A case of Langerhans cell histiocytosis mimicking child abuse

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Langerhans cell histiocytosis is a rare non-malignant disease with clinical heterogeneity. The disease may present with various clinical findings and may imitate many other conditions. In this report we describe a 34-month-old girl who presented with chronic otitis and otorrhea, skull fracture, rash, vulvar edema, erythema and erosion in labia majors which initially suggested child abuse but the patient was diagnosed with Langerhans cell histiocytosis.

Key words: Langerhans cell histiocytosis, child abuse.

Langerhans cell histiocytosis (LCH) is a myeloid cell-based neoplasm characterized by the reactive clonal increase of Langerhans cells and consisting of the components of autoimmune inflammatory disease¹. Moreover, this disease is genetically related to BRAF-V600E gene mutation influencing the MAPK signaling pathway. Incidence rate is 8.9/million per year among children under the age of fifteen ². Potential symptoms encountered in LCH diagnosis in adults and children may also be observed in numerous diseases. Differential diagnosis is based on the presentation of typical histopathological findings or CD1a positive cells³.

Tissue damage that was caused by someone else due to any reason is defined as physical abuse ⁴. The awareness of conditions that simulate child abuse enables the ability to diagnose correctly. We presented a case of LCH mimicking child abuse in this report.

Case Report

A 34-month-old girl was admitted to hospital with fever, rash, ear discharge and vaginal secretion. Medical history revealed crusted rash on the scalp and intertriginous areas and ear drainage from six months of age. Systemic and topical antibiotic treatments were prescribed several times with the diagnosis of acute otitis media and seborrheic dermatitis in other hospitals. At the age of one, laser polypectomy was performed through both of the external auditory canals. In addition to these complaints, vaginal secretion was observed for the last five months, and fever and rash were noted for the last five days. The patient, whose parents were third-degree relatives, had a one-year-old healthy sister.

On physical examination, her general condition was moderate and she was apparently agitated. Remaining data about her physical examination were as follows; body temperature 38.1 °C, respiratory rate 24/min, pulse 101/min, arterial blood pressure 100/60 mm Hg. Apparent and locally yellowish clotted plaque was seen on the scalp, and broad erythematous skin was observed beneath it. Petechial-purpuric rashes were seen on the whole trunk and extremities. Bilateral external auditory canal was obliterated with polypoid lesion and purulent discharge. Purulent discharge, vulvar edema, erythema in labia majors, and local erosion were detected on intertriginous areas on the examination of genitourinary system. Other system examinations were normal.

Routine laboratory investigations were as follows; hemoglobin (Hb) 8.9 gr/dl, white
blood cell count (WBC) 14,900/mm$^3$ (60% neutrophil, 30% lymphocyte, 8% monocyte, %2 eosinophil), platelet count 661,000/mm$^3$, C-reactive protein (CRP) 174 mg/dl (0-4 mg/dl), erythrocyte sedimentation rate (ESR) 71 mm/h, aspartate aminotransferase (AST) 15 U/L (<48 U/L), alanine aminotransferase (ALT) 73 U/L (0-56 U/L), and gamma-glutamyl transpeptidase (GGT) 261 U/L (<23 U/L). Other biochemical evaluations and coagulation tests were normal. Urinalysis revealed plenty of leukocyte and urine leukocyte esterase strip were 3(+). Cerebrospinal fluid (CSF) examination was normal. Serologic tests for human immunodeficiency virus (HIV), Ebstein-Barr virus (EBV), cytomegalovirus (CMV) and syphilis were all negative, and serum quantitative immunoglobulin levels were normal according to her age.

Ceftriaxone was empirically commenced for the suspicions of urinary tract infection and chronic vulvitis. Cultures of blood and CSF were resulted as negative. Temporal computed tomography (CT) scan revealed numerous fractures in right temporal and bilateral mastoid bone. Cranial CT was performed due to suspicion of child abuse and no cerebral hemorrhage or contusion were revealed. Abdominal ultrasonography(USG) showed heterogeneity of liver and these findings were interpreted as a lesion that might develop depending on contusion, infiltrative disease or infectious process. Whole body direct bone survey graphics were normal.

A diagnosis of physical child abuse was suspected because of the bilateral fracture in temporal CT, the abdomen USG image that compatible with the liver contusion and purulent genital discharge, vulvar edema, erythema in labia majors and the local erosion that was detected on the examination of genitourinary system. Further social history of family was questioned by social workers. Contrast-enhanced temporal magnetic resonance imaging (MRI) revealed soft tissue intensities in both of the squamous and mastoid parts of temporal bone, surrounding the external auditory canal and extending to the tympanic cavity. Diffuse contrasting was observed in the meningeal structures at the levels of both temporal lobes (Fig. 1). Histiocytosis X was firstly considered in the differential diagnosis. Histopathological analysis of the biopsy specimen taken from the lesions on the scalp and axillary area demonstrated an infiltration developing in a way of filling the upper dermis under the epidermis, having the diffuse developmental
pattern that extending to the lower dermis, and consisting of atypical cells with big vesicular anisocoric nucleus, small nucleolus, and large eosinophilic cytoplasm (Fig. 2). Giant cell formation was not recorded. The patient was diagnosed as LCH according to these findings. In the bone scintigraphy taken for staging the disease, focally increased activity involvement was recorded on the area complying with the adjacency of left temporal bone—sphenoid bone, and the distribution of activities was measured to be at the desired level and symmetry in the lines of epiphysis and for other bones in the body. Bone marrow aspiration revealed no involvement. The treatment protocol with vinblastine and prednisone was administered to the patient. She has been on follow-up for eight months uneventfully, her MRI findings returned to normal.

**Discussion**

Langerhans cell histiocytosis has been defined as a heterogeneous clinical condition occurring with the clonal proliferation of bone marrow-based dendritic cells of the epidermis. The disease may involve any organ or tissue. Although the involvements of skeletal system and skin are the most commonly seen, parenchymal organs, lymph nodes, and central nervous system may also be involved. Nowadays, LCH classification is made by taking into account the number of affected areas (single or multi-systemic and local or multifocal) and involvement of the risk organs. Bone marrow, liver, and spleen involvement are considered as high stage disease. Ear involvement and rash related to the skin involvements that are commonly seen in this disease were present in our patient whose complaints started when she was six months old. The patient was evaluated as having systemic form as expected for that age group. The involvement of eyes and endocrine system which may be observed in the systemic form was not present in our patient. Similar to this case, the cases with chronic otitis and otorrhea depending on the temporal bone were reported, previously.

Although the physical examination findings are numerous in child abuse, ecchymosis, bite marks, fractures, injuries in internal organs, and head trauma are the most commonly reported findings. Skull fractures may be present due to accidents as well as the child abuse. Especially more than one complicated skull fracture and compression fractures are the indications increasing the probability of child abuse. In the present case, child abuse was included in the differential diagnosis due to the bilateral fracture in temporal CT and the ultrasonographic images that were compatible with the liver contusion. A study that included 25 pediatric LCH patients reported that, LCH imaging findings are not specific, they might be similar to the infection, inflammation or neoplastic diseases, and they might even imitate...
the traumas not related to accidents. Skin findings of LCH may also vary to some extent. The differential diagnosis should include especially seborrheic dermatitis, psoriasis, atopic dermatitis and scabies. Acrodermatitis enteropathica, ‘blueberry muffin’ type rash, and molluscum contagiosum were reported as the findings that might be rarely confused with LCH. Recurrent skin findings of our patient had been evaluated as seborrheic dermatitis, and the treatment was administered according to this diagnosis. Petechial rash is a rare symptom seen in LCH. In this case, the probability of child abuse was considered due to the presence of distinct vulvar, labial edema and laceration, her unfavorable general hygienic condition, and the detection of the images compatible with fracture and contusion. Since the detection of vulvar lesions in the age group of children is a highly stimulant factor in terms of child abuse, professional social service evaluation was made. It was reported that, a 4-year-old patient with bilateral vulvar vesicular lesions and surrounding erythema, who developed diabetes insipidus during the follow-up, was diagnosed as vulvar LCH with central nervous system involvement.

Langerhans cell histiocytosis is diagnosed through the clinical, radiological findings and histological and immunophenotyping analysis of the biopsy taken from the lesion or involved organ if any in systemic involvement. The present case had temporal MRI findings compatible with histiocytosis X, bilateral external auditory canal obliteration, hepatic, vulvar and skin involvement. For this reason, systemic form of LCH was considered and diagnosis was confirmed by means of typical histopathological findings on biopsy. When the cases of child abuse are early noticed, the abused children can be saved from chronic abuse or death. For this reason, detailed medical history and analyses of physical examination findings are quite significant for the early diagnosis of child abuse. LCH may present with various clinical findings and may imitate many other conditions. LCH should be considered in cases of chronic otitis and otorrhea that are resistant to the medical treatment, and ear involvement may result with formation of polyps and also the disease should be kept in mind in young children who had complaints similar to the findings of child abuse.

REFERENCES