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LHERMITTE-DUCLOS DISEASE RECOVERY AFTER SUBTOTAL RESECTION; CASE REPORT

Lhermitte-Duclos Hastalığının Subtotal Rezeksiyondan Sonra İyileşmesi; Vaka Raporu

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ABSTRACT

Lhermitte-Duclos Disease (LDD) is a rare benign hamartomatous disorder that typically manifests unilaterally in the cerebellum, leading to thickening of the molecular and granular cell layers of the cerebellar cortex. It most commonly occurs in young adulthood, typically in the third or fourth decade of life. Since LDD is a slowly progressive disease, conservative management is an option in the absence of mass effect, whereas total excision is recommended in symptomatic patients. In our case, subtotal resection was performed due to indistinct surgical margins observed intraoperatively. Postoperative cranial magnetic resonance imaging (MRI) demonstrated regression of the lesion. No recurrence or residual tumor was detected at the 7-month follow-up. During this period, the patient remained neurologically stable without newly developed symptoms or deficits. However, long-term follow-up is essential to monitor for potential recurrence.

Keywords: Cerebellum, Lhermitte Duclos, Subtotal Resection

ÖZET

Lhermitte-Duclos Hastalığı (LDH) genç erişkinlikte, genellikle 3-4. dekatta çok nadir görülen tipik olarak serebellumu tek taraflı tutan, serebellar korteksin moleküler ve granüler hücre katmanlarında kalınlaşma gösteren iyi huylu hamartomatöz bir hastalıktır. LDH yavaş ilerleyen bir hastalık olduğundan kitleye ait bulgu yoksa konservatif olarak tedavi edilebilirken, kitle etkisi olan hastalarda total eksizyon önerilmesine rağmen olgumuzda ameliyat sırasında bu ayrım net olarak yapılamadığı için subtotal rezeksiyon yapıldı. Postoperatif kontrolünde kraniyal manyetik rezonans (MRG) incelemesinde lezyonun gerilediği görüldü. Hastanın postoperatif 7. ay kontrollerinde nüks veya rezidü izlenmedi. Hastanın 7 aylık takibinde yeni gelişen nörolojik semptom veya defisit yoktu. Ancak uzun dönem takiplerde nüks açısından hastalar yakın takip edilmelidir.

Anahtar Kelimeler: Lhermitte Duclos, Serebellum, Subtotal Rezeksiyon

INTRODUCTION

Lhermitte-Duclos Disease (LDD) was first described by Lhermitte and Duclos in 1920 (Lhermitte & Duclos, 1920). It is a rare disorder primarily affecting young adults, most commonly occurring in the third or fourth decade of life. Although its exact pathogenesis remains unclear, LDD is classified as a benign hamartomatous lesion that typically involves the cerebellum unilaterally, leading to thickening of the molecular and granular cell layers of the cerebellar cortex and exhibiting a slow growth pattern (Carter et al., 1989; Yeşildağ et al., 2005). While LDD is generally unilateral, (Khandpur et al., 2019). Reported a case associated with Cowden syndrome that demonstrated bilateral cerebellar involvement (Khandpur et al., 2019).

CASE REPORT

A 10-year-old male patient presented to our clinic with a two-month history of headache, vomiting, and imbalance. Neurological examination revealed a Glasgow Coma Scale (GCS) score of 15, intact cranial nerve function, and the presence of right-sided ataxia and dysmetria. Cranial MRI demonstrated a lesion originating from the right cerebellar hemisphere with minimal extension into the left cerebellum, causing slight compression of the fourth ventricle. The lesion measured 56×48 mm, appeared hypointense on T1-weighted images, and showed no hyperintensity on T2-FLAIR or contrast enhancement. LDD was suspected based on imaging findings (Figure 1).

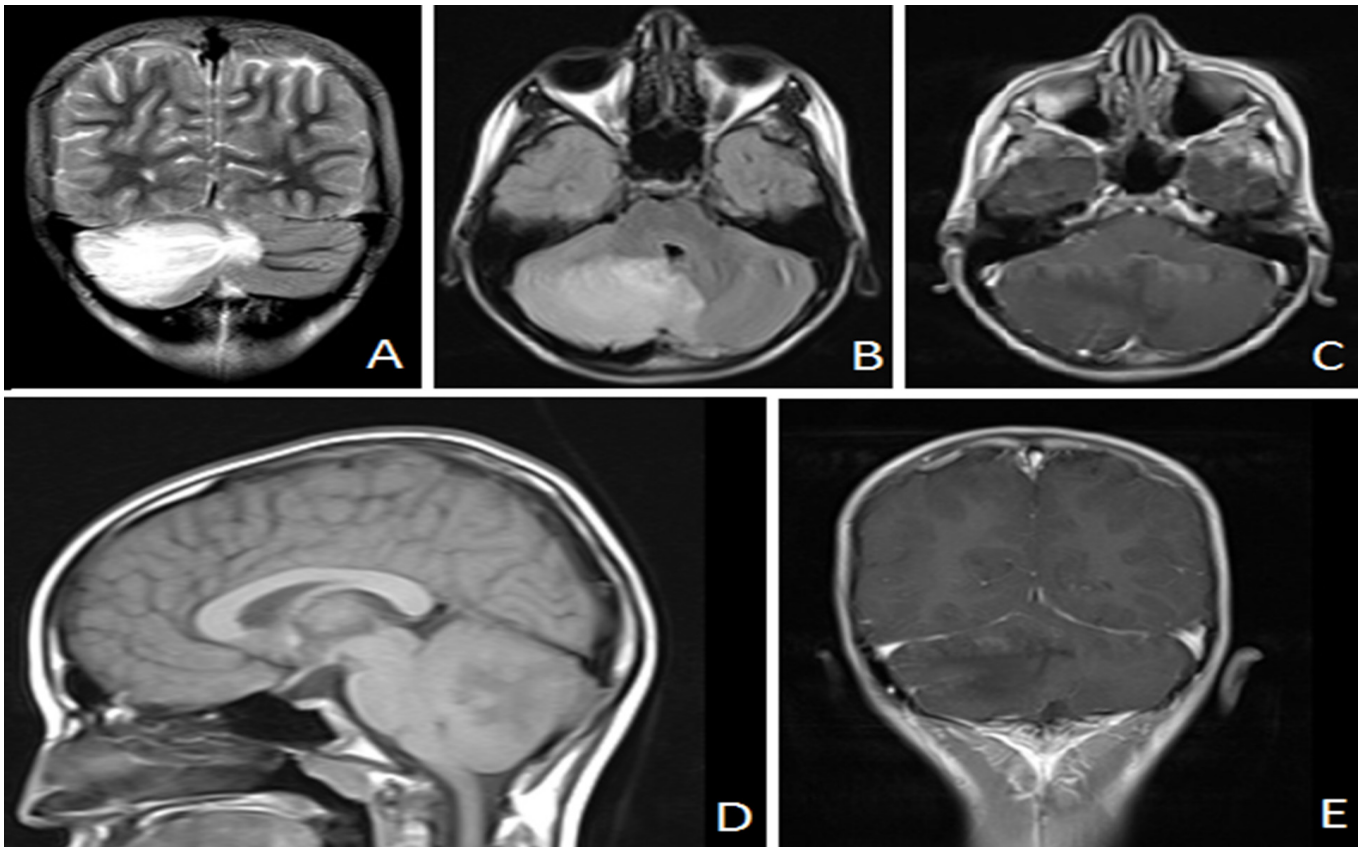

Figure 1: Patient's MR screen A: Coronal plane (T2 Weighted) B: Axial plane (T2 FLAIR) C: Axial plane (Contrasty) D; Sagittal plane (T1 Weighted) E: Coranal plane (Contrasty)

Due to the patient's clinical and radiological findings, surgical intervention was planned. Written informed consent was obtained from the patient's legal guardian regarding the surgical procedure and the potential use of clinical data in academic research. The patient underwent a right suboccipital craniectomy in the sitting position. The dura was opened according to standard surgical protocol, and the mass was subtotally resected due to its indistinct borders from the surrounding cerebellar tissue. No intraoperative complications were encountered. Postoperative cranial computed tomography (CT) findings are shown in Figure 2.

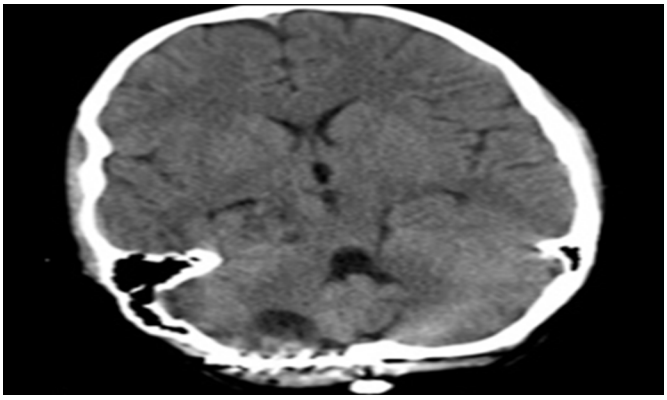

Figure 2: Cranial CT examination taken within 24 hours postoperatively.

Postoperatively, the patient exhibited improvement in symptoms, with a reduction in headache, ataxia, and dysmetria. No signs of increased intracranial pressure were observed during follow-up. Pathological examination confirmed a hamartomatous cerebellar lesion (Figure 3). As the patient's clinical symptoms did not regress further, he was discharged with a recommendation for close follow-up.

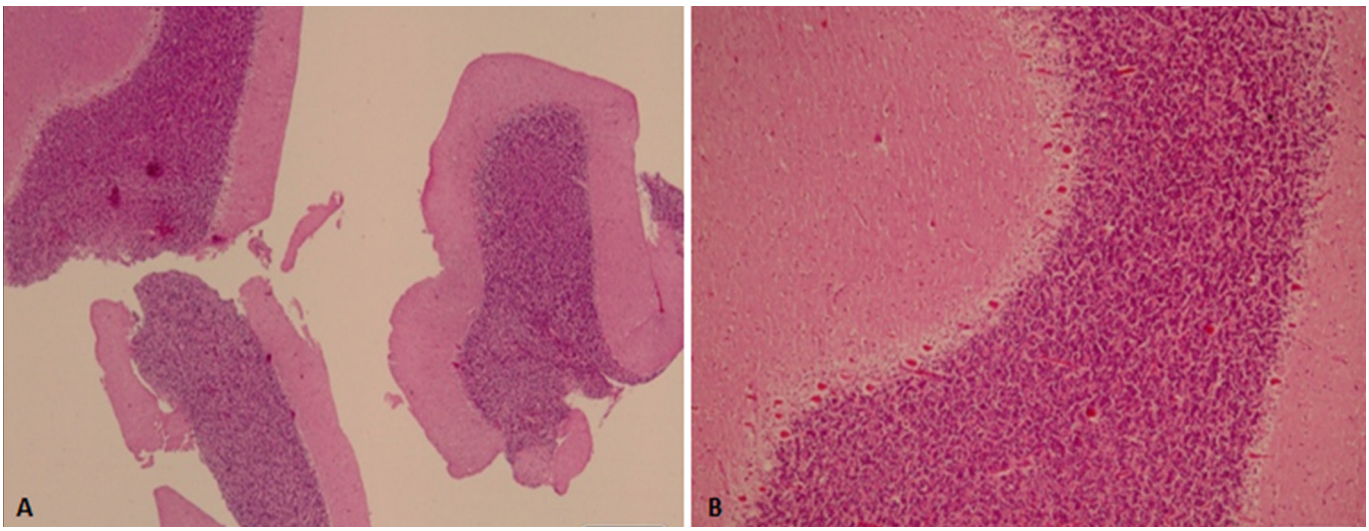

Figure 3: Widespread expansion appears in the internal granular layer and the molecular.

At the one-month postoperative follow-up, the patient's headache and vomiting had resolved, and his neurological examination was normal, with marked improvement in ataxia and dysmetria. Cranial MRI at this time revealed a residual lesion measuring 23×16 mm in the right cerebellar hemisphere (Figure 4).

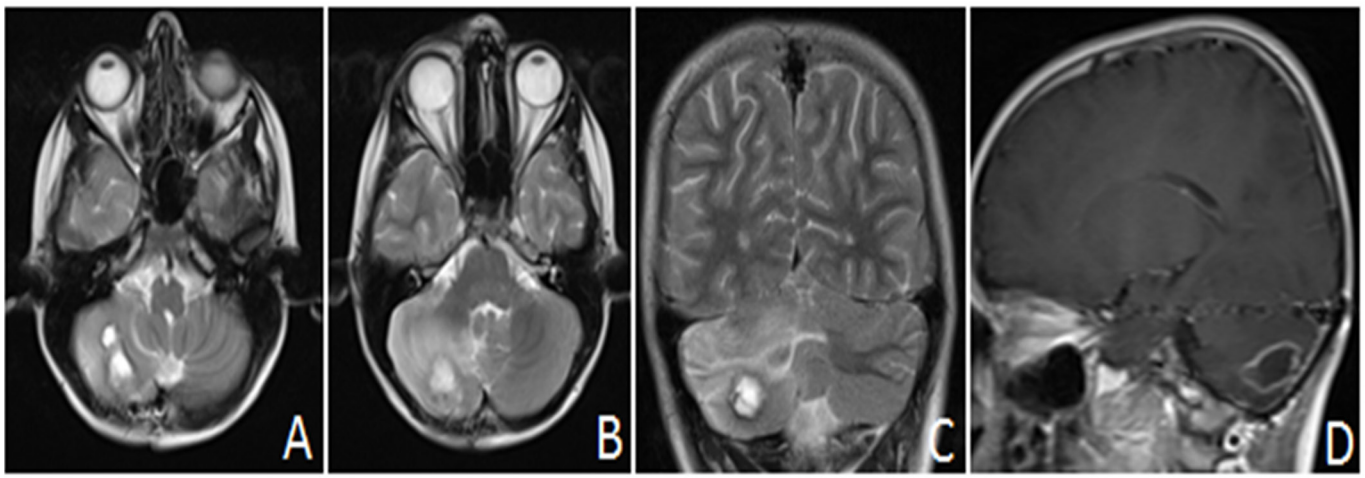

Figure 4: A-B-C; T2 MR images in the axial and coronal plane at the 1st month of postoperatif, D ;contrast MRI imaging in the sagittal plane.

Given the patient's clinical improvement, a conservative approach was adopted. Monthly clinical follow-ups revealed no new neurological deficits. By the seventh postoperative month, cranial MRI demonstrated complete regression of the lesion, with no evidence of residual mass (Figure 5).

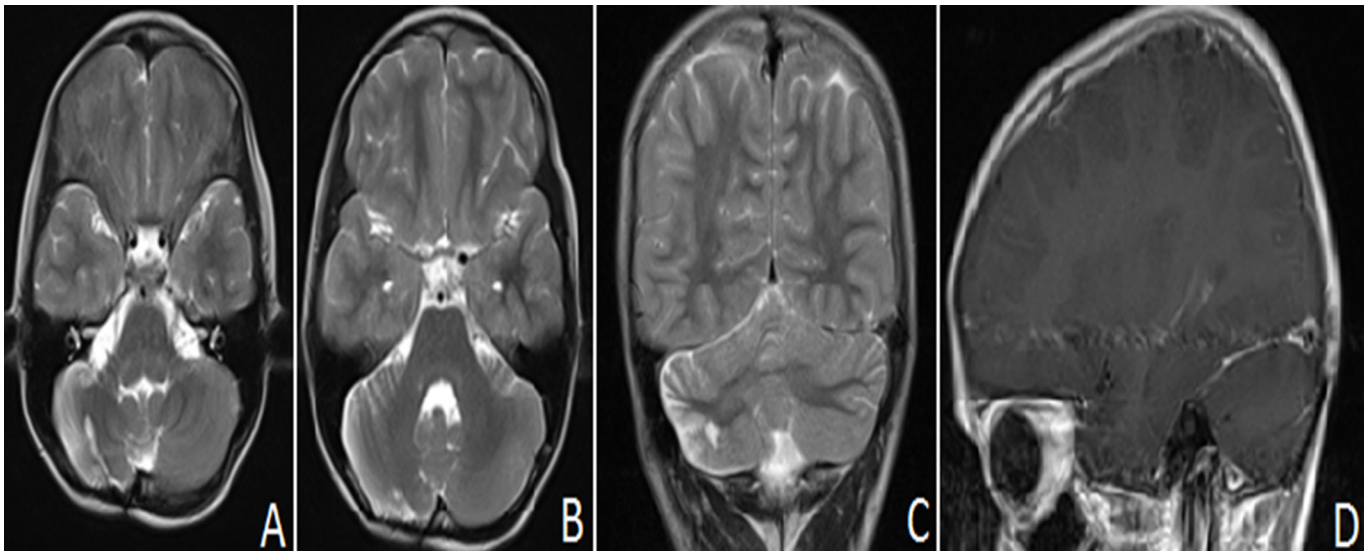

Figure 5: Control cranial MR examination of the case at the postoperative 7th month: A-B-C; T2-weighted sequence D; image in contrast sagittal plan.

DISCUSSION

LDD is a rare, hamartomatous lesion of cerebellar tissue. Although it is usually a lesion characterized by unilateral location, bilateral cases have also been reported in the literature (Khandpur et al., 2019; Borni et al., 2019). The disease typically manifests in adulthood, particularly in the 30s and 40s, and does not show a difference in prevalence between males and females (Nowak et al., 2001; Klisch et al., 2001). The pathology of this lesion, which is generally considered benign, has not been fully elucidated (Yesildag et al., 2005). Its coexistence with Cowden syndrome with mucocutaneous lesions and multiple hamartomas has also been described (Khandpur et al., 2019; Akiyama et al., 2006). We present a 10-year-old boy patient with no pathological findings to suggest Cowden syndrome. The diagnosis of many patients can be made by preoperative MRI examination (Xu et al., 2005; Haris et al., 2008). In MRI examination, it is seen as a hypointense in T1 sequences, hyperintense in T2, a lesion in which there is no contrast enhancement. The mass with parallel linear streaking without serious contrast enhancement is an important radiological finding of the disease. The presence of contrast involvement does not rule out the diagnosis (Akiyama et al., 2006). Depending on the edema effect of the mass; cereballar dysfunction may be presented with signs of obstructive hydrocephalus and increased intracranial pressure as a result of 4th ventricular compression (Yesildag et al., 2005; Berkman et al., 1998). In the cranial MRI examination of our case, in the right cerebellar hemisphere; There was a mass lesion with no contrast enhancement, with linear parallel streaks, with edema effect. The patient had mass-related headache and cerebellar dysfunction findings. LDD is rarely encountered in posterior fossa lesions. Since the differential diagnosis is in the pediatric age group, it should be differentiated from posterior fossa lesions such as medulloblastoma, pleocytic astrocytoma and hemangioblastoma (Chen et al., 2002). Since LDD is a slowly progressing disease, if there are no mass-related findings, it can be treated conservatively, while total excision is recommended in patients with mass effect (Nowak et al., 2001; Berkman et al., 1998). However, complications can be seen after total resection due to the fact that mass tissue cannot be clearly distinguished from cerebellar normal tissue. In some cases reported in the literature, since this distinction could not be made clearly, cerebellar dentate nucleus damage due to wide resection or postoperative cerebral mutism

due to vasospasm was observed (Khandpur et al., 2019). To prevent this situation, resection and high resolution fiber tractography with intraoperative MRI are recommended (Cheng et al., 2019; Fernandes-Cabral et al., 2016; Afshar-Oromieh et al., 2010). (Wang et al., 2010) published a series of patients who underwent subtotal resection. In this series, 9 of 12 patients reported that obstructive hydrocephalus findings improved in patients after subtotal resection. Since this distinction could not be made clearly during the operation of our case, resection was performed subtotally. Since there was no worsening in clinical findings in the postoperative period, clinical monitoring was taken. At the 7th-month follow-up examination, the patient's right-sided ataxia had resolved. No additional pathology was detected in the cranial system, cerebellar examination, motor, or sensory examination. Follow-up cranial MRI revealed regression of the lesion. No additional pathology requiring surgical intervention was identified. Although LDD is classified as WHO Grade 1 tumor, there is relapse case report in the literature (Banerjee & Gleadhill 1979). In another relapse case, it was reported that the patient was administered temozolomide therapy (Khandpur et al., 2019). Chemotherapeutic treatment was not applied to our case. LDD is presented because it is a rare disease that should be considered in the differential diagnosis of posterior fossa lesions of the pediatric patient group. The clinical findings of the patient may improve after subtotal resection. However, in terms of recurrence, strict following of the patient is required.

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