

CASE REPORT

Large Porencephalic Cyst with Epilepsy in an Adult: A Rare Clinical Entity

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ABSTRACT

Porencephalic cyst is the presentation of cysts or cavities in the brain as a result of developmental defects or acquired lesions, as associated with ischemia. Porencephalic cysts are most frequently located in the Sylvian fissure region and are typically associated with the subarachnoid space or ventricular system. Mental retardation, spastic quadriparesis, optic atrophy, and seizures may be seen. The cysts may be bilateral or unilateral. We discuss in this case report the follow-up and treatment of an adult patient presenting with porencephalic cysts related to late-stage epileptic attacks. The seizures are seen frequently in the early stage, though not all patients present with seizures. The patients with a normal neurological examination may be followed without surgical procedures. Those with resistant epilepsy may undergo surgery as well as medical therapy.

Key words: Porencephalic cyst, Cortical developmental malformations, Epilepsy

Received: September 15, 2012 • Accepted: April 19, 2013

ÖZET

Epileptik Yetişkinde Büyük Porensefalik Kist: Nadir Bir Klinik Tablo

Porensefalik kist, gelişimsel defektlere veya iskemi gibi edinsel lezyonlara bağlı olarak beyin içerisinde kistler veya kaviteler bulunmasıdır. Porensefalik kist en sık silvian fissür bölgesine yerleşir ve tipik olarak subaraknoid aralık veya ventriküler sistem ile bağlantılıdır. Mental retardasyon, spastik kuadriparezi, optik atrofi ve nöbetler görülebilir. Kistler unilateral veya bilateral görülebilir. Biz yazımızda geç dönem epileptik ataklarla gelen porensefalik kist tanılı yetişkin hastanın takip ve tedavisini tartıştık. Nöbetler genellikle erken dönemde görülür ve tüm hastalar epilepsi ile başvurmaz. Normal nörolojik bulgulara sahip hastalar takip edilir. Nörolojik defisit ve dirençli epilepsi görülürse cerrahi tedavi ve ilaç tedavisi uygulanabilir.

Anahtar kelimeler: Porensefalik kist, Kortikal gelişimsel malformasyonlar, Epilepsi

Geliş Tarihi: 15 Eylül 2012 • Kabul Ediliş Tarihi: 19 Nisan 2013

INTRODUCTION

Porencephalic cysts are cavities that are formed due to damaged brain tissue associated with ischemia and they are filled with cerebrospinal fluid (CSF). These cavities are triangular and cover the cerebral cortex, and are considered to be an irrigation region like the middle cerebral artery^[1-3]. Radiological imaging techniques have facilitated the detection of cortical developmental malformations. They are generally imaged as hypodense foci on computerized tomography (CT) or hypoechogenic sites on ultrasound results. The intracystic fluid fills the cavity rather than occupying a place, and presents no or very little mass effect^[4-6]. Treatment may include physical therapy, medication for seizure disorders, and a shunt for hydrocephalus. In the case of porencephalic cysts, which sometimes involve the large sites of the frontal or parietal lobe, slight hemiparesis or sensation defects, which may be detected with a detailed neurological examination, and a level of growth asymmetry may develop as neurological deficits^[4,6]. Severe malformations may be seen with pronounced developmental retardation and intermittent seizures with early onset. Slight malformations are detected on the diagnostic assessments after the seizures originating in any period. Some patients with this disorder may develop only minor neurological problems and have normal intelligence. The seizures are seen frequently in the early stage, and in most cases, startle epilepsy is refractory to antiepileptic drugs. Surgical management of intractable epilepsy in porencephalic cyst patients is an important method for controlling epilepsy^[1,3,5].

CASE REPORT

A 24-years-old male patient presented with intermittent epileptic seizures that lasted for approximately three months and occurred several times a day. It was apparent that these attacks were immediate, and during them, the patient focused upon a fixed point. According to the patient's self-report, the attacks began with exaggerated motion, especially of the arms. These attacks lasted for 1-2 minutes; the patient suffered amnesia and could not recall what he did during the attacks. The histories taken demonstrated that these attacks were consistent with complex partial seizures. There was no knowledge of other diseases, administration of medication, trauma, or operation history. His blood and electrolyte values were in normal ranges. The neurological examination and electroencephalography (EEG) test results were normal (Figure 1). The CT and magnetic resonance imaging

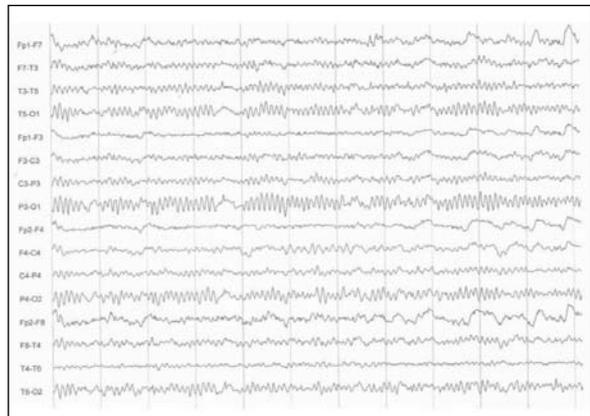


Figure 1. EEG test results shows rhythmic activity with specific high amplitude.

(MRI) revealed a plain contour cystic lesion characterized by hyperintense signal in the T2-weighted sets (with CSF isointense), which caused a detected appearance on the corpus callosum covering the right lateral ventricle, especially on the occipital horn. In the contrast and edema examination of the lesion periphery, no staining was detected (Figure 2). No surgical operation was performed in view of the normal neurological and EEG findings. Following the diagnosis, with the administration of anti-epileptic medications, the epileptic attacks were considered to have ceased at the 14th month follow-up. The clinical progression of the patient was good, and he has been followed non-surgically.

DISCUSSION

In the pediatric age group, mental-motor retardation, epilepsy and other neurological disorders are associated with malformations of cortical development, and in conjunction with the improving neuro-radiological methods, the frequency of their detection has gradually increased^[1,5,6]. The data on the frequ-

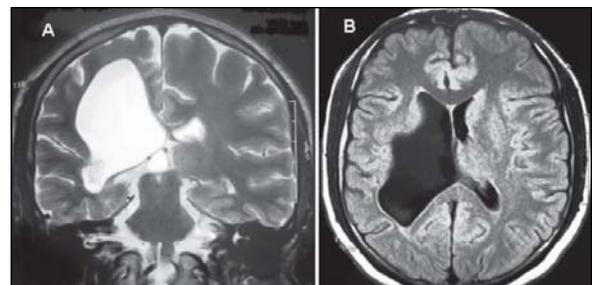


Figure 2. T2-weighted MR images in the coronal (A) and T1-weighted MR images in the axial plane show porencephalic cysts (B).

ency of the malformations in cortical development are limited, and the incidence rates are not known exactly. The basis of these malformations, including porencephaly, consists of neuronal proliferation and migration and recessions in the organizational step in brain development. Genetic and prenatal environmental factors during the developmental steps contribute to the development of malformations, and the reason for this condition has not been completely revealed^[2,4,6]. It is known that environmental factors affect neuroblast migration before the 16th week of pregnancy, and those neuroblasts are the genetic origin of most neuronal migration disorders. Porencephalic cysts are cavities that are filled with CSF formed by the damaged brain tissue after ischemia. The most severe form, alobar holoprosencephaly, accompanies many dysmorphic characteristics and is also clinically severe. Lobar and semilobar forms present relatively milder clinical courses^[1,3,5]. Porencephalic cyst can develop to focal injury during the fetal period, often follows vascular territories. It can develop as secondary due to infarct, hemorrhage or infectious etiologies such as cytomegalovirus (CMV). Congenital porencephalic cysts originate from intrauterine vascular or infectious injury. In the placenta of monozygotic twins, in one of the fetuses, the association of vessel anastomosis-localized ischemia may have caused immediate hypotension and hypoxia, leading to the organ damage and associated abnormalities observed in a presented case^[3,6]. Clinical findings in cortical developmental malformations such as porencephaly are variable, and the most substantial findings vary from mental-motor retardation and epilepsy to focal neurological disorders due to the functions of the affected location^[3,5,6]. Epilepsy consists of partial or generalized attacks related to the spread of the epilepsy lesion. The seizures are seen frequently in the early stage, though not all patients present with seizures, and various responses to the antiepileptic agents may be seen in different patients. Motor findings may vary from medium hemiplegia to severe atonic diplegia, and the mental findings may vary from a normal mental course or slight learning disability to severe mental retardation^[1,5]. The patients with diffuse disorders may present with early symptoms, and the prognosis is poor. Microcephaly may frequently accompany. The patients with lateral focal disorder present with slight motor, cognitive and speech disorders^[1,4]. EEG may help in the diagnosis, but the findings are not specific. MRI is a valuable method for the detection of such pathologies, and provides the

detection and diagnosis in 50-70% of the disorders. Volumetric MRI may be employed, and continual thin sectional images are required for the most appropriate anatomical assessment in T1-weighted series. The abnormal MRI findings and clinical EEG information have demonstrated 84% efficiency in detecting porencephalic cysts in the reported studies^[1,4,6]. Porencephalic cysts frequently lead to symptoms and signs related to intracranial hypertension. These mass effects may be determined with imaging methods. It has been reported that the cystoperitoneal shunting procedure may resolve the symptoms in patients with clinical presentation of pronounced pressure. In the anastomosis of sterile fluid in the porencephalic cavity with the ventricle, it has been considered that the mass effect of the cavity may be ruled out by filling the cavity with CSF within the scope of "communicating vessels". The patients with a normal neurological examination may be followed without surgical procedures^[1,5,6]. Those with resistant epilepsy may undergo surgery as well as medical therapy. A kind of surgical intervention is applied in the patients with medication-resistant epilepsy that develops with all modifications, including hemispherectomy and hemispherotomy. This technique is most frequently applied in pediatric patients and is employed in cases of large porencephalic cysts related to ischemia or trauma, hemi-megalencephaly, infantile hemiplegia, and Rasmussen encephalopathy^[3,5,6]. The prognosis for individuals with porencephaly varies according to the location and extent of the lesion. Some patients with this disorder may develop only minor neurological problems and have normal intelligence, while others may be severely disabled. Others may die before the second decade of life. The seizures are seen frequently in the early stage, though not all patients present with seizures, and various responses to the antiepileptic agents may be seen in different patients.

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