Hand-Schuller-Christian Disease: A Case Report

HAND-SCHÜLLER-CHRISTIAN HASTALIĞI

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Abstract

Langerhans cell histiocytosis (LCH) is a disease of unknown cause, characterized by the benign proliferation of a distinct cell type which is similar to Langerhans cell. Hand-Schuller-Christian disease is the chronic, progressive and multifocal form of LCH. It is characterized by four findings: bone lesions, diabetes insipidus, exophtalmus and mucocutaneus lesions.

A 3-year-old boy attended to our clinic with seborrheic dermatitis like skin lesions on his back and scalp, and subcutaneus tumoural lesions on the cranium. In detailed examination exophtalmus, osteolytic lesions of the cranial bone, and otitis media were detected. The histopathology of the lesions was consistent with LCH.

In this report we present a case of Hand-Schuller-Christian disease who was misdiagnosed and treated as seborrheic dermatitis several times.

Key Words: Langerhans cell histiocytosis, seborrheic dermatitis

Özet

Langerhans hücreli histiyositozis (LCH), Langerhans hücresine benzeyen farklı bir hücre tipinin benign proliferasyonu ile karakterize sebebi bilinmeyen bir hastalıktır. Hand-Schüller-Christian hastalığı LCH'ın kronik, progresif ve multifokal bir formudur. Kemik lezyonları, diabetes insipitus, ekzoftalmus ve mukokutanöz lezyonlar olmak üzere dört bulgu ile karakterizedir.

Üç yaşında erkek olgu sırtında ve saçlı derisinde seboreik dermatit benzeri lezyonları ve kafada subkutanöz tümorleri nedeniyle kliniğimize başvurdu. Detaylı incelemede olguda ekzoftalmus, otitis media ve kraniumda osteolitik lezyonlar saptandı. Lezyonlardan alınan biyopsinin histopatolojik incelemesi LCH ile uyumluydu.

Bu yazıda defalarca seboreik dermatit yanlış tanısı ve tedavisi alan Hand-Schüller-Christian hastalığı tanılı bir olguyu sunuyoruz.

Anahtar Kelimeler: Langerhans hücreli histiyositozis, seboreik dermati

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Langerhans cell histiocytosis (LCH), is a rare disorder characterized by the abnormal proliferation of Langerhans cells. Hand-Schuller-Christian disease is the chronic, progressive and multifocal form of LCH and is characterized by four findings: bone lesions, diabetes insipidus, exophtalmus and mucocutaneus lesions.[1,2]

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In this report we present a case of Hand-Schuller-Christian disease that was misdiagnosed and treated as seborrheic dermatitis for several times.

Case

A 3-year-old boy attended to our clinic for the rash on his scalp and trunk that had been present for the last year. The patient was healthy at birth and had an unremarkable past medical history. The patient's father indicated that his son had recurrences of drainage from his right ear and a mass on his scalp for a few months and both of the skin eruption and drainage from the ear were resistant to the given therapies. We have learned that before coming to our clinic, he was operated in another hospital where he had applied for the subcutaneus

mass localized on the right parietal region in the cranium. The pathological examination of the mass revealed the diagnosis of "Rhabdomyosarcoma". When the histological specimens were examined once again in another center , the diagnosis showed histiocytosis.

There was no pathology on physical examination except for exophtalmus of the both eyes. On dermatological examination, on the scalp he had lesions covered by clingy, yellowish scales overlying on an erythematous surface. On the 1 cm inferolateral of the right eye, there was an 8 mm papular lesion with a haemorrhagical crust (Figure 1). On his back, there were a few erythematous papular lesions with clingy yellowish scales (Figure 2). Our patient had been treated several times in different hospitals for the existing lesions and was misdiagnosed as "seborrheic dermatitis". He did not respond to the given treatments.

The hematological examination revealed micrositer anemia. The routine chemistry panel and vazopressin levels were normal, but X-ray graphies of cranium showed multiple osteolytic lesions (Figure 3). On the cranial computer tomoghraphy multiple osteolytic lesions and a mass filling the right temporal fossa and damaging the adjacent bones were observed. The biopsy of the lesions on his scalp revealed an infiltration in the papiller and upper reticuler dermis. The cells in the infiltrate resembled the Langerhans cell histiocyte which were polygonal shaped, with a reniform nucleus and slight eosinophilic cytoplasm (Figure 4 and 5). In immunohistochemical examination, Langerhans cells were stained with S-100 positively (Figure 6). These findings were consistent with LCH.

Depending on both clinical and histopathological findings, the patient was diagnosed as "Hand-Schuller-Christian Diseae", which is the chronic multifocal and progressive form of LCH.

The patient is now followed by the oncology clinic of pediatrician and vinblastine and prednisone were started as a treatment.

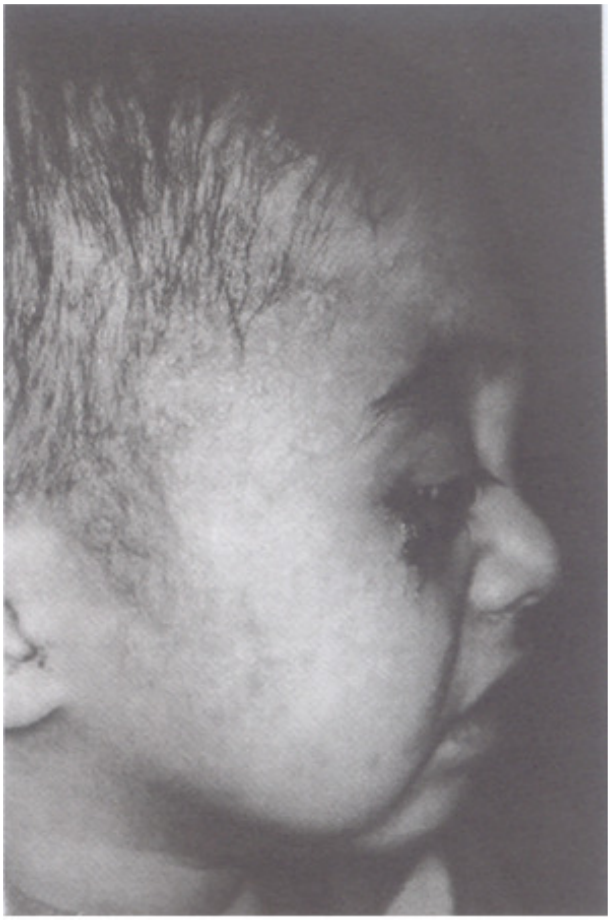

Figure 1. The appearance of the lesions on the scalp and inferior of the right eye.

Discussion

Langerhans cell histiocytosis (LCH); is a disease of unknown cause, characterized by the abnormal proliferation of a distinct cell type which is similar to Langerhans cell (pathological Langerhans cell) which takes S-100, CD-1 stains and contains cytoplasmic Langerhans granules.[1-5]

The exact cause of disease is still unclear. But there has been some hypothesis carried out. The most acceptable one is the viral theory of which HHV-6 is accused. The other suggested theory is the immunological one but until now it hasn't been shown a consistent immunological defect that confirms this theory. The neoplastic theory is also not supported since the appearance of the cells and the course of the disease does not support this theory.[1-3]

Figure 2. The appearance of the lesions on the back of the patient.

Hand-Schuller-Christian Disease (HSCD); is the chronic, progressive, multifocal form of LCH. In most of the cases, it begins in the ages of 2-6. The disease is characterized by four findings: bone lesions, diabetes insipidus, exophtalmus, and the mucocutaneus lesions. It is uncommon to find all the four findings in one person. The beginning findings are frequently diabetes insipidus, exophtalmus and the cutaneus lesions. Bone lesions are common findings which are frequently found on cranial bones and especially on the temporoparietal region and characterized by limited osteolytic area. Over the bone defect there may be soft tissue nodules. Bone lesions on the mastoid area cause otitis media. Diabetes insipidus is found over 50% of the cases and is suggested as a good prognostic factor. Exophtalmus is found in 30-40% of the cases and is seen both unilateral and bilaterally. Mucocutaneus lesions occur in about 1/3 of the cases. The skin lesions are especially localized on the laterals of the chest, back and the temporoparietal region. The intertiginous areas and the

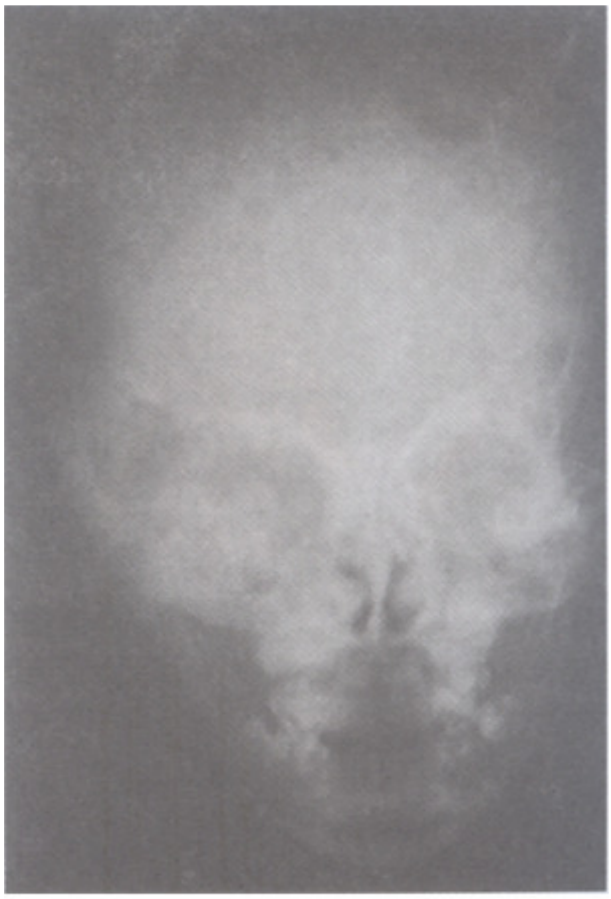

Figure 3. The appearance of the osteolytic lesion.

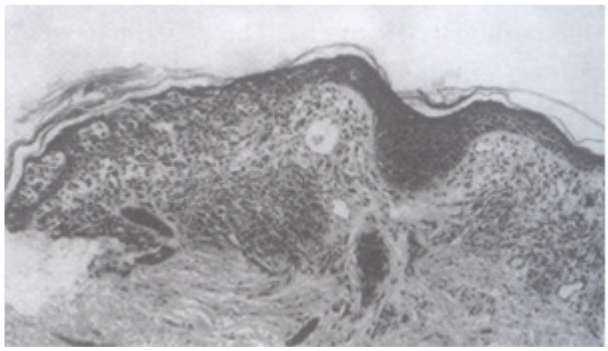

Figure 4. The histopathological appearance.

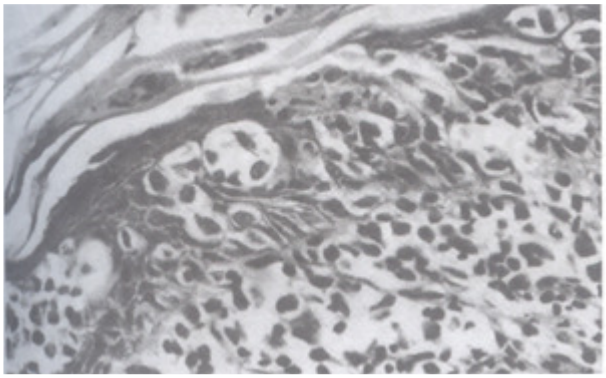

Figure 5. The appearance of the Langerhans cell.

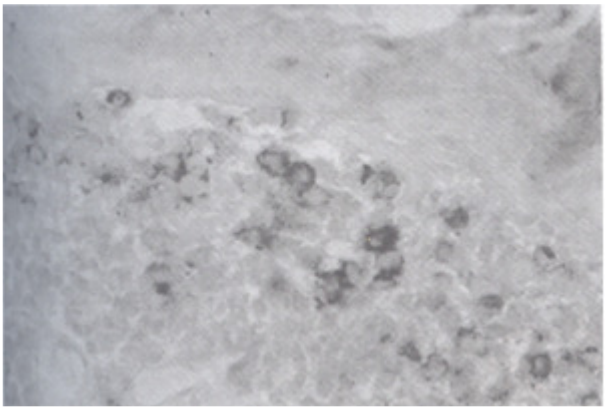

Figure 6. The cells that were stained with S-100 positively.

mucosal surfaces can also be involved. The systemic involvement is found in a few patients. The pulmoner system involvement occurs in 20-30% of the cases, while hepatosplenomegaly and lymphadenopathy are found rarely.[1,3-5]

Histopathologically; the different clinical forms of the disease show a common cell type; that is likely to Langerhans cell, which is a large cell with a reniform nucleus, slight eosinophilic cytoplasm. Electron microscobic studies show the cells contain Birbeck granules in the 50% of the cells. The cells are stained with S-100 and CD-1 positively.[1-8]

HSCD is a chronic disease, and is progressive and is fatal if not treated.[1,3-5]

In the differential diagnosis; skin lesions can be confused with some of the diseases, especially "seborrheic dermatitis". The other diagnosis that have to be considered are; moniliasis, Darier's disease, lichen nitidus, staphylococcal pyodermi, varicella, eosinophilic folliculitis, benign cephalic histiocytosis, generalized eruptive histiocytosis.[1-3,6-9]

Our case was diagnosed as Hand-Schuller-Disease because of the coexistence of the cutaneous lesions that are found on his back and scalp; the subcutaneus tumours in the cranium, the osteolytic lesions in the cranial bones, exophtalmus and otitis media. Because it is a rarely seen case and it can be confused especially with seborrheic dematitis, we decided to present this case.

As it is seen in our case; LCH and seborrheic dermatitis can easily be confused,we have to consider other diseases and especially LCH in the cases that is similar to seborrheic dermatitis which don't respond the treatment carried out.

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