

Pericallosal Lipomas: Aetiology of Symptomatic Epilepsy or Incidental Thalamic Involvement with Idiopathic Generalized Epilepsy?: Two Case Reports

Perikallosal Lipomalar:

Semptomatik Epilepsi Nedeni mi Yoksa İdyopatik Jeneralize Epilepsiyle
İnsidental Talamik Tutulum mu?: İki Olgu Sunumu

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Summary

Intracranial lipomas are regarded as congenital malformations of the meninx primitiva. Here we present two patients with pericallosal lipomas extending to the thalamus. They had generalized tonic-clonic seizures. Electroencephalograms showed generalized spike-wave discharges. It is known that the thalamus is important in the formation of sleep spindles and spike-wave discharges. Thalamic involvement by lipomas may play a role in the generation of generalized spike-wave discharges and epileptogenesis in these patients.

Key words: Lipoma; seizures; spike-wave discharges; thalamus.

Özet

İntrakranial lipomaların meninks primitivanın doğumsal malformasyonları olduğu düşünülmektedir. Bu yazıda, jeneralize tonik klonik nöbetleri olup elektroensefalogramlarında jeneralize diken dalga deşarjları gösteren ve talamusa uzanım gösteren perikallosal lipomalı iki olgu sunuldu. Talamusun uyku içciklerinin ve diken dalga deşarjlarının oluşmasında rolü olduğu iyi bilinmektedir. Talamik tutulum gösteren lipomalarda da jeneralize diken dalga deşarjlarının oluşmasındaki ve epileptogenezdeki olası rolü değerlendirilmiştir.

Anahtar sözcükler: Lipoma; nöbet; diken dalga deşarjı; talamus.

Introduction

Intracranial lipomas are regarded as congenital malformations. They account for 1% of all intracranial tumors.^[1] Maldevelopment of the meninx primitiva at the lamina terminalis appears to be the cause.^[2] They are mostly located in midline (pericallosal) regions, but other locations have also been reported, including the quadrigeminal/superior cerebellar, suprasellar/interpeduncular, cerebellopontine angle, and

Sylvian cisterns.^[1,2] Lipomas are generally asymptomatic but headaches, convulsions, and cranial nerve palsies have been reported in patients with intracranial lipomas.^[1,3] Thalamus, especially the thalamic reticular nucleus (nRT), was frequently responsible for the formation of generalized spike-wave discharges (SWDs).^[4] Involvement of the posterior inferior part of the thalamus is responsible for the generation of SWDs, instead of the medial anterior region, including thalamocortical cells and surrounding reticular nucleus.^[5]

Submitted (Geliş): 31.10.2013

Accepted (Kabul): 24.02.2014

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Here we present two patients with generalized seizures including absence seizures and generalized 3-Hz SWDs on electroencephalograms (EEG) in which electrophysiological and clinical features may be misrelated as the result of thalamic involvement by pericallosal lipomas rather than incidental co-existence.

Case Report

Case 1– A 21-year-old female patient was referred to our center in 1996 for intractable seizures and epilepsy surgery. She had had febrile convulsions when she was 6 months old and 4 febrile seizures had occurred by the age of 4 years. Recurrent seizures had started at age 16. Her neurological and physical examinations were normal. She and her mother reported two types of seizures: short episodes of unconsciousness and staring occurring daily and generalized tonic-clonic seizures (GTCSs) that lasted 1-2 minutes, 1-4 times per year despite high-dose carbamazepine (CBZ) therapy.

Routine EEGs were either normal or showed nonspecific slow waves. Her sleep-deprived EEG was recorded in our outpatient EEG laboratory and 3-Hz SWDs with absence seizures were seen (Figure 1a). Magnetic resonance imaging (MRI) revealed pericallosal lipoma (1x1.5x3.5 cm) (Figure 1b). Her MR angiography was normal. The CBZ therapy was switched to valproic acid (VPA) for idiopathic generalized epilepsy based on the EEG findings and seizure types. Her absence seizures ceased but after the occurrence of a few GTCSs lamotrigine was added to the VPA. She became seizure-free and follow-up EEGs were normal after combined therapy. She was seizure-free during the 12-year follow-up under the combined therapy but drug withdrawal trials were unsuccessful. Follow-up MRI performed after 10 years showed that the lesion had not changed.

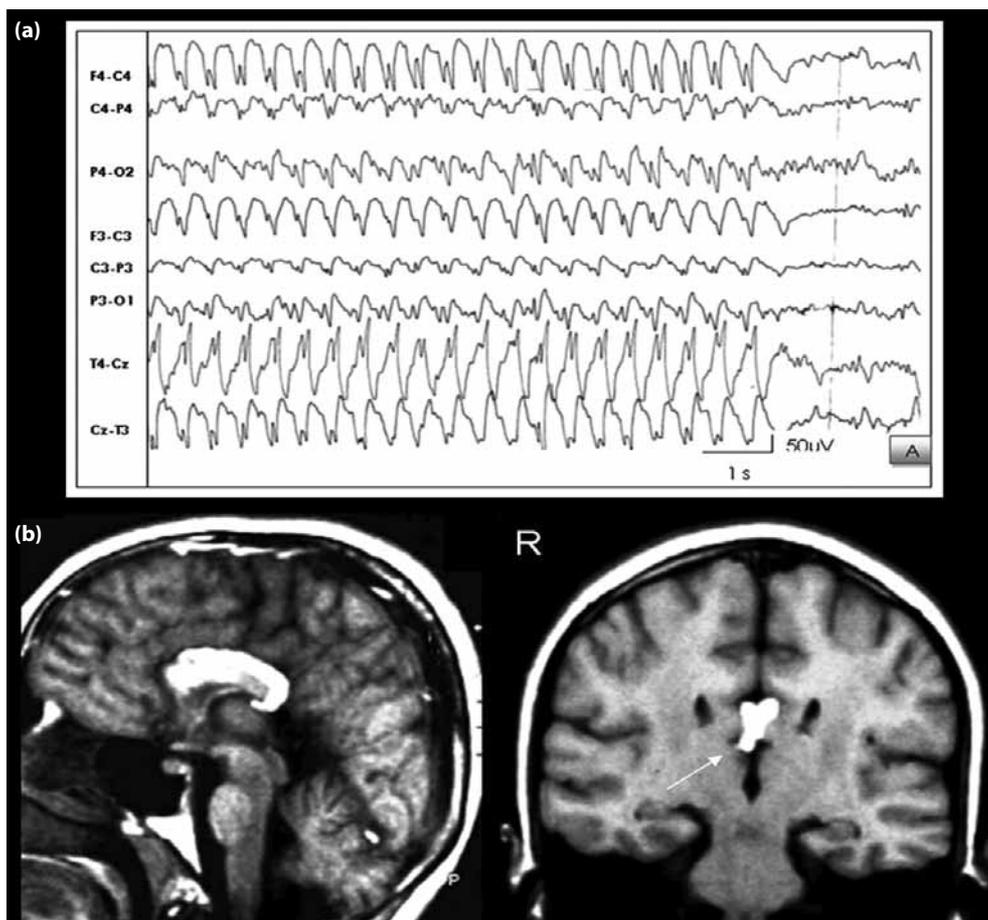


Fig. 1. (a) Generalized 3-Hz spike-wave discharges during absence seizures are seen in the routine EEG of Case 1. (b) Brain MRI (T1-weighted images with axial, coronal, sagittal sections) of Case 1 showing a pericallosal lipoma touching the right thalamus (arrow).

Case 2– An 18-year-old male diagnosed with partial epilepsy had been followed up by a pediatric neurologist for 5 years. Febrile convulsions were reported in his past medical history. In addition, the patient had suffered loss of consciousness as a result of a traffic accident at the age of 10 years. He had been hospitalized in the neurointensive care unit for 7 days. Recurrent seizures had started at age 13 as GTCs lasting 2 minutes. EEGs had shown slow waves in the posterior part of the right hemisphere. CBZ had been started and increased to a dosage of 600 mg/d. Bilateral synchronous SWDs had been reported in follow-up EEGs without seizure recurrence. After 4 years of treatment, his seizures recurred following a CBZ withdrawal trial.

When he was 18 years old, he was admitted to our clinic. He reported 2 GTCs in the previous year despite CBZ therapy. Routine and sleep-deprived EEGs were performed and generalized SWDs were seen (Figure 2a).

He had two cranial MRIs in 5 years and there were no encephalomalacic or sequelae lesions but there was a perical-

losal lipoma in the corpus callosum adjacent to the right thalamus located in the quadrigeminal cistern (Figure 2b). The patient was treated for idiopathic generalized epilepsy and CBZ therapy was switched to VPA recently. After VPA treatment he had one relapse.

Discussion

MRI scans show hyperintensity on T1-weighted images (T1WI) and hypointensity on long-TR images in patients with intracranial lipomas. Lipomas are isointense to hypointense on fast spin-echo (FSE)-T2WI and hyperintense on FLAIR.^[1] In the patients presented here, pericallosal lipomas were diagnosed based on brain MRIs and their MRIs showed that the lipomas extended to the thalamus. Intracranial lipomas have fibrous capsules and higher vascularity, and they are adherent to the cerebral tissue.^[3] It can be speculated that the normal functioning of the thalamus might be disrupted by pericallosal lipomas because of the pressure of buried fibrous capsules in the thalamus and abnormal vascularity in both cases. EEGs of our patients revealed generalized SWDs. Only four adult patients with intractable

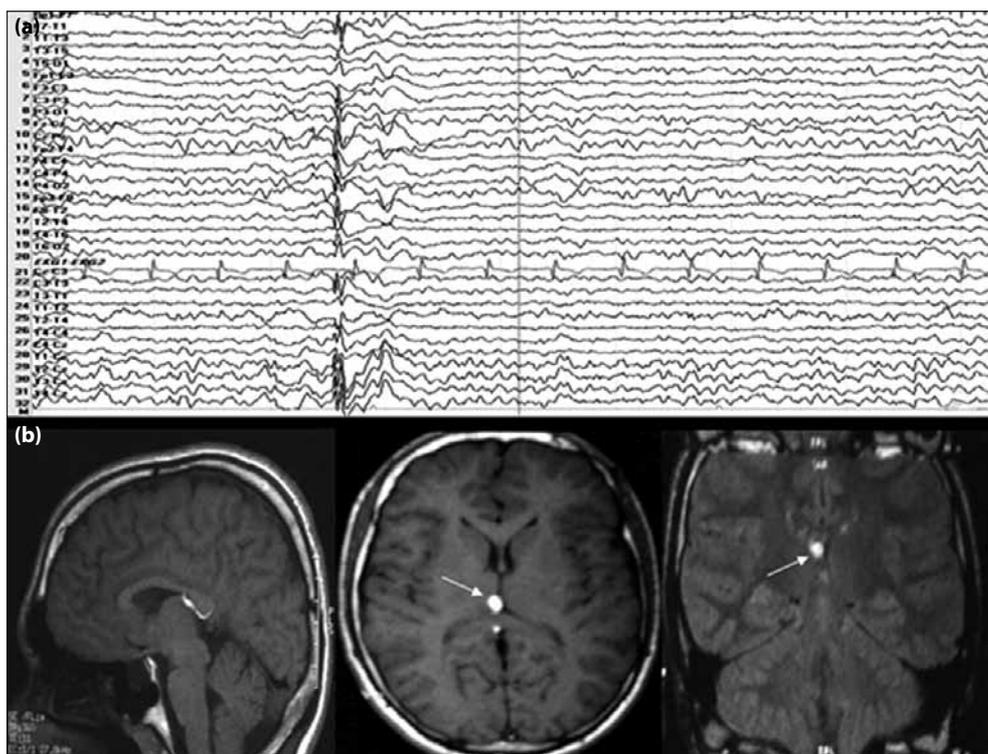


Fig. 2. (a) Generalized spike-wave discharges in the routine EEG of Case 2. (LF: 0.1 Hz, HF: 70 Hz). (b) Brain MRI (T1-weighted sagittal, axial, FLAIR sequences) of Case 2 showing the pericallosal lipoma reaching the right thalamus (arrows).

idiopathic generalized epilepsy (IGE) were reported due to unilateral thalamic lesions with generalized SWDs on EEG have been reported.^[4]

It is known that the thalamus plays a crucial role in the formation of sleep spindles and generalized 3-Hz SWDs.^[5] The thalamic reticular nucleus is mostly responsible for forming SWDs and sleep spindles by reciprocal excitatory and inhibitory activities between thalamocortical and nRT neurons.^[5] During sleep spindles and spike-wave seizures a burst of synchronized firing in thalamocortical neurons causes excitatory postsynaptic potentials (EPSPs) in nRT neurons and then a burst of synchronized firing of GABAergic nRT neurons causes inhibitory postsynaptic potentials (IPSPs) in thalamocortical neurons.^[5] There is a circuitry between the cortex and thalamus. Removal of the thalamus, cortex, or both eliminates SWDs.^[5] In our cases, although involvement of the posterior thalamus (not posterior-inferior) was documented in MRIs, EEGs showed that integrity of the cortico-thalamic circuitry was not disrupted physiologically. EEG and clinical findings were compatible with IGE. MRI lesions probably caused the misdiagnosis of symptomatic generalized epilepsy and treatment failure with CBZ. Surgical treatment generally fails and results in high morbidity and mortality because of the high vascularity and buried lipomas in the cortex.^[3]

Symptomatic generalised epilepsy can be misdiagnosed and inappropriate medication can be started in some cases because of the existence of a lesion. Antiepileptic treatment should be started according to epilepsy syndrome consid-

ering seizure types and EEG findings. Without considering MRI findings, case 1 can be diagnosed as juvenile absence syndrome. Lipomas can be found incidentally in some cases and we cannot rule out the possibility that our patients may have idiopathic generalized epilepsy and intracranial lipomas, which are incidentally found in MRIs.

Conclusion

To avoid giving improper medication and performing unnecessary surgery, a treatment plan should be drawn up according to epilepsy syndrome considering EEG findings and seizure types in patients with intracranial lipoma in whom lipomas may be an incidental finding or may accompany to IGE.

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