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Schizencephaly in Adulthood: A Report of Two Cases

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ABSTRACT

Objective: *To present two clinical cases of adult patients without a prior diagnosis of schizencephaly who were admitted to the emergency department due to acute neurological events.*

Methods: *A brief literature review is provided along with the presentation of two clinical cases involving adult patients with no prior history of the disease, who were diagnosed with schizencephaly after being admitted to the emergency department.*

Keywords

Schizencephaly, Adult-onset presentation, Incidental Findings, Neuronal Migration Disorders, Congenital brain malformations, Neuroimaging.

Introduction

Schizencephaly is a congenital malformation of the central nervous system resulting from a neuronal migration disorder. It is characterized by the presence of a cerebral cleft that spans the thickness of the hemisphere, connecting the ventricular lumen with the subarachnoid space [1,2]. It is a rare condition (1.5/100,000 people) with an undefined etiology, although it has been associated with infectious causes and genetic mutations [2,3]. Clinically, schizencephaly presents with the characteristic triad of neuronal migration disorders: seizures (mostly focal), intellectual disability, and motor impairment; therefore, it is generally diagnosed at an early age [3,4]. The classification of this entity consists of 4 types, depending on whether the malformation is open-lip or closed-lip, and whether it is unilateral or bilateral, as well as the combination of these conditions [2-4].

Case Presentation

Case 1

A 41-year-old male with a history of diabetes mellitus and no known neurological history. He was admitted to the emergency department presenting with partial and focal seizures in the left lower limb, which later generalized to all four extremities. Upon admission, the patient was neurologically intact, with no abnormalities found during the physical examination. During his hospital stay, additional focal seizure events were observed, which were initially managed with phenytoin, showing an adequate response. A non-contrast head CT scan was performed (Figure 1), reporting a right closed-lip schizencephaly involving the temporal lobe. Management was adjusted by the neurology department, and the patient was discharged on anticonvulsant therapy.

Case 2

An 82-year-old female with a history of systemic arterial hypertension and sequelae of poliomyelitis since childhood. She presented to the emergency department following a traumatic brain injury; five days later, she developed emesis and headache. Upon admission and throughout her stay in the emergency

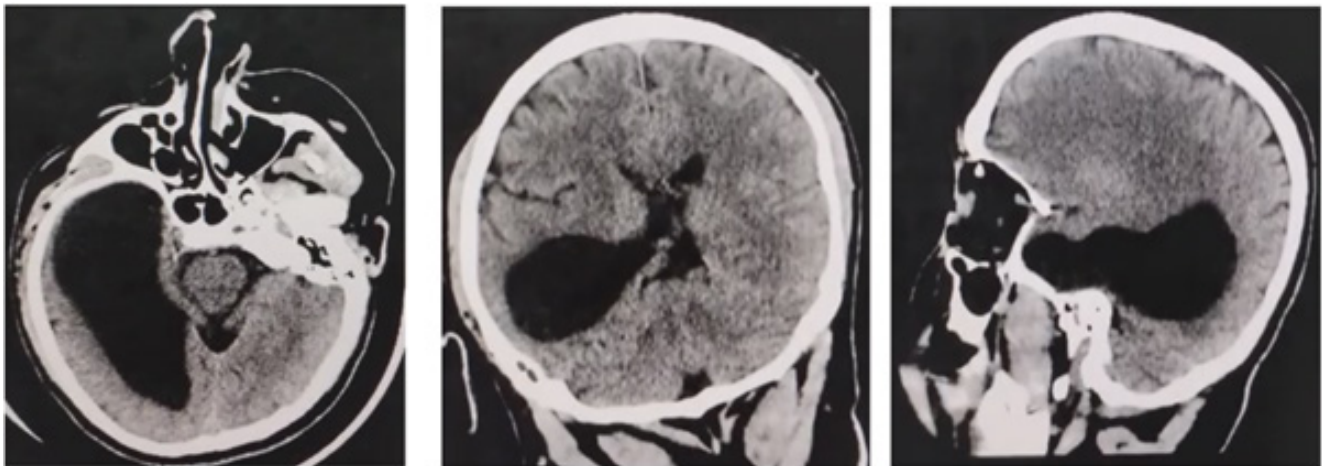

Figure 1: Axial, coronal, and sagittal planes showing a right closed-lip schizencephaly involving the temporal lobe.

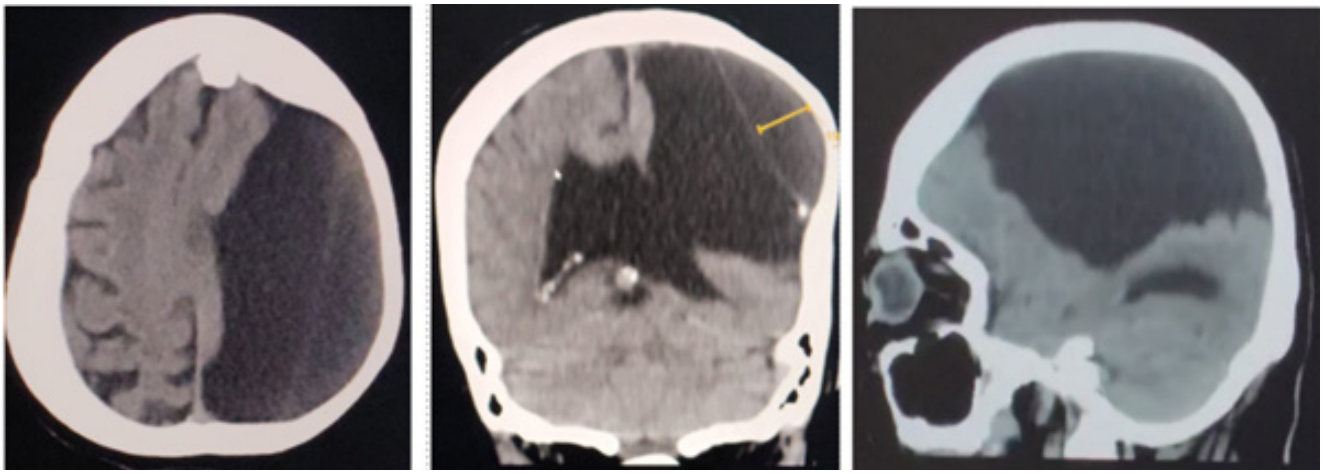

Figure 2: Axial, coronal, and sagittal planes showing a left open-lip schizencephaly.

department, no neurological deficits were found during physical examination. A non-contrast head CT scan documented an open-lip schizencephaly (Figure 2). The patient was discharged with a referral for neurology follow-up.

Discussion

The presentation of schizencephaly in adulthood is uncommon, as most cases are identified in the pediatric population. These cases are remarkable because they highlight the wide clinical spectrum of the disease, ranging from new-onset seizures to entirely asymptomatic incidental findings in elderly patients. While Magnetic Resonance Imaging (MRI) is the gold standard for detailed classification, non-contrast Computed Tomography (CT) remains a fundamental tool for initial identification in the emergency setting. These findings emphasize that schizencephaly should be considered in the differential diagnosis of acute neurological events, regardless of the patient's age.

Conclusion

Schizencephaly is a rare condition that may present late in life or as an incidental finding. It should be considered in the differential diagnosis of patients with neurological symptoms in the emergency department. When the presentation is late-onset, treatment should be based on symptom control; however, if it is a finding without clinical manifestation, it may not require treatment.

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