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### Abstract

Schizencephaly is a cerebral malformation characterized by a cleft lined with gray matter crossing the entire hemisphere, from the lateral ventricle to the outer surface of the cortex; it would be due to a defect in cell migration during cerebral embryogenesis. Its association with epilepsy is frequent, as well as motor and language disorders and mental retardation. We report the case of a 3-year-old girl who had two open schizencephalic clefts which were objectified on cerebral computed tomography. She also had behavioral and mood disorders, probably related to temporal lobe seizures, which were much improved by sodium valproate.

**Keywords:** Behavioral disorders- Epilepsy- Mood disorders- Temporal lobe- Schizencephaly.

### Résumé

La schizencéphalie est une malformation cérébrale caractérisée par une fente bordée de substance grise traversant tout l'hémisphère, du ventricule latéral à la surface externe du cortex ; elle serait due à un défaut de migration cellulaire au cours de l'embryogenèse cérébrale. Son association à l'épilepsie est fréquente, de même que des troubles moteurs, du langage et un retard mental. Nous rapportons le cas d'une fille de 3 ans qui avait deux fentes schizencéphaliques ouvertes qui ont été objectivées à la tomodensitométrie cérébrale. Elle présentait en plus des troubles du comportement et de l'humeur sans doute en rapport avec des crises du lobe temporal et qui ont été bien améliorés par le valproate de sodium.

**Mots-clés :** Epilepsie- Lobe temporal- Schizencéphalie- Troubles du comportement- Troubles de l'humeur.

### Introduction

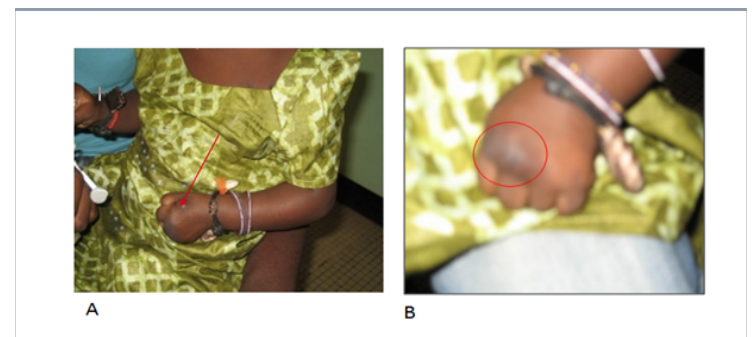
Schizencephaly, from the Greek “skhizein” (splitting, fissure, division) and “cephalon” (head) is a rare cerebral malformation resulting from defects in cell migration. It is characterized by a slit lined with gray matter crossing the entire hemisphere, from the lateral ventricle to the outer surface of the cortex. It can be unilateral or bilateral, closed or open. It can manifest as motor disorders, psychomotor retardation and epilepsy [1, 2].

### Observation

We report the case of a female three-year-old child brought to a neurology consultation at the Principal Hospital in Dakar for seizures probably of the myoclonic type, a left hemispheric deficit, language delay, behavioral disorders and mood disorders. Behavioral disorders were recurrent episodes of self-harm (self-biting, violent banging of the head against the

wall) and were favored by anger. Indeed, mood disorders made of an angry tendency and extreme nervousness were also noted in the child.

Physical examination showed spastic left hemiplegia, delayed psychomotor development, and a scar on the back of the left hand that resulted from self-biting scenes (the child bit her left hand violently each time she was angry) (figures 1 and 2).

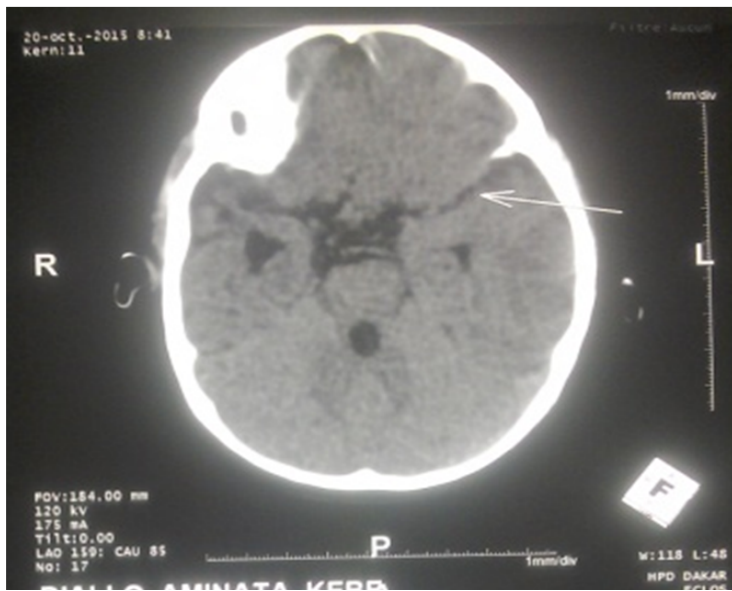


**Figure 1(A et B):** Self-bite scars indicated in A by the red arrow and in B by the red circle.



**Figure 2:** Right schizencephalic cleft on this axial slice.

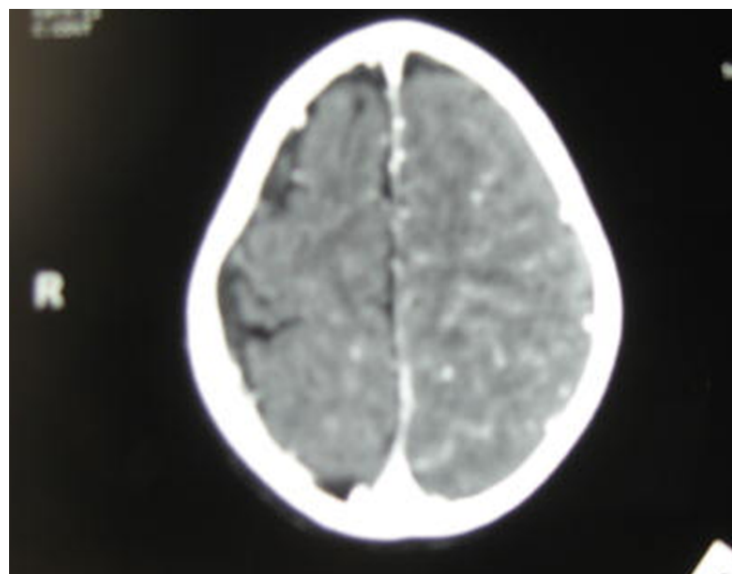
On the paraclinical level, cerebral CT showed bilateral schizencephaly with open clefts and right hemispheric atrophy (figures 3, 4, 5 and 6).



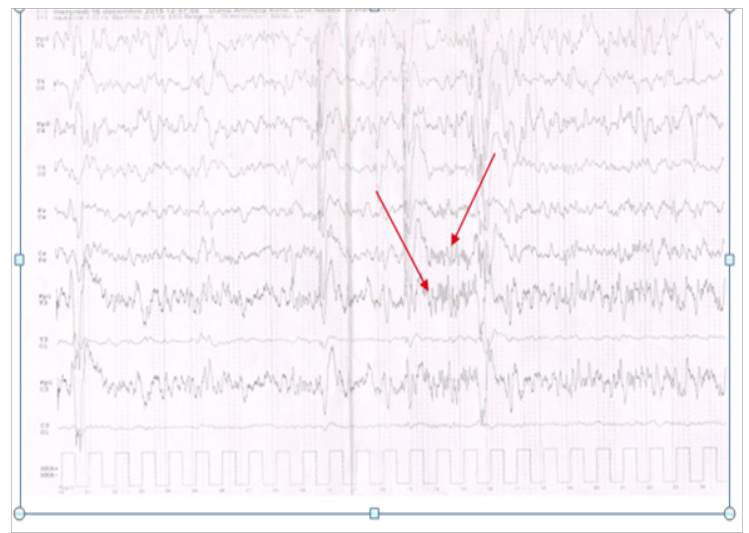
**Figure 3 :** Left schizencephalic cleft on this axial slice.



**Figure 4 :** Right cleft with cortical-subcortical right hemispherical atrophy on this coronal slice.

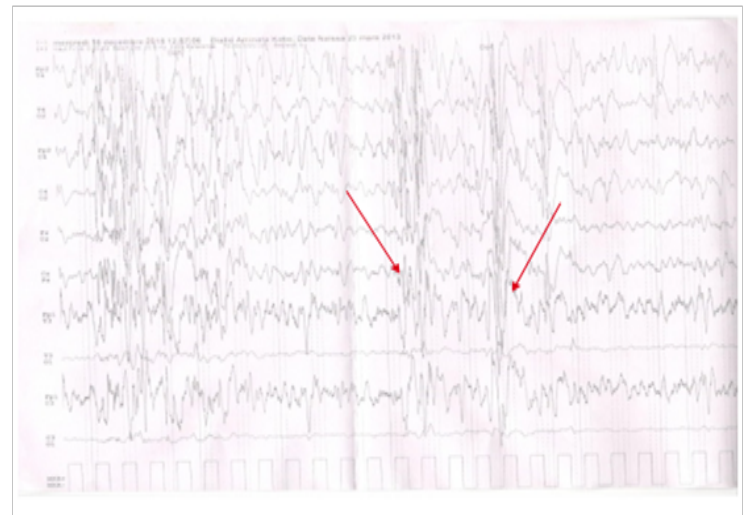


**Figure 5 :** Clear right hemispheric hemi atrophy on this axial slice.



**Figure 6 :** Sleep spindles present on the left only (indicated by the red arrows) and attesting to an asymmetry in favor of a more marked right hemispheric suffering.

The electroencephalogram showed right hemispheric suffering associated with bursts of diffuse and repetitive slow wave spikes (figure 7).



**Figure 7:** Diffuse and repetitive puffs of atypical slow wave spikes in favor of diffuse irritative cerebral suffering (indicated by red arrows).

Treatment with sodium valproate (25mg/kg/d) was instituted. The evolution was favorable because after 6 months of treatment, seizures were considerably reduced, as well as behavioral and mood disorders. The child “was more fulfilled with a clear propensity to play, smile and share” according to the words of her elder brother who brought him for consultation. It is important to point out that previously a traditional treatment, to supposedly fight against «evil spirits which made the child violent», had been instituted in vain.

### **Discussion**

Schizencephaly is a rare malformation, about 5% of all cortical malformations [3]. It would be due to disorders of neuronal migration and cortical organization occurring during cerebral embryogenesis. The first exhaustive anatomic-pathological descriptions were reported by Yakovlev and Wadsworth in 1946 during an autopsy series [4]. The exact etiology is not known. Vascular factors (ischemia of layer V of the cortex), infectious

(cytomegalovirus), genetic (existence of familial and sporadic cases associated with mutations of the EMX2 gene), toxic (antenatal use of cocaine or other alpha-stimulants, intoxication oxycarbonate) and traumatic (maternal trauma) have been considered [3, 5, 6]. We were unable to identify etiological factors in our case.

The severity of the clinical symptomatology depends on the width of the cleft but also on any associated lesions. The most common manifestations are: motor disorders, epilepsy, mental retardation and language disorders. All these manifestations, rather usual in schizencephaly, were found in our patient who, in addition, presented mood and behavior disorders. These latter could be related to the temporal location of the cleft and would thus be more related to the expression of temporal epileptic seizures, when we know the importance of psychiatric-like manifestations in this type of focal epilepsy [7]. The good response to treatment with sodium valproate is an argument in favor of this hypothesis. It also made possible to concretely invalidate the false supernatural interpretation initially given by the family to the child's behavioral problems and which had justified recourse to traditional treatment for a certain time.

### **Conclusion**

The epilepsy-schizencephaly association is common. In our case, manifestations of psychiatric appearance and the mood disorders, having been the subject of an attempt at supernatural interpretation, would rather be linked to the expression of crises of temporal lobe origin, given the location at this level of the cleft. The good evolution of these symptoms under sodium valproate really tends to confirm this hypothesis.

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