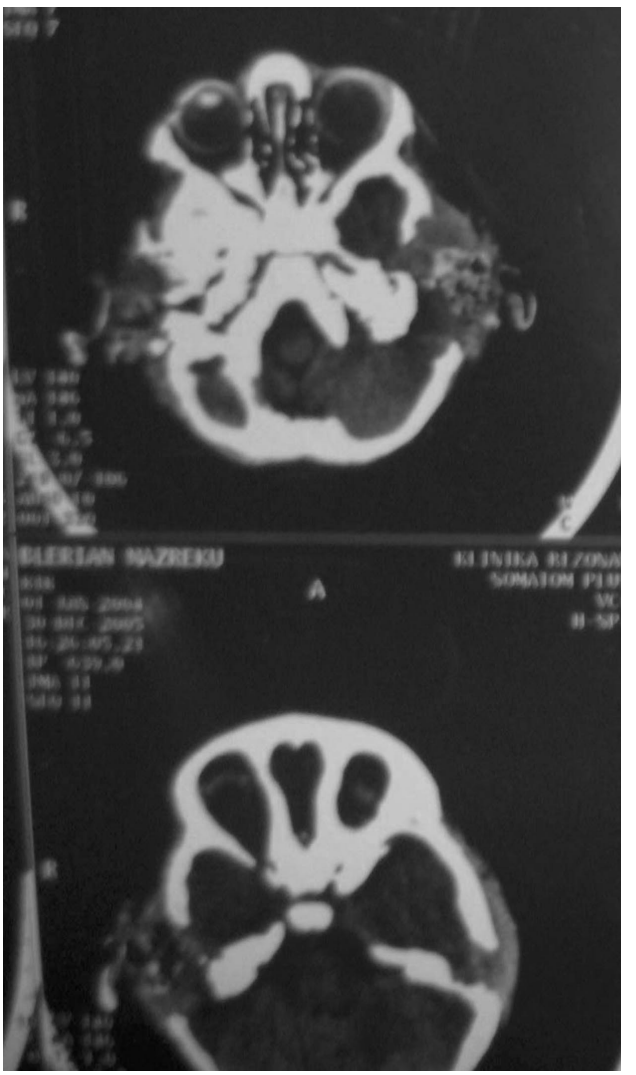

Figure-1: Computed tomography of the cranium shows bone destruction of both temporal bones; both tympanic clefts are filled with soft tissue masst.

landmarks. The mastoid cells were filled with granulation tissue that was easily curetted. Histopathology revealed granulation tissue, with fibrin deposits, cell detritus, capillary proliferation, and masses of granulocytes, lymphocytes, monocytes, fibrocytes, and giant inflammatory multinuclear cells. The immunohistochemical analysis was as follows: S100-positive in dispersed cell groups (Fig. 2), CD20-positive in normal B-lymphocytes (Fig. 3), CD45 positive (Fig. 4), and Ki67 in about 5% of the cell population (fig. 5). Desmin and cytokeratin were negative. Based on the histopathology and immunohistochemistry, the diagnosis of LCH was established. The child received chemotherapy with high-dose methylprednisolone (30 mg/kg i.v. over 3 consecutive days given during the first course of treatment only) and vinblastine (6 mg/m2 i.v. weekly for a period of 6 months). In the first 6 months, the patient had an improvement that was also seen in the

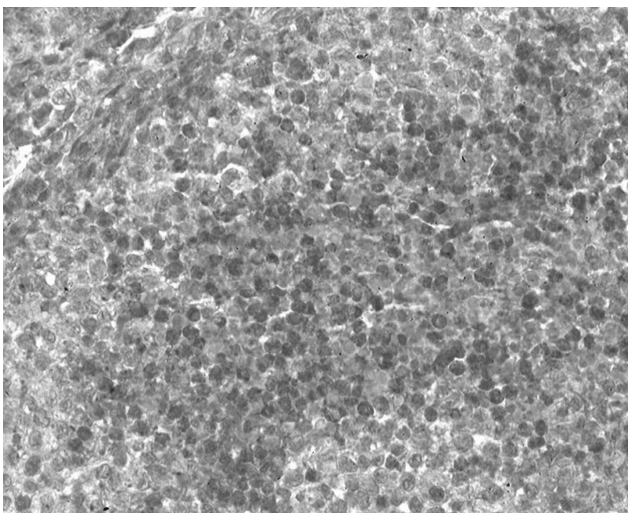

Figure-2: Histopathology of the mastoid cavity analyzed with S-100 immunohistochemistry marker. (Courtesy of Dr. Fisnik Kurshumliu, Pathology Institute of University Clinical center of Kosova, Prishtina)

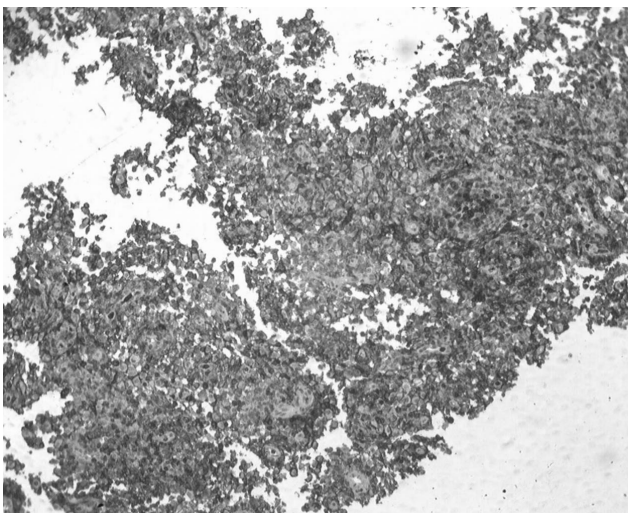

Figure-4: Histopathology of the mastoid cavity analyzed with CD 45 immunochemistry marker. (Courtesy of Dr. Fisnik Kurshumliu, Pathology Institute of University Clinical center of Kosova, Prishtina).

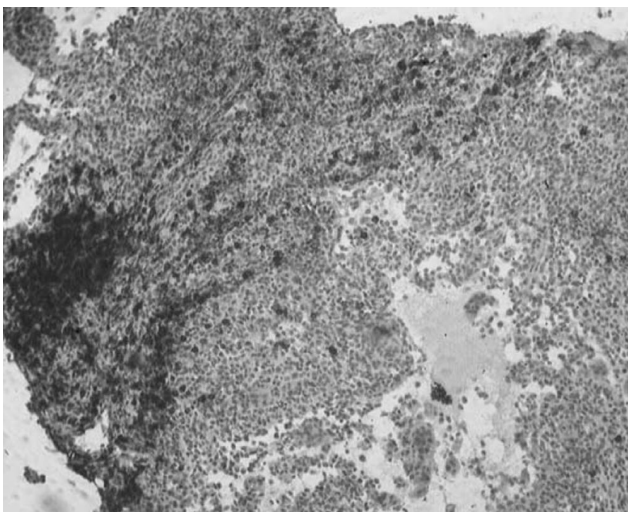

Figure-3: Histopathology of the mastoid cavity analyzed with CD 20 immunohistochemistry marker. (Courtesy of Dr. Fisnik Kurshumliu, Pathology Institute of University Clinical center of Kosova, Prishtina)

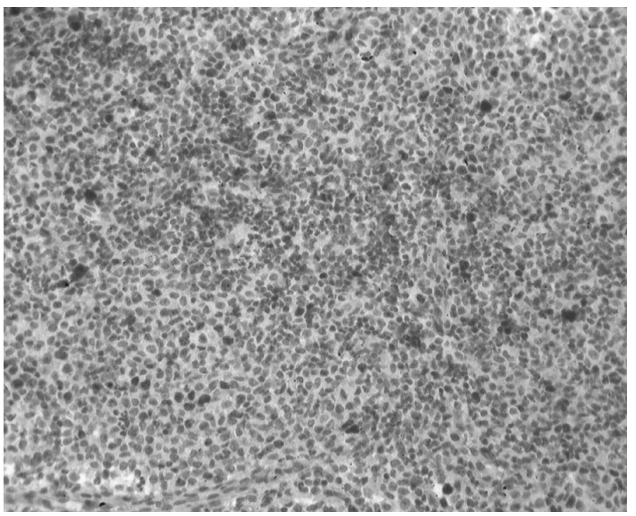

Figure-5: Histopathology of the mastoid cavity analyzed with Ki-67 immunochemistry marker. (Courtesy of Dr. Fisnik Kurshumliu, Pathology Institute of University Clinical center of Kosova, Prishtina).

temporal bone CT, but despite appropriate treatment, patient died because of generalisation of the lesions in other bones.

DISCUSSION

In 1953, Liechtenstein published an article on histiocytosis X, or LCH, a term he introduced to indicate that three previously described entities-EG, HSC disease, and LS disease-were the same entity and that its cause was still unknown.3 Involvement of the temporal bone in the course of LCH has been described in 15 to 61% of all cases. In the series of 62 patients published by Fernandez-Latorre et al., temporal bone involvement was present in 22.6%, and bilateral temporal involvement was seen in [4] (28%) of the 12 patients with temporal bone occurrence.4 In our case, the middle ear and temporal bone involvement was bilateral and occurred at an early age. Chronic otitis media with granulation tissue in the external auditory canal was also present. The retroauricular swelling led us to consider the differential diagnosis of subperiosteal abscess, as a complication of acute mastoiditis. With bilateral involvement, one must always suspect an idiopathic disease, such as LCH.

The main diagnostic procedures are imaging. X-rays of the mastoids always show osteolytic lesions of the temporal bone. Sometimes osteolytic lesions can be detected in other skull bones and the mandible, and may be solitary or multiple. CT and magnetic resonance imaging (MRI) can also be very useful for differentiating between LCH and other osteolytic lesions, such as osteomyelitis, epidermoid cyst, and metastases [5].

The CT image displayed in bone window clearly shows fibrous dysplasia as focal areas of ground-glass appearance, but more importantly there is preservation of the bony cortex even in the significantly enlarged surrounding bony structures, a characteristic feature of fibrous dysplasia.6 In our case, CT of both temporal bones showed osteolytic lesions, with no involvement of other skull or facial bones. The definite diagnosis was established after histopathology examination of bilateral biopsies.

The treatment of temporal LCH is combined chemotherapy, radiotherapy, and surgery. The role of otosurgery is limited to aural polyp removal and eventual curettage of the granulation tissue. Mastoidectomy is a surgical intervention with very high risk because of the lack of main temporal bone landmarks. Similarly, radiotherapy has been associated with significant early and delayed complications, especially in young patients and should be reserved exclusively for patients in whom vital structures are at risk [2, 4, 6, 7].

Based on these facts, the most suitable therapy is chemotherapy, which gives a good remission rate, and surveillance.

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