

LANGERHANS CELL HISTIOCYTOSIS IN CHILD TEMPORAL BONE: A CASE REPORT

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Abstract

Langerhans Cell Histiocytosis (LCH) is one of the rare disease with atypical clinical manifestations. LCH must be watched out for and considered a differential diagnosis of the other otolaryngology diseases. A 2-years-10-months old boy with a crusted lump in the right ear canal accompanied by a lump behind the right ear that is expanding, bleeding for 3 months, and the patient experience loss of weight. Examination of the ear found a black mass covered with crusts with a firm, fixed consistency, filling the lateral one-third of the right external acoustic canal to the right auricle concha. A solid mass was obtained from the lateral one-third of the right external acoustic canal on a head CT scan, medially obliterating the external acoustic canal to the right internal acoustic canal. FNAB found nonspecific inflammation. Performed mass excision surgery and anatomical pathology examination. The patient was referred to the pediatric haemato-oncology division and underwent chemotherapy. Langerhans cell Histiocytosis in the temporal bone is a rare disease with atypical clinical manifestations, so it should be considered as one of the differential diagnoses if a mass is found in the external acoustic canal.

Keywords: Histiocytosis, Temporal bone, Vinblastine

Introduction

Langerhans cell histiocytosis (LCH) or histiocytosis X is a group of idiopathic disorders with a characteristic feature of an increased number of histiocytes. Histiocytes are a type of white blood cell, which act as destroyers of foreign material in blood and tissues. Recent studies have shown the involvement of Langerhans cells, leading to the use of the term LCH instead of the term histiocytosis X. [1] These findings were further investigated and it was found that Langerhans cells play a big role in this disorder, hence the term Langerhans Cell Histiocytosis (LCH) is used. Langerhans cells are cells that accumulate in various parts of the body as antigen-presenting cells. Lesions in LCH are heterogeneous in cell composition, anatomic distribution, clinical behavior, and prognosis.[1]

The incidence of LCH as a rare disease is 0.5-5.4 cases per million persons per year.[2] An epidemiological study reported a male predominance of 1.2 to 1.5, with the highest incidence at 1 to 3 years of age. Recent studies have reported a correlation between the

How to cite: Andrio Raymos, Sally Mahdiani, Lina Lasminingrum (2022) Langerhans Cell Histiocytosis in Child Temporal Bone: A Case Report, (7) 09. Doi: 10.36418/syntax-literate.v7i9.13763

E-ISSN: 2548-1398

Published by: [Ridwan Institute](#)

incidence of LCH with maternal and neonatal infections, non-immunization at a young age, family history of thyroid disease, in vitro fertilization, eating disorders, and transfusions in infancy. Disseminated LCH is reported in individuals with lower socioeconomic conditions.[3]

LCH can occur in various organs or systems in the human body, and the most involved organ is bone in about 80% of cases, with temporal bone involvement in 4% of cases.[4] Other organs that may be involved include skin (33%), pituitary gland (25%), liver, spleen, hematopoietic system, lungs (15%), lymph nodes (5 - 10%), and central nervous system other than the pituitary gland (2-4%).[4,6] Characteristics of LCH, which can involve multiple organ systems, create challenges in diagnosing this disease and its rarity also results in limited data related to LCH.[5] This case report aims to add knowledge in diagnosing this disease so that LCH patients can be treated well.

Case Presentation

A 2-years-10-months old boy was referred with a diagnosis of right mastoid tumor. A crusted and bleeding lump was found in the right ear canal 3 months ago, accompanied by a lump behind the right ear which has been getting bigger since 2 months ago and is painful. There is a history of picking at the right ear and a history of weight loss. There was no history of fainting, seizures, nausea, and vomiting.

On physical examination, the child was conscious, with no shortness of breath or cyanosis. Heart rate is equal to the pulse rate of 100 beats per minute, respiratory rate 24 times per minute, axillary temperature 36.7°C, and SpO₂ 99% of room air.

In the right ear, a black mass covered with crusts that bleed easily, with a firm, fixed consistency, attached to the lower wall of the ear canal, tenderness that filled the auricle concha and covered the lateral one-third of the external acoustic canal was seen (Fig.1A). In the right retro auricular, a mass was palpable with a firm consistency, 5 cm in diameter, ill-defined borders, and tenderness (Fig.1B). Left ear within normal limits. There were no abnormalities in the nose and oropharynx. The maxillofacial was symmetrical and there were no signs of cranial nerve deficits and no regional lymph node enlargement was found in the neck area.

Laboratory results showed a decrease in hemoglobin and an increase in leukocytes. Other laboratory results were within normal limits. FNAB in the right retro auricular mass found debris, fibrin, and many inflammatory cells of lymphocytes, histiocytes, and PMN.

A CT scan of the head (Fig.2) showed a solid mass originated from the subcutis of the right external acoustic canal, medially obliterating the external acoustic canal to the right internal, right mastoid air cell, right mastoid antrum, right epitympanic recess wall, and right prussak space. Destruction of the right mastoid bone was seen. No intracranial metastases were seen. The patient was consulted to the neurosurgery department and diagnosed with suspicion of cholesteatoma infection with a differential diagnosis of the epidermal cyst and suggested for a re-biopsy. A joint conference was held between the otology division and the oncology division of the ENT-HN Faculty of Medicine Universitas Padjadjaran, the patient was diagnosed with epidermal cyst differential diagnosed with a brachial cyst at the mastoid region and right external acoustic canal. It was decided in this patient to undergo mass excision and biopsy.

Mass excision surgery was performed by incision at 1 cm from the retro auricular sulcus, then dissection of the area around the mass and removal of the tumor mass, mastoid drilling, and identification of the facial nerve. The remaining mass was removed, sufficient deep fascia was taken to be used as a graft and placed on the *aditus ad antrum*, then meatoplasty was performed on the right external acoustic canal, and the surgical wound was closed layer by layer.

Intra-operative findings include right mastoid planum, mastoid tegmen, posterior wall, external acoustic canal, and then erosion on malleus and incus. The mastoid antrum and tympanic cavity are filled with red encapsulated masses, not easily bleed, and there is a dehiscence of the facial nerve.

Histopathology of temporal bone lesion biopsy specimens revealed a dense monotonous cell arrangement underlying the squamous epithelium. These cells are cells with an eosinophilic cytoplasm with a grooved, kidney-shaped nucleus, which is characteristic of LCH. Eosinophils and multinucleated giant cells were also found (Fig.3A). Immunohistochemical staining was performed, namely CD1a, S100, and langerin, and a positive result was obtained which is pathognomonic of LCH (Fig.3B).

The patient was referred to the pediatric haemato-oncology division of the Department of Pediatrics and underwent a 12-month treatment protocol consisting of chemotherapy with intravenous bolus injection of vinblastine for the first 6 weeks ($6\text{mg}/\text{m}^2$) in combination with prednisolone ($40\text{mg}/\text{m}^2$). Initial therapy consisted of oral prednisolone $40\text{mg}/\text{m}^2$ in 3 divided doses for 4 weeks with tapering off for 2 weeks. The patient tolerated therapy well and there was an improvement in symptoms.

Result and Discussion

Langerhans cell histiocytosis in the temporal bone is a rare condition with the incidence of LCH in the temporal bone only 4% of all LCH cases in the world. LCH can occur in all populations, but LCH in the temporal bone is mainly found in boys and children (about 75-90% of temporal LCH patients are boys) with a peak incidence at 1 to 3 years old.[6] In this case, the patient characteristics matched the epidemiology of temporal bone LCH, i.e. the patient was a boy aged 2 years and 10 months which was still in the peak incidence of temporal bone LCH. Children with LCH under 2 years of age have a worse prognosis than children over 2 years of age.

The most common clinical presentation in LCH patients is painful bone lesions and rash. In addition, non-specific symptoms that can also be found include fever, poor appetite, weight loss, fatigue, the patient becoming sensitive, and changes in behavior.[7] In this case, there was a history of weight loss and the presence of scabs and lumps behind the right ear. Based on the literature, bone involvement in LCH is characterized by pain and swelling in the involved area, so a lump and scab in the right ear may be a sign of bone involvement in the area around the mastoid.[7]

Ear and skull involvement was found in the form of a black mass covered with crusts that bleed easily, elastic consistency, fixed to the ear canal wall, with tenderness covering the external acoustic canal. These masses are similar to lesions caused by other, more common conditions such as cysts or cholesteatoma. Misdiagnosis of LCH due to involvement of the

outer and middle ear is similar to other, more common conditions. Previous studies reported a misdiagnosis rate of up to 72.7% for LCH because of this similarity.[8] Therefore, if an external acoustic intracanal mass or retro auricular mass is found, it is necessary to consider LCH as one of the differential diagnoses.

Confirmation of the mass by cytological examination can help establish the diagnosis of LCH. The classic finding on LCH cytology is the discovery of many Langerhans cells mixed with a population of eosinophils, neutrophils, lymphocytes, plasma cells, multinucleated giant cells, and macrophages. Sometimes Langerhans cells have poorly defined cytoplasmic curvatures and processes, these things may be difficult to identify. This may have resulted in findings on FNAB biopsy results in patients being interpreted as having non-specific inflammation with cystic lesions so that the differential diagnosis of LCH is an epidermal cyst, brachial cyst, and right ear cholesteatoma.[9]

Confirmation of the mass by histopathological biopsy can help in the diagnosis of LCH. A common histopathological finding in LCH is Langerhans cells resembling epithelioid cells with dominant and eosinophilic cytoplasm and reniform nuclei. These cells are easier to identify and often show diffuse infiltration, particularly in bone and skin. Eosinophils are also commonly scattered throughout the infiltrate. However, the presence of eosinophils is not a must for the diagnosis of LCH.[1] Diagnosis with biopsy also can be helped with CD207 (langerin) and CD1a detection with the immunohistochemical method.[1,10]

The pathogenesis of LCH is still being debated today. Debate exists regarding the pathogenesis of LCH, whether because of neoplastic transformation or activation of the immune system in the form of activated epidermal Langerhans cells. Recent studies suggest that LCH is better considered as a result of the neoplastic transformation of myeloid cell precursors, so LCH is treated with chemotherapy. [11]

Therapy modalities that can be used in LCH cases include operative or surgical therapy, radiotherapy, chemotherapy, and steroid injections. LCH therapy can be determined based on risk factor stratification. The patient had bony lesions with low-risk areas, LCH with lesions in this area is rarely fatal, but more than half require >1 treatment. Based on the HSL protocol, III the current therapy is vinblastine and prednisone combination therapy for 1 year. The frequency of reactivation may decrease with 1-year duration of therapy. [6,10] In this patient, chemotherapy was done according to LCH III protocol, and received an initial post-treatment recovery for 6 weeks.

Age of onset determines the prognosis in LCH patients, with a better prognosis in adult patients. Localized LCH has a better prognosis than LCH involving multiple organs.[10] The patient is a pediatric patient, but the lesions are localized, so it is hoped that appropriate therapy with standards will be able to provide a good outcome.

Conclusion

LCH is a rare disease in the form of neoplastic abnormalities in Langerhans cells. The diagnosis of LCH still faces great challenges because of the diverse clinical manifestations and similar to the conditions commonly found, so LCH should still be used as one of the differential diagnoses if masses are found in the external acoustic canal.

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First publication right:

Syntax Literate: Jurnal Ilmiah Indonesia

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