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CASE REPORT / OLGU SUNUMU

Late Remission of Seizures After Functional Hemispherotomy in a Child With Congenital Middle Cerebral Artery Infarct



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Doğuştan Arteria Serebri Media Enfarktı Olan Epileptik Hastada Nöbetlerin Fonksiyonel Hemisferotomi Sonrası Geç Dönem Remisyonu

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Summary

Hemispherotomy can be a useful procedure in hemispheric syndromes, such as Rasmussen's encephalitis, congenital stroke, or hemimegalencephaly. The seizures associated with these syndromes usually remit immediately after hemispherotomy. Presently described is a patient who had a functional hemispherotomy for a congenital middle cerebral artery stroke and whose seizures remitted 2½ years after the operation. His postoperative electroencephalogram showed contralateral abnormalities. However, these disappeared with the remission of seizures. One may argue that the disconnection of the abnormal hemisphere could result in the cessation of the epileptogenic potential of the healthy hemisphere by terminating secondary epileptogenesis.

Keywords: Electroencephalogram; epilepsy; hemispherotomy.

Özet

Hemisferotomi Rasmussen ensefaliti, doğuştan inme ve hemimegalensefali gibi hemisferik sendromlarda oldukça yararlı bir yöntemdir. Bu hastalıklar kaynaklı nöbetler genellikle hemisferotomi sonrası hemen dururlar. Bu yazıda, doğuştan arteria serebri media enfarktı kaynaklı nöbetleri olan, hemisferotomi operasyonu yapılan ve nöbetleri ameliyat sonrası 2.5 yıl sonra duran bir olgu sunuldu. Hastanın ameliyat sonrası elektroensefalografisi (EEG) kontralateral düzensizlikler içermekteydi. Nöbetlerin durması sonrası yapılan EEG'de kontralateral düzensizliklerin kaybolduğu görüldü. Bu olguda enfarkt sekelli hemisferin sağlıklı hemisferden ayrılmasının, sağlıklı hemisferin epileptojenik potansiyelini sekonder epileptogenesisi engelleyerek durdurduğu düşünülebilir.

Anahtar sözcükler: Elektroensefalografi; epilepsi; hemisferotomi.

Introduction

Congenital ischemic cerebral infarction leads to a loss of brain parenchyma and gliosis, which may cause drug-resistant epilepsy.[1] Presently described is the case of a 10-year-old boy with perinatal ischemic stroke and drug-resistant epilepsy who was treated with a functional hemispherotomy. His seizures subsided 2½ years after the operation, which is an unusually long period of time.

Case Report

The patient was a 10-year-old boy with left-sided hemiplegia and severe epilepsy, secondary to a congenital right middle cerebral artery infarction (Fig. 1). His seizures were characterized by left arm tonic activity lasting for only a few seconds, happening up to 100 times a day. During seizures, he occasionally experienced a brief loss of consciousness and bladder control. An interictal electroencephalogram

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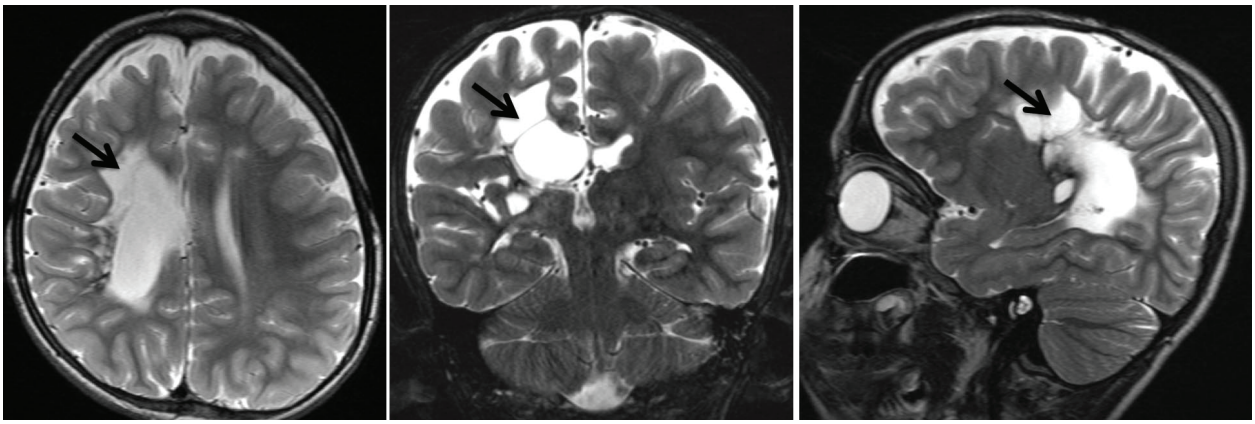


Fig. 1. Right hemisphere gliosis and cyst formation in the right middle cerebral artery territory. Black arrows indicate porencephalic cavity.

(EEG) showed bilateral epileptiform discharges more intense on the right side (Fig. 2). Ictal EEG was characterized by diffuse flattening of interictal activity on all electrodes for a few seconds (not shown). Fluorodeoxyglucose positron emission tomography (PET) was also performed (not shown) and did not indicate any metabolic abnormality on the opposite hemisphere.

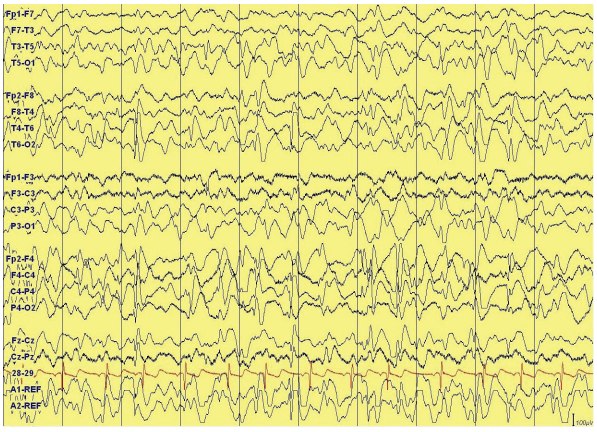


Fig. 2. Preoperative interictal EEG showed bilateral epileptiform discharges (right>left hemisphere).

Prior to surgery, the patient had been taking clobazam (10 mg+15 mg+15 mg), levetiracetam (3x500 mg), and topiramate (3x25 mg). This antiepileptic treatment had led to no improvement in his seizures. A right functional hemispherotomy was performed. Postoperative magnetic resonance imaging (MRI) showed complete disconnection of the right hemisphere (Fig. 3). However, his seizures continued with a similar frequency for 2½ years after the surgery, despite an increase in the doses of antiepileptic medications. Figure 4 shows EEG performed 2 years after surgery that was consistent with right hemispheric slowing and sharp waves, as well as left hemispheric, centroparietal-

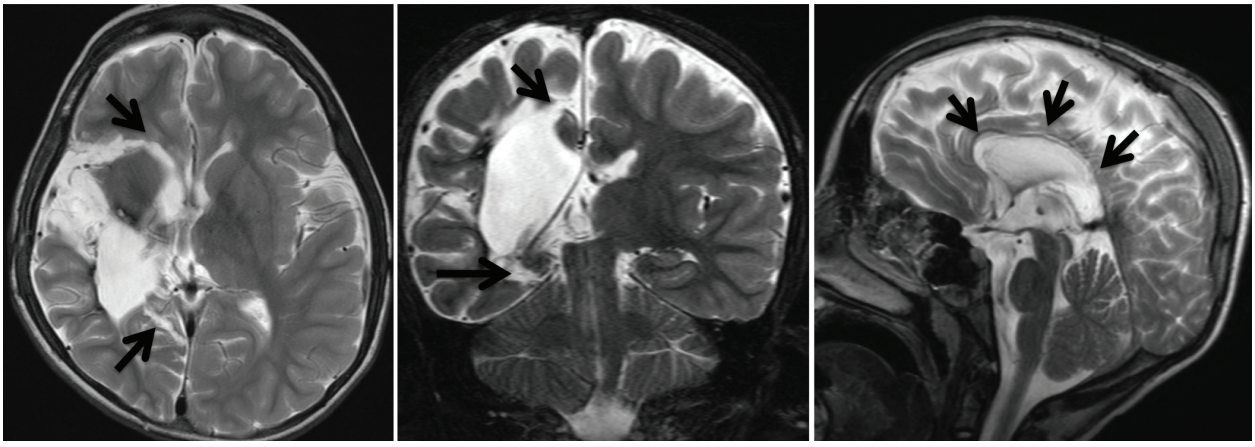


Fig. 3. Postoperative magnetic resonance image illustrating total disconnection of the right hemisphere. Black arrows show exact disconnection sites.

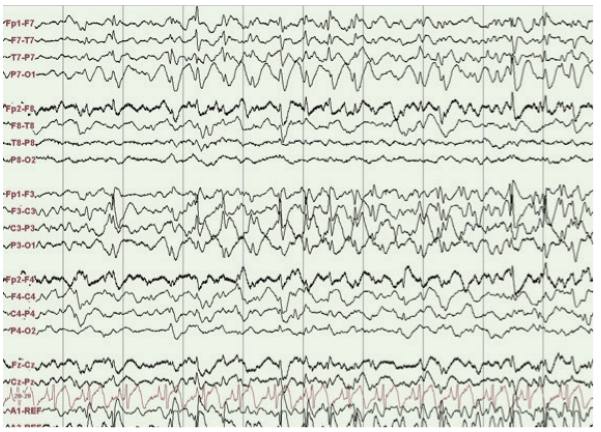


Fig. 4. EEG performed 2 years after surgery. Right hemisphere slowing and sharp waves can be seen, as well as left hemispheric, centroparietal-dominant, sharp waves occurring in chains.

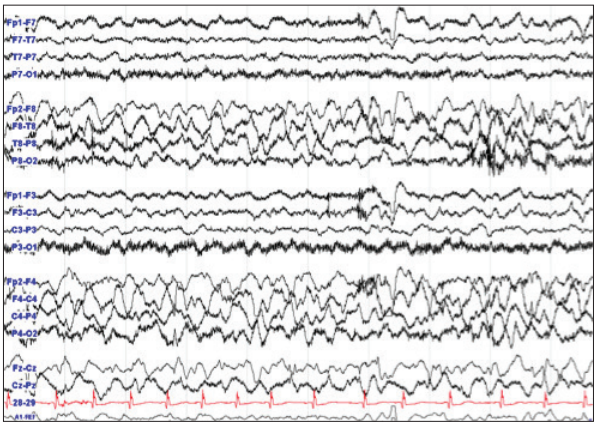


Fig. 5. EEG performed 3 years after surgery demonstrating no epileptiform activity in the left hemisphere, with right hemisphere slowing and sharp waves.

dominant, sharp waves repeating in chains. Two-and-a-half years later, however, his seizures stopped completely, and a repeat EEG on postoperative year 3 showed no epileptiform activity on the healthy side (left hemisphere) (Fig. 5).

Discussion

Perinatal cerebral artery occlusion is responsible for ischemic cerebral infarction leading to brain cavitation and gliosis. The resulting porencephalic cysts are frequently associated with hemiplegia and epilepsy, with 6% to 7% being refractory to medical treatment.[1]

Due to widespread damage produced by the ischemic insult, the epileptic focus can either correspond to the cystic lesion or be localized at a distant site. There may also be the possibility of diffuse foci. When this is the case, diffuse epileptic abnormalities are not necessarily an expression of independent epileptic activity, but may simply represent a secondary bilateral synchrony originating from the porencephaly itself. It may also represent the development of secondary epileptogenesis in the contralateral hemisphere.[2] Generalized interictal epileptiform abnormalities and generalized ictal patterns with no independent seizure discharge arising from the opposite hemisphere are not necessarily a contraindication to hemispherectomy in patients with vascular congenital hemiparesis.[3] A paradoxical lateralization of the EEG to the "good" hemisphere in children with unispheric encephaloclastic lesions is also described.[4] In some cases where there is good clinical, functional, and static imaging lateralization, particularly if associated with large encephaloclastic porencephaly, a review of the previous EEG history may be helpful in evaluating the significance of the contralateral findings.[5] In our patient, despite bilateral interictal EEG findings and non-localizable ictal EEG, clinical semiology and MRI findings suggested that he was a good candidate for hemispherotomy.

Contralateral epileptiform abnormalities may continue after hemispherotomy, but they usually diminish starting a year after surgery. In one study, seizures stopped despite the presence of contralateral interictal EEG abnormalities.[6] However, our patient's seizures continued in the presence of contralateral EEG abnormalities. Epileptic activity without clinical correlation may be observed in postoperative EEG's in areas of the brain that have been disconnected.[7] Mechanisms are unknown, but the generalized and contralateral epileptiform discharges may be manifestations of potentially reversible secondary epileptogenesis resulting from an interaction between the early lesion and the developing brain.[8,9]

Careful review of MRI can show subtle abnormalities in the contralateral hemisphere. These findings may include mild sulcal anomalies in patients with cortical dysplasia and mild white matter loss and signal abnormality in the hemisphere opposite the side with extensive cystic encephalomalacia due to perinatal infarction.[3] However, our patient had a normal opposite hemisphere on 3 Tesla MRI. The fluorodeoxyglucose PET performed before the surgery also revealed no abnormalities on the opposite hemisphere.

Residual non-disconnected tissue causing electroen-

cephalographic epileptic activity with corresponding clinical seizures may be observed in technically difficult hemispherectomies, such as those associated with hemimegalencephaly, or when there is not a full anatomic disconnection.[7] A study performed in children with hemimegalencephaly showed that cerebral blood flow of the opposite (normal) hemisphere was normal at birth; however, it increased in the third month and was only normal after hemispherotomy. In addition, the earlier the child was operated on, the more rapidly it normalized.[10] In this case, although one cannot be sure if total disconnection was achieved without a second-look operation, we may assume that it was complete as the seizures stopped eventually. We think that secondary epileptogenesis of the hemisphere was terminated with the total disconnection of the epileptogenic hemisphere. The reason for the seizures continuing for an unusually long period in this case could be the older age of our patient at the time of the operation. This may have allowed the healthy hemisphere to be bombarded by the epileptiform discharges from the contralateral hemisphere for a longer time period.

Informed Consent

Written informed consent was obtained from the patients family who participated in this study.

Peer-review

Externally peer-reviewed.

Conflict of Interest

None declared.

Authorship Contributions

Concept: U.I., M.M.Ö.; Design: U.I., M.M.Ö.; Supervision: M.M.Ö.; Materials: U.I., B.T., M.M.Ö.; Data collection &/or processing: U.I., B.T.; Analysis and/or interpretation: U.I., B.T., M.M.Ö.; Literature search: U.I.; Writing: U.I., B.T.; Critical review: B.T., M.M.Ö.

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