Solitary Langerhans' Cell Histiocytosis of the Occipital Lobe: A Case Report

Oksipital Lob Soliter Langerhans Hücre Histiyositozu: Bir Olgunun Sunumu

EMİN ÖZYURT, SÖHRET ALI ÖÇUZOĞLU, AHMET HİLMİ KAYA, TANER TANRİVERDİ, TİBET KAÇİRA

University of Istanbul, Cerrahpasa Medical Faculty, Department of Neurosurgery (EO, SAO, AHK, TT, TK)

Received: 24.12.2001 ∙ Accepted: 5.3.2002

Abstract: Objective: Langerhans' cell histiocytosis is a disease that can involve many organs or remain localized to one organ. The hypothalamus is the most common site of solitary involvement of the central nervous system, and other locations have only been reported rarely. This report describes an unusual case of solitary Langerhans' cell histiocytosis of the occipital lobe, and discusses the clinical findings, radiological features, and treatments for this condition.

Methods: A 4-year-old girl presented with epilepsy of unknown origin. Electroencephalography and diagnostic imaging identified a lesion in her left occipital lobe. The patient was surgically treated with transoccipital lesionectomy and posterior mesial temporal resection.

Results: The histopathological diagnosis was lymphoplasmacytic infiltration of the leptomeninges and the underlying brain. Postoperatively, the patient had transient hemiparesia for 3 weeks, but recovered well otherwise. At 10 months post-surgery, there was no clinical or radiological evidence of recurrence. No adjuvant therapy was needed.

Conclusion: The small number of similar cases in the literature provide little information on which to base decisions about adjuvant therapy after surgical excision. Our patient's outcome, and the results in most cases with long follow-up indicate that total excision of the lesion is satisfactory and sufficient treatment.

Key words: Central nervous system, Langerhans' cell histiocytosis, solitary lesion


Yöntem: Dört yaşında kız çocuklu eplepsi nedeni ile klinigimize başvurdu. Elektroencefalografisi ve görüntüleme tectikleri sonrasında saptanan oksipital lob yereşimli tümoral lezyonu transoksipital yolla eksiye edilerek, posterior mesial temporal bölgeye de eksizyon uygulandı.


Sonuç: Cerrahiden sonra yardımcı tedavilere gerek duyulup duyulmamasına karar verdirebilecek kadar literatürde yeterli vaka bildirimmesine rağmen, total olarak çıkarılan vakalarda cerrahinin yeterli ve tatmin edici olduğunu düşünmekteyiz.

Anahtar Kelimeler: Merkezi sinir sistem, langerhans hücre histiyositozu, soliter lezyon.
INTRODUCTION

Langerhans' cell histiocytosis (LCH) is a rare disease characterized by clonal proliferation of histiocytes in various tissues (20). The central nervous system (CNS) is a rarely affected, and neurological dysfunction occurs in only 1-4% of patients with LCH. When the disease does involve the CNS, the pituitary-hypothalamic axis and the cerebellum are the areas most frequently affected. Solitary CNS involvement outside the hypothalamus and without infiltration of the meningeal structures and the cranial vault is rare. To date, only 14 cases have been reported in the literature (1,3,7,8,10,11,13,18). In this paper, we describe a case of solitary LCH of the occipital lobe, and discuss the clinical findings, radiological features, and treatments for this condition.

CASE

A 4-year-old girl presented to our clinic with epilepsy of unknown origin. Her first seizure had occurred 4 months prior to presentation, and was a generalized tonic attack with status epilepticus that lasted for 45 minutes. She was started on antiepileptic medication after this initial event. However, she then experienced three consecutive partial epileptic attacks while on the medication. Each of these attacks started with gastrointestinal discomfort and progressed to right-sided tonic-clonic seizures of the face, arm, and leg that lasted for 5 minutes.

Upon admission to our center, the patient's neurological examination and laboratory findings revealed nothing abnormal. Interictal electroencephalography showed disorganized electrical activity in the left temporal and occipital lobes. Magnetic resonance imaging (MRI) revealed a lesion in the left occipital lobe. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images, and we noted surrounding edema that extended to the mesial temporal region. There was marked enhancement after contrast injection (Figures 1a, b, c). The tentative preoperative diagnosis was ganglioglioma. The patient was taken to surgery, where she underwent left occipital craniotomy, transoccipital lesionectomy, and posterior mesial temporal resection.

Postoperatively, she exhibited right hemiparesia that lasted for 3 weeks. Histopathological examination of the surgical specimen revealed lymphoplasmacytic infiltration of the leptomeninges and underlying brain. The infiltrating lymphocytes and plasma cells were diffusely distributed, but some pseudofollicular architecture was noted. The inflammatory infiltrate also contained large amounts of pale eosinophilic cytoplasm and one or more large ovoid nuclei. Emperipolysis with lymphoid cells were observed in some of these cells. Many of the larger cells stained positive on immunohistochemistry studies for S100.
and CD68, and showed weak positivity for CD1. Positivity for CD1 was confirmed by studies with peanut agglutinin, which stained the cells strongly. Immunohistochemical staining for Ki67, a known marker of cellular proliferation, highlighted a moderate proportion of the nuclei of the all cells within the lesion. All the features mentioned above were with Langerhans' cell histiocytosis.

A pediatric hematologist consulted the patient during her postoperative course, and no systemic findings of the disease were detected. Adjuvant therapy was not considered. At 10 months post-surgery, she showed no clinical or radiographic evidence of recurrence (Figures 2a, b, c).

**DISCUSSION**

LCH is a disease characterized by abnormal proliferation of histiocytes in one or many organs.

---

Figures 2a and 2b: At the tenth postoperative month, T1- and T2- weighted axial MR images, respectively; 2c: sagittal MR image revealed no recurrence.
The skeletal system, skin, lung, liver, CNS, spleen, reticuloendothelial system, and gastrointestinal tract are the sites most often involved (17). Although the cause of this illness is not clear, immunological abnormalities such as decreased suppressor T-cell activity, increased immunoglobulin synthesis, and decreased thymic function have been documented in affected individuals (12,14,15). Several investigators have detected clonal CD1a+ histiocytes in all the LCH lesions they tested, which suggests that the condition may be a clonal neoplastic disorder with highly variable biological behavior and clinical severity (21). Recent reviews have proposed further stratification of the clinical form of the disease. One proposed system is based on extent of disease, with categories listed as “restricted versus extensive, with or without organ dysfunction” (2). A second is based on location, with categories of “skin isolated, monostotic, poliostotic, and multisystem disease” (19).

The most common intracranial site of LCH involvement is the hypothalamus-pituitary axis, and the clinical presentation in these cases is the triad of diabetes insipidus, exophthalmus, and a lytic skull lesion (6,9). The second most frequent intracranial site is the cerebellopontine pathway (6).

Imaging material from an LCH-CNS study done on 38 patients in 1997 illustrates the wide array of parenchymal and extraparenchymal lesions that can occur within the CNS (5). Most of the cases (87%) exhibited more than one type of lesion. The MRI appearances of the lesions were not specific for LCH, and the diagnoses could not be made without associated clinical features. Since these lesions can be located in the gray or white matter, and they may or may not show contrast enhancement, the list of differential diagnoses for LCH is large. It includes such diverse pathologies as multiple sclerosis, acute disseminated encephalomyelitis, leukodystrophy, infection, metastasis, sarcoidosis, infarction, and neoplasia (6). As mentioned above, the finding of a solitary space-occupying LCH lesion in the CNS that does not involve the hypothalamus, meninges, or cranial vault is very rare. To our knowledge, only 14 such cases have been reported in the literature (1,3,7,8,10,11,13,18). Most of these lesions were in frontal, temporal, choroidal, parieto-occipital, parietal, and insular locations, and the majority of patients had no other systemic findings.

From a surgical point of view, pure solitary non-hypothalamic LCH-CNS lesions appear to be a distinct form that can be totally excised. As a result, the prognosis is favorable. Recent reports of cases with follow-up periods as long as 17 years have noted success with surgical excision alone, and radiotherapy and chemotherapy have only been advocated in situations where subtotal resection was performed (4,7,13). Among the reports in the literature, only one describes local recurrence and one describes a fatal outcome (16,18).

In conclusion, there are too few documented cases of solitary non-hypothalamic LCH-CNS lesions to make definitive decisions about adjuvant therapy; however, the outcomes in our case and others suggest that surgical excision with close follow-up is a safe and adequate treatment.

Correspondence: Prof. Dr. Emin Özyurt Cerrahpasa Tip Fakültesi Beyin Cerrahisi ABD P.K. 4, 34301 Cerrahpasa İstanbul, Turkey E-mail: ahmethilmikaya@yahoo.com Telephone Number: 0090-212-6320026

REFERENCES


Eosinophilic granuloma. A different behaviour in children than in adults.

**Plasschaert F, Craig C, Bell R, Cole WG, Wunder JS, Alman BA.**

Localised Langerhans-cell histiocytosis of bone (eosinophilic granuloma) is a benign tumour-like condition with a variable clinical course. Different forms of treatment have been reported to give satisfactory results. No recurrences were noted in the skeletally immature group even after biopsy alone. By contrast, four of 13 skeletally mature patients had a recurrence and required further surgery. This suggests that eosinophilic granuloma has a low rate of recurrence in skeletally immature patients.