Epilepsy with Giant Interhemispheric Lipoma, Callosal Agenesis and Cortical Dysplasia Complex: Case Report

Epilepsiyle Birlikte Dev İnterhemisferik Lipom, Kallozal Agenezi ve Kortikal Displazi Kompleksi: Olgu Sunumu

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ABSTRACT Intracranial lipomas are rare conditions. Most lesions are small and found incidentally during neuroradiological investigations. Also, they are usually incidental findings in patients with epilepsy. In this report, we present a patient in whom epilepsy was the main clinical manifestation of interhemispheric lipoma, associated with agenesis of the corpus callosum and bilateral mesial frontal cortical displasia as shown by magnetic resonance (MR) imaging. The semiological and video-EEG monitoring findings were consistent with the left hemispheric involvement. Mass effect, associated cortical abnormalities and callosal agenesis could be responsible for epileptogenesis.

Key Words: Lipoma; epilepsy

ÖZET İntrakraniyal lipomlar nadir görülürler. Çoğu zaman küçük boyutlarda olup, nöroradyolojik incelemeler sırasında rastlantısal olarak tespit edilirler. Epilepsili hastalarda da sıklıkla tesadüfen tespit edilen lezyonlardır. Sunulan olguda; epilepsi, interhemisferik lipomun temel klinik bulgusu olup, lezyonla birlikte manyetik rezonans (MR) görüntüleme ile ortaya konan korpus kallozum agenezisi ve bilateral mezial frontal kortikal displazi bulunmaktadır. Semiyoloji ve video-EEG görüntüleme bulguları sol hemisferik tutulumla uyumlu bulunmuş, kitle etkisi ve eşlik eden korpus kallozum agenezisi ile bilateral mezial frontal kortikal anomalilerin epilepsi gelişimden sorumlu olabileceği düşünülmüştür.

Anahtar Kelimeler: Lipom; epilepsi

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Intracranial lipomas are rare conditions, with an incidence of 0.1-1% of all intracranial tumors.[1-6] Most lesions are small and found incidentally during neuroradiological investigations.[6] In symptomatic cases, clinical features are consist of seizures, headache, developmental delay, hemiparesis, cranial nerve defects and hydrocephalus.[7] The most common location is the pericallosal cistern, accounting for about 50% of cases, and they also can be located in the quadrigeminal/superior cerebellar cistern, suprasellar/interpeduncular cistern, cerebellopontine angle and sylvian cistern.[8,9] Intracranial lipomas are often associated with various anomalies of the central nervous system, such as agenesis/dysgenesis of the corpus callosum, absence of septum pellucidum, cranium bifidum, spina bifida, encephalocele, myelomeningocele, hypoplasia of vermis, and malformation of the cortex.[9]

In this report, we present a patient in whom epilepsy was the main clinical manifestation of interhemispheric lipoma, associated with agenesis of the corpus callosum and bilateral mesial frontal cortical displasia as shown by magnetic resonance (MR) imaging.

CASE REPORT

A 25-year-old, left-handed male patient was admitted with headache with increasing seizure frequency recently. His seizures started at the age of 12 years. The patient was born after a normal full term pregnancy and psychomotor development was slow. His sister had presented one febrile seizure but otherwise normal and there was no consanguinity. Neurologic examination revealed slight right lower limb monoparesis and positive Babinski sign. Psychiatric status was normal, although with a moderately limited intellect, which was not measured. He had been treated with oxcarbazepine 900 mg/day, but he was not taking his pills regularly.

According to 2010 ILAE seizure classification,[10] a focal seizure originated from the left hemisphere was recorded by ictal video-EEG monitoring. The seizure characteristics were right hemifacial and upper limb tonic then right hemiclonic convulsions with loss of consciousness.[11] The ictal EEG was consistent with the left anterior temporal focal and left hemispheric epileptogenic seizure pattern.

MRI findings revealed a giant interhemispheric lipoma. There was no corpus callosum (Figure 1A). The cortices nearby the lipoma on both sides were dysplastic appearing pachygyric and thickened (Figure 1C). Branches of both anterior cerebral arteries were tortuous and traversing through the lipoma (Figure 1B, D). Findings all were consistent with a giant callosal lipoma associated with callosal agenesis and cortical dysplasia.

DISCUSSION

Intracranial lipomas are congenital malformations believed to result from abnormal persistence and maldifferentiation of meninx primitive during the development of the subarachnoid cisterns.[9,12] This theory explains the characteristic subarachnoid and cisternal locations.

Approximately, 50% of intracranial lipoma cases were found to be asymptomatic in a review of 85 cases.[4] In the remained cases, seizures were reported as the most common presenting symptoms. Hemispheric lipomas are more likely to be symptomatic than lipomas in the midline[13]. However, the frequency of epilepsy in callosal lipoma cases were found as high as 82% in a previously report.[14] The nature of the seizures in patients with lipomas of the corpus callosum were firstly analyzed in a study from 1980.[15] In this study, Gastaut et al. reported four patients with seizures and lipomas of the corpus callosum. However, clinical and EEG characteristics of three patients pointed out the temporal lobe epilepsy. In a recent study, among the 3500 consecutive patients who underwent video-EEG monitoring, just five patients with

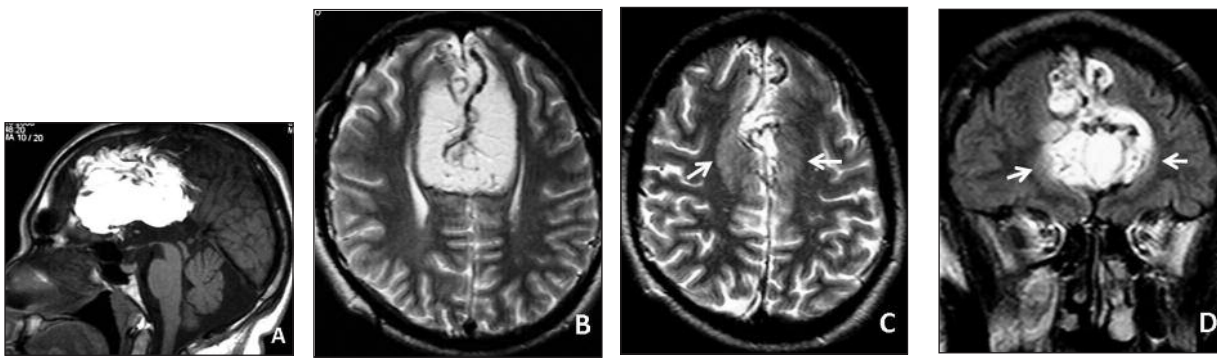

FIGURE 1A-D: Sagital T1-weighted image (A) shows callosal lipoma with callosal agenesis. Axial T2-weighted image (B) reveals bilateral tortuous anterior cerebral arteries traversing through the lipoma and lateral ventricles parallel to each other. Axial T2-weighted image (C) above the lipoma shows pachygyria on mesial aspects of frontal lobes (arrows). Coronal FLAIR image (D) again shows tortuous branches of anterior cerebral arteries embedded inside the interhemispheric lipoma and smooth surface of nearby cortices (arrows). Findings all are compatible with callosal lipoma associated with callosal agenesis and cortical dysplasia.

intracranial lipomas were identified[7]. In these five patients, one patient's lesion was thought to be responsible for the seizures. In a pediatric intracranial lipoma case series of 20 patients, only one patient was examined because of epilepsy.[16] According to recent data, the incidence of epilepsy seems to be much lower than that claimed in earlier reports, and intracranial lipomas are usually incidental findings in patients with epilepsy. No detailed information is also found in the literature about the semiological and video-EEG findings from such patients.

Pathophysiology of the seizures in patients with intracranial lipoma could be explained by various mechanisms, such as a mass effect, associated cortical abnormalities and callosal agenesis. Zettner and Netsky proposed that explanation of the seizures was the infiltration of the adjacent cortical surface by the thick capsule of the lipoma mass.[14] Ultrastructurally, along the border between the thick connective tissue of the lipoma and cortical surface, disruption of the basal lamina, prominent astrocytosis, and abundant axonal and synaptic profiles were observed.[17] There are also reports of cortical abnormalities, such as polymicrogyria, pachygyria, subcortical nodular heterotopia associated with cerebral lipoma.[17] Development of the lipoma was suggested to contribute to the formation of the focal cortical dysplasia because of physical interruption and focal perfusion insufficiency.[18] Interhemispheric lipomas are almost associated with hypogenesis or agenesis of the corpus callosum. Persistence and maldifferantiation of the meninx and formation of the lipoma are thought to interfere with genesis of the callosal fibers.[9] This interhemispheric disconnection is responsible for a facilitatory and disinhibitory action that favors the occurrence of seizures originated from the lesion.[13]

In our case, a giant interhemispheric lipoma accompanied with pachygyria of the bilateral was mesial frontal lobes and callosal agenesis. Nonetheless, the semiological and video-EEG monitoring findings were consistent with the left hemispheric involvement, and all aforementioned mechanisms could be responsible for epileptogenesis in our patient. We did not consider surgery as a choice of therapy due to the vascular structures embedded within. In addition, the mechanisms of epilepsy probably involved not only vascular but also bilateral cortical malformation. Although the abnormality persisted, the seizures stopped on 1200 mg/day oxcarbazepine treatment in 1-year follow up.

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