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Case Report

Schizencephaly Diagnosed After Childhood: Report of Two Cases

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Abstract

Schizencephaly is a rare congenital malformation of the brain in which there are abnormal clefts lined by cortical gray matter that allow communication between the subpial space and the lateral ventricle in the cerebral hemispheres, as a result of a neuronal migration disorder. Due to its rarity, the study of this condition has been limited, and knowledge about its etiology, diagnosis, management, and prognosis is still unclear, with the vast majority of published studies occurring in pediatric patients. We present two cases, one is a 32-year-old man and the other is a 17-year-old woman who attended our unit, in both cases the diagnosis of schizencephaly was established. Since most of what is known about schizencephaly has been reported in children; Through this case report, we intend to offer our experience in the evaluation, diagnosis, and management of this rare condition in adults.

***Keywords:** seizures, electroencephalogram, brain, epilepsy, schizencephaly, antiepileptic*

Introduction

Schizencephaly is a congenital malformation of the brain in which there are abnormal clefts lined by cortical gray matter that allow communication between the subpial space and the lateral ventricle in the cerebral hemispheres. (1) It is a rare condition, with a 2005 population-based study a prevalence of 1.54 per 100,000 births. (2)

Case Report

Mr. S is a 32-year-old man with a history of epilepsy since the age of 16, treated with phenytoin and with satisfactory treatment adherence. However, he reported monthly tonic-clonic seizures in the left side of the body with altered alertness and aura consisting of paresthesias of the affected limbs.

In February 2020, Mr. S was taken to our institution due to convulsive status epilepticus lasting 40 minutes. Upon arrival at the Shock Trauma Unit, orotracheal intubation was decided because status epilepticus persisted. A simple cranial computed tomography scan was performed, in which morphological abnormalities were observed in the right cerebral hemisphere, consistent with a communication between the subarachnoid space and the ventricular system at the level of the right temporal lobe (Figure 1). The imaging finding was compatible with open-lip schizencephaly, which caused retraction of the occipital horn of the left lateral ventricle. All other structures were found normal. It was established that schizencephaly was the central diagnosis of our patient, explaining its clinical manifestations. Mr. S was admitted to the intensive care unit where he was also diagnosed with healthcare-associated pneumonia and acute kidney injury for which he received medical treatment in addition to starting antiepileptics such as valproic acid and levetiracetam. The patient was extubated 3 days after admission and continued surveillance in the neurology ward. He was discharged home after completing the antibiotic regimen and had no new convulsive events. The electroencephalogram was normal and we decided to continue follow-up as outpatient consultation.

Miss B is a 17-year-old woman who in July 2019 went to the Emergency Room for several focal tonic-clonic seizures in the 3 hours prior to her arrival at the Hospital. She refers a history of having presented similar episodes in 2017 and 2018 with an approximate frequency of one every 3 months. She denied taking any treatment or seeking medical help. In the emergency department, treatment with phenytoin was started. The computed tomography scan of the skull was reported normal, but the magnetic resonance imaging of the brain showed an irregular cleft of the subarachnoid space in the right frontal pole, this cleft extends from the cortex to the ipsilateral lateral ventricle and the connection between them were observed. It can be seen covered with gray matter throughout its length. (Figure 2, Figure 3)

With the aforementioned findings, the diagnosis of open-lip schizencephaly in association with left frontal polymicrogyria was established. The electroencephalogram turned out to be normal.

In the follow-up consultations, she reported not having any more seizures since his stay in the emergency department of this hospital during July 2019 and in the physical examination we only found an increased left patellar reflex and ipsilateral extensor plantar response. The remainder of his neurological examination was normal.

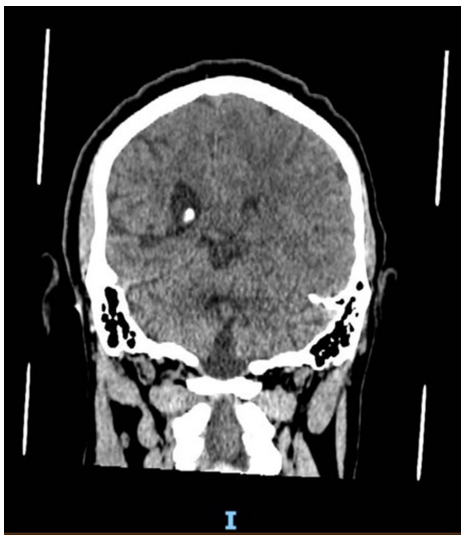

Figure 1: *Simple tomography of the brain, coronal section in which schizencephaly with open lips is observed.*

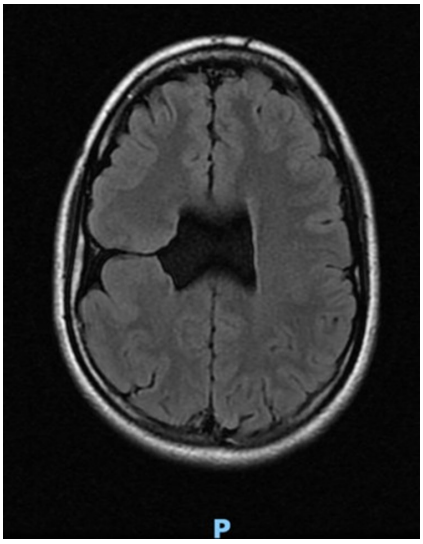

Figure 2: *Simple MRI of the brain, axial slice, showing open-lip schizencephaly.*

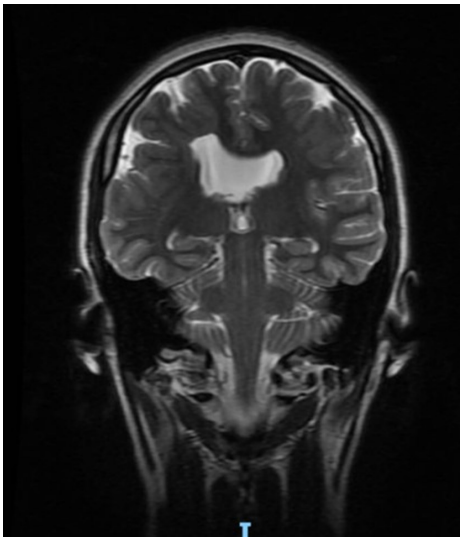

Figure 3: *Simple magnetic resonance imaging of the brain with open lip schizencephaly and retraction of the right lateral ventricle.*

Results & Discussions

Schizencephaly is a condition with a variable clinical presentation, which can range from being asymptomatic, neurocognitive dysfunction, focal seizures, motor disturbance. (3)

When comparing our patients with a diagnosis of schizencephaly, both cases presented focal and generalized tonic-clonic seizures as a frequent complaint. Both cases responded favorably to antiepileptic treatment. For some reason the diagnosis in both patients was not made during childhood. It has been reported that clefts consistent with schizencephaly are frequently missed (4) which of course makes it difficult to timely diagnose schizencephaly, a condition with a highly irregular clinical presentation and otherwise uncomplicated.

We consider the contrast between the imaging findings of these cases noteworthy: while the plain CT scan of Mr. S's brain shows no abnormalities other than the defining cleft of schizencephaly, the plain MRI of Ms. B's brain yielded several additional structural abnormalities such as polymicrogyria. (4,5)

Conclusion

As mentioned above, most of the information available on schizencephaly comes from studies with pediatric patients. These cases represent the rare occurrence of schizencephaly that is diagnosed after childhood, so through these reports we intend to offer our experience in the treatment of these patients. Also through the comparison of the cases of Mr. S and Mrs. B, we aim to offer examples of how this same condition can present with various clinical and imaging findings, so clinicians should be aware that we can find different clinical features among affected patients.

Conflict of Interest

The authors declare no conflict of interest.

Acknowledgement

None

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