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Acquired Cerebral Hemiatrophy: Dyke-Davidoff-Masson Syndrome - A Case Report

Akkiz Serebral Hemiatrofi: Dyke-Davidoff-Masson Sendromu - Bir Olgu Sunumu

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ABSTRACT

A rare syndrome, Dyke-Davidoff-Masson Syndrome (DDMS), with a diagnostic conundrum, and the way it was solved is presented. A 13-year-old boy presented with recurrent seizures for the past 10 years. He had been treated with anticonvulsant medication which was satisfactory at first but later the seizures recurred. Recently, the frequency of the seizures increased with preictal dizziness and postictal drowsiness. Physical examination revealed mild left hemiparesis and left deviated gait irregularity. He was mentally alert but had not achieved all the developmental milestones as compared to normal child of his age. CT and MRI scan of the head showed hemiatrophic cerebral parenchyma with prominent sulci and encephalomalacia. 24-hour intensive video EEG monitoring revealed suppression of alpha rhythm and local slow wave activity on the side of the atrophic hemisphere. PET-CT showed highly functional left cerebral hemisphere and less functional right cerebral hemisphere. The patient underwent functional hemispherectomy under neurophysiological monitoring and the nonfunctional brain tissues were resected while selectively preserving the functional areas detected by fMRI and PET-CT scan. During follow up, the patient was seizure free as well as without difficulties in performing his daily activities and communications. Functional hemispherectomy for DDMS patient has a good prognosis.

KEYWORDS: Dyke-Davidoff-Masson syndome, Cerebral hemiatrophy, Seizure, Functional hemispherectomy

ÖZ

Nadir bir sendrom olan ve tanı koyması güç olan Dyke-Davidoff-Masson Sendromu (DDMS) ve tanı konma şekli sunulmaktadır. 13 yaşında bir erkek son 10 yıldır tekrarlayan havalelerle başvurdu. Antikonvülsan ilaçlarla tedavi başlangıçta tatminkar olmuş ama daha sonra havaleler tekrarlamıştı. Son zamanlarda havale sıklığı artmış ve preiktal başdönmesi ve postiktal uyku hali başlamıştı. Fizik muayene hafif sol hemiparezi ve sola deviasyonlu yürüme bozukluğu gösterdi. Mental olarak uyanıktı ama kendi yaşındaki normal bir çocuğun gelişimsel referans noktalarının tümüne ulaşmamıştı. Baş BT ve MRG taraması belirgin sulkuslar ve ensefalomalasi ile hemiatrofik serebral parankim gösterdi. Yapılan 24 saatlik yoğun video EEG izleme, atrofik hemisfer tarafında alfa ritim baskılanması ve yerel yavaş dalga aktivitesi ortaya çıkardı. PET-BT çok işlevsel bir sol serebral hemisfer ve daha az işlevsel bir sağ serebral hemisfer gösterdi. Hastaya nörofizyolojik izleme altında işlevsel hemisferektomi yapılıp işlevsel olmayan beyin dokuları rezeksiyonu gerçekleştirilirken fMRG ve PET-BT taraması ile saptanan işlevsel bölgeler selektif olarak korundu. Takip sırasında hasta havale geçirmedi ve ayrıca günlük faaliyetleri ve iletişimini devam ettirmekte zorluk çekmedi. DDMS hastası için işlevsel hemisferektominin prognozu iyidir.

ANAHTAR SÖZCÜKLER: Dyke-Davidoff-Masson sendromu, Serebral hemiatrofi, Havale, İşlevsel hemisferektomi

INTRODUCTION

The Dyke-Davidoff-Masson Syndrome (DDMS) was first described by Dyke, Davidoff and Masson in 1933 (4). The study conducted by Albert Zilkha consisting of 5,000 CT studies of the head over a 2-year period found 10 patients with cerebral hemiatrophy (14). Unal et al. reported male sex dominance in their study which was the largest series of DDMS in the literature (13). DDMS is characterized by cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses and elevation of the sphenoid wing and petrous ridge, in association with contralateral hemiplegia, seizures, mental retardation. These findings are due to cerebral injury that may occur early in life or in utero. Thus etiology of DDMS may be classified as congenital or acquired (4). In the congenital type, there is usually no apparent etiologic factor and the symptoms are present at birth or shortly thereafter. In this category, the cerebral damage most likely occurs during intrauterine life which might be due to intrauterine vascular occlusion. In the acquired type, the symptoms are related to central nervous system damage that occurs in the perinatal period or later. Among the etiologic factors are trauma, infection, vascular abnormality, and ischemic and hemorrhagic conditions (1, 4, 8, 11, 12), coarctation of the midaortic arch, subependymal germinal matrix, amniotic bands and intraventricular hemorrhage of premature infants (10, 11, 14). CT or MRI is the gold standard for the diagnosis (1, 10). However, rapidly advancing technologies of invasive

and noninvasive functional imaging of the human brain, such as EEG, functional magnetic resonance imaging (fMRI) and positron emission tomography (PET) aid the correct diagnosis and proper management. One such rare case is reported here that was encountered in West China Hospital along with its clinical manifestations, radiological and electrophysiological features, diagnosis, differential diagnosis and appropriate treatment.

CASE ILLUSTRATION

A 13-year-old boy presented with recurrent seizures for past 10 years. This normally delivered child had seizure and left hemiparesis after bacterial pneumonia at the age of 9 months. During seizure he was unconscious with steady gaze, swallowing activities, crocidismus, but without tremor, muscle stiffness and urine or bowel incontinence. He was treated with anticonvulsant medication which was satisfactory at first but due to noncompliance the medication had poor effect and seizure recurred from time to time. Recently frequency of the seizure increased and had occurred once every 4~5 days, lasting usually for 20~30 minutes with preictal dizziness and postictal drowsiness. Physical examination revealed mild left hemiparesis, left deviated gait irregularity, and a congenital pinkish nevus in the forehead. He was mentally alert but had not achieved all the developmental milestones as compared to normal child of his age. Computed Tomography (CT) of head showed hemiatrophic cerebral parenchyma with prominent sulci and encephalomalacia. There was overdevelopment of the frontal and ethmoid sinuses and of the mastoid air cells. Elevation of the petrous ridge was also present. Dilatation of ipsilateral lateral ventricle and displacement of the midline structures toward the atrophic side were seen. The subarachnoid space was widened on the affected side (Figure 1). Head Magnetic Resonance Imaging (MRI) scan revealed right cerebral hemiatrophy with pachygyria, encephalomalacia, gliosis, porencephaly, loss of white and gray matters; hypoplastic cerebral peduncle, thalamus and internal capsule in the atrophic side; right ventricular enlargement and midline shift toward the atrophic side (Figure 3). 24-hour intensive video EEG monitoring revealed suppression of alpha rhythm in association with local slow wave activity on the side of the atrophic hemisphere. Right and left side EEG waves were highly asymmetrical. Right side EEG had relatively dominant slow waves on the anterofrontal, frontal, parietal, anterotemporal and midtemporal regions. Also the amplitudes on the right side were higher than the left (Figure 4). PET-CT showed highly functional left cerebral hemisphere and less functional right cerebral hemisphere (Figure 5, Figure 6). After detailed presurgical evaluation, the patient underwent functional hemispherectomy under neurophysiological monitoring and the nonfunctional brain tissues were resected while selectively preserving the functional areas detected by fMRI and PET-CT scan. Postoperative CT of the head after the right hemispherectomy showed a fluid filled right frontal area as well as preserved right occipital area and right basal ganglia (Figure 2). The patient was seizure free after the hemispherectomy, however, there was CSF leakage through surgical wound which affected its healing and needed twice debridement of the wound, continuous CSF drainage, and several dressings. He had left hemiparesis which was much improved in his follow up examinations. During one year follow up, he was seizure free as well as without difficulties in performing his daily activities and communication.

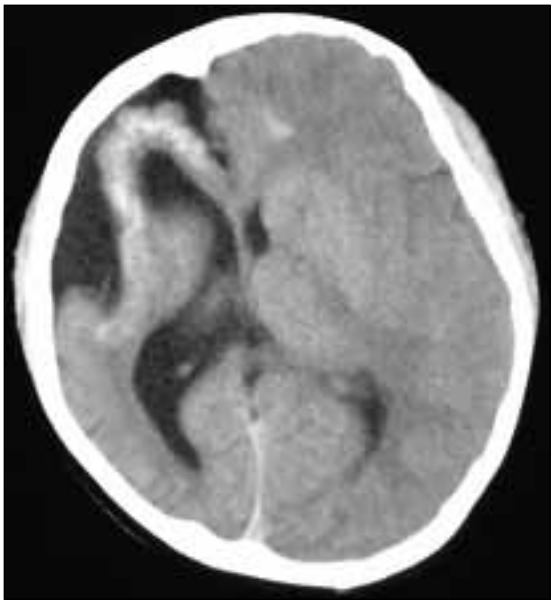

Figure 1: Preoperative CT showing the right cerebral hemiatrophy with enlarged right ventricle, ipsilateral calvarial thickening, right deviation of the falx, left cerebral hypertrophy and compression of the left lateral ventricle.

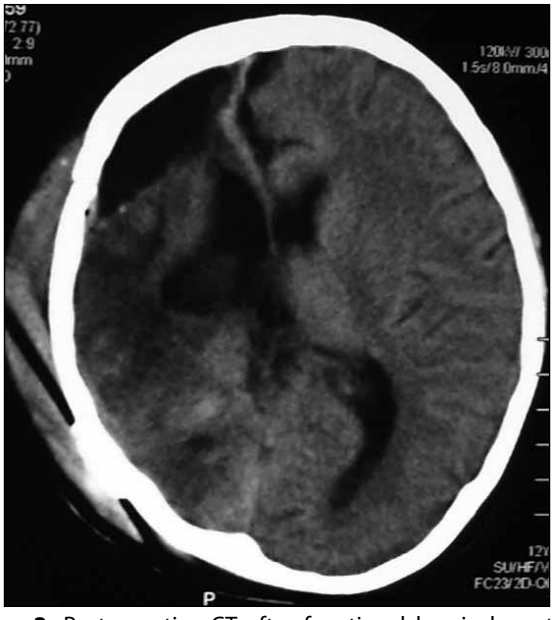

Figure 2: Postoperative CT after functional hemispherectomy showing fluid filled right frontal area as well as preserved right occipital area and right basal ganglia.

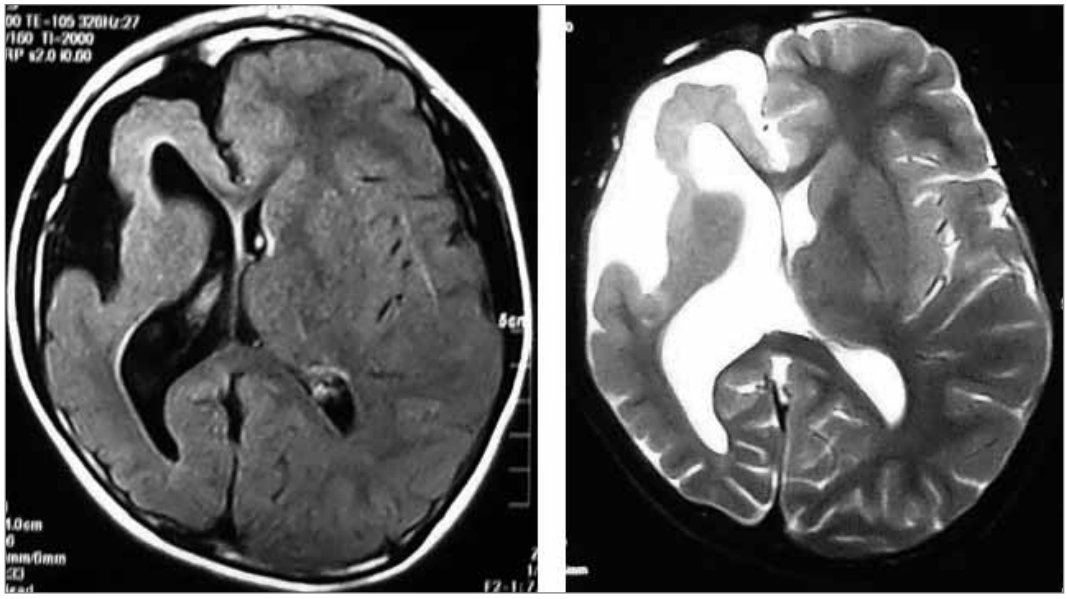

Figure 3: MRI (T1WI: right; T2WI: left) showing enlarged right ventricle, midline shift toward the atrophic side and hypoplastic right cerebral peduncle, thalamus and internal capsule, hypertrophic left hemisphere with left ventricular compression.

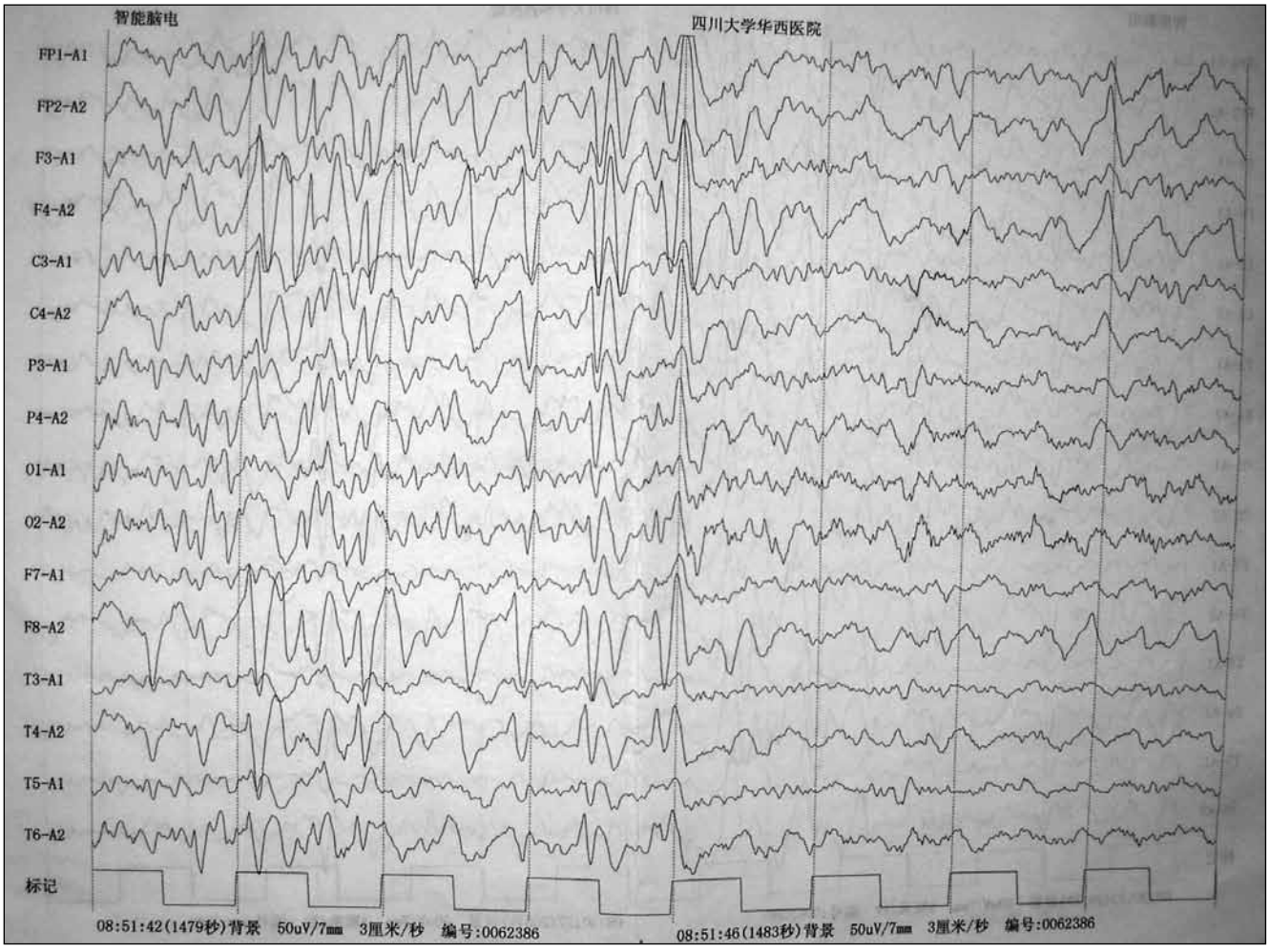

Figure 4: EEG: Right and left side EEG waves were highly asymmetrical. Slow waves were relatively dominant in FP2-A2, C4-A2, F4-A2, F8-A2 and T4-A2. Also the amplitudes in the right side were higher than left.

DISCUSSION

Cortical hemiatrophy, hemiparesis, and seizures are the typical features of DDMS (4). The patient may develop seizure at first and then hemiparesis which was seen in this case or vice versa.

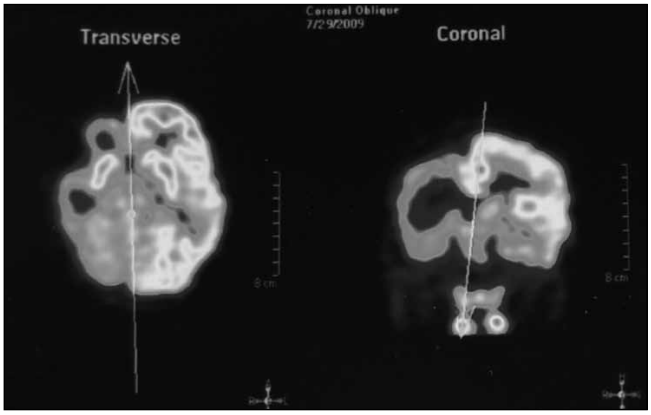

Figure 5: PET-CT. Transverse (right) and coronal (left) sections showing the highly functional left cerebral hemisphere and less functional right cerebral hemisphere.

Mental retardation may not present or appear years after the onset of hemiparesis (14). As Jules Cotard firstly recorded that unilateral cerebral atrophy in infancy does not necessarily lead to aphasia (9), DDMS patient not necessarily be aphasic. Radiological features of DDMS are unilateral loss of cerebral volume and associated compensatory bone alterations in the calvarium, like thickening, hyperpneumatization of the paranasal sinuses and mastoid cells and elevation of the petrous ridge and greater wing of the sphenoid bone (2). This case presented with most of the typical features of DDMS: cerebral hemiatrophy, seizures, contralateral hemiparesis, and mild mental retardation. Moreover compensatory cerebral hypertrophy was seen at contralateral hemisphere. The etiological factor in this patient was infection in the childhood that led the febrile seizure and recurrent seizures which inhibited the development of affected cortex. Garg et al. had also reported a similar etiological relation of cerebral hemiatrophy with febrile seizures (5). The prominent sulci and encephalomalacia in the atrophic hemisphere reflect a late onset of brain insults due to abnormal neuronal and glial proliferation or apoptosis during cortical development

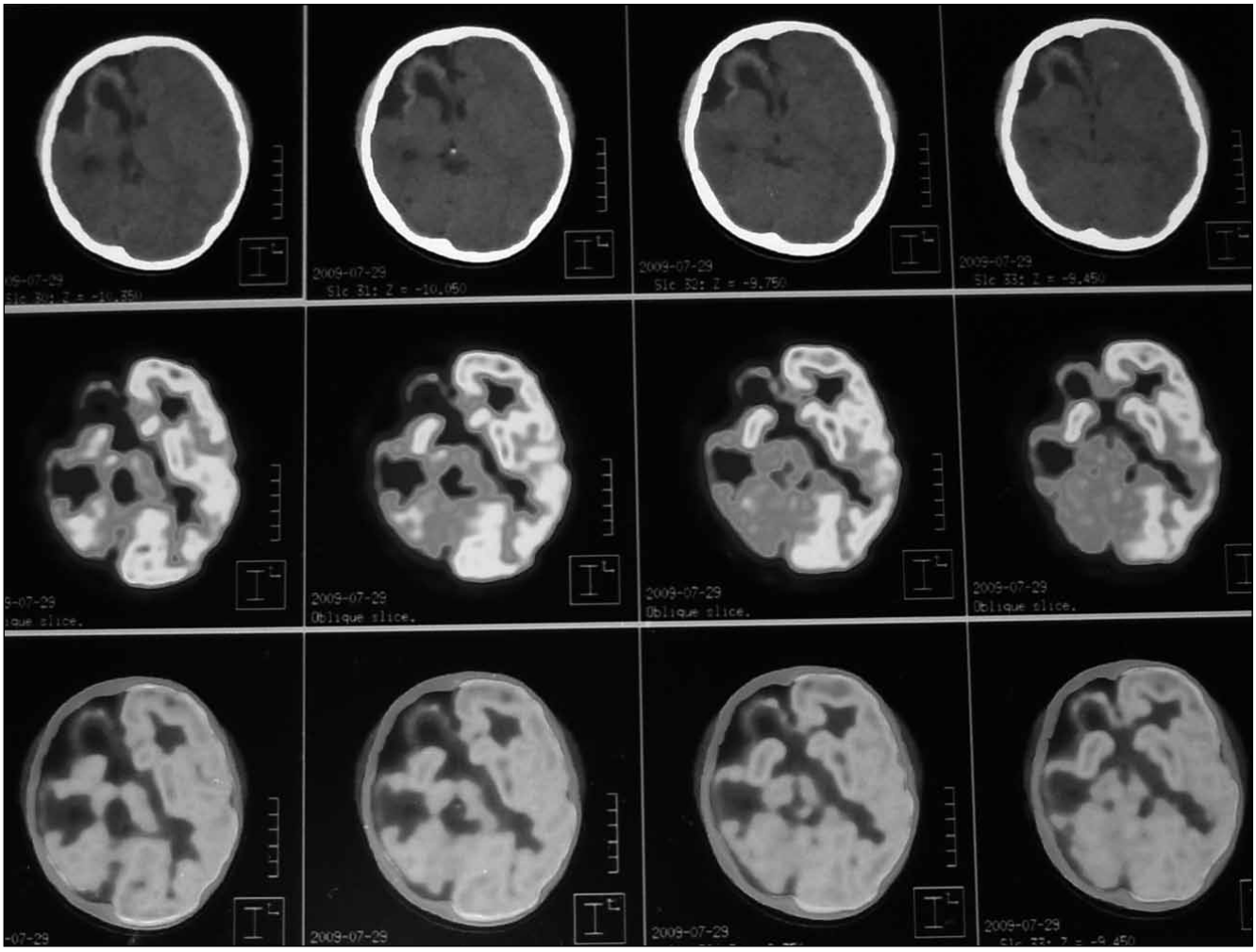

Figure 6: Transverse PET-CT showing presence of functional activity in the occipital lobe as well as discrete functional activity of frontal and temporal lobes of the right cerebral hemisphere.

which were the consequence of intracranial infection in this case. On the other hand, if the brain insults occurs during embroyogenesis, when the formation of gyri and sulci is incomplete, no prominent sulci will be present (2). Typical skull changes develop when insult to the brain occurs during the first 18 months to 2 years of life. In cerebral atrophy, whether unilateral or bilateral, focal or diffuse, the common factor is loss of brain tissue, although the nature and extent of the underlying pathologic processes vary widely. Adaptation to unilateral decrease of brain substance may consist calvarial thickening affecting particularly the diploic space with loss of convolutional markings of the inner table of the skull (4, 14), overdevelopment of the frontal and ethmoid sinuses and of the mastoid air cells, elevation of the petrous ridge, dilatation of one lateral ventricle and displacement of the midline structures toward the atrophic side and widened subarachnoid space on the affected side. EEG showing suppression of alpha rhythm in association with local slow wave activity on the side of the atrophic hemisphere in this case as well as noted by others (7) might be due to the pathologic process itself or due to loss of brain tissue. Conditions that are associated with cerebral hemiatrophy such as Rasmussen encephalitis, Sturge-Weber syndrome, some brain tumors, Silver's syndrome, linear nevus sebaceous syndrome and progressive multifocal leucoencephalopathy should be differentiated. Cerebral hemiatrophy without seizure most likely cause due to cerebrovascular disease (7). Medically intractable patients are the candidates for the surgical treatment in an attempt to achieve better seizure control. Functional imaging like PET and fMRI can visualize alterations in cerebral perfusion or metabolism that provides important information regarding the localization of eloquent cortex adjacent to the lesions or planned resection line (3). Functional hemispherectomy after detailed presurgical evaluation is reasonable for the panhemispheric syndromes associated with intractable seizures (3, 6). In this patient CSF leakage occurred which also affected wound healing around the leakage area which was efficiently managed by twice debridement of wound, continuous CSF drainage, and several dressings. In the year-long follow up after functional hemispherectomy he was seizure free and without difficulties in performing his daily activities and communications.

In the present study, the classical clinical features of the DDMS is described with its radiologic and electrophysiologic characteristics. CT or MRI is gold standard for diagnosis. Functional evaluation by fMRI, PET should be performed while planning the surgery. Functional hemispherectomy for the DDMS patient has a good prognosis. Further study in the future is necessary to optimize the treatment.

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