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Double cortex syndrome: A striking case of X-linked subcortical band heterotopia

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Abstract

Double cortex syndrome is a rare X-linked dominant disorder affecting females. It typically presents with developmental delay and epileptic seizures. Brain MRI features are characteristic and strongly suggestive of the diagnosis. We report the case of a 6-year-old girl who presented to the pediatric clinic with speech delay and generalized tonic-clonic seizures. MRI revealed sub cortical band heterotopia.

Keywords: Epilepsy; X-Linked; Band heteretopia; MRI.

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Case presentation

We report the case of a 6-year-old girl who presented to the pediatric clinic with speech delay and generalized tonic-clonic seizures since the age of three. History revealed a full-term vaginal delivery with a normal APGAR score and first-degree parental consanguinity. An initial brain CT scan was performed and returned unremarkable. A recent electroencephalogram (EEG) showed findings suggestive of epilepsy. Axial and coronal T2-weighted MRI sequences showing thickening of the frontoparietal cortex (Figure 1A and 1B - blue arrow). A coronal TIR MRI sequence (Figure 2) showing thickening of the frontoparietal cortex, organized into two parallel bands of gray matter separated by a thin layer of white matter (red arrow), creating a "double cortex" appearance.

Discussion

Band Heterotopia, also known as <<Double Cortex>> syndrome is a rare form of diffuse gray matter heterotopia primarily affecting females due to a genetic anomaly in the DCX gene [1]. Clinically, it manifests as seizures and developmental delays, generally appearing during the first years of life [3].

Developmental disorders in children are increasingly common in the age group between 3 and 17 years [2].

Neurological and physical examinations may be normal in some cases; however, hypotonia, dysarthria, poor fine motor control, or, in rare cases, a pyramidal syndrome may also develop. During the embryonic development of the cerebral cortex, any disruption—whether genetic, toxic, infectious, or ischemic—can lead to a "neural migration disorder", which is a broad term that encompasses conditions such as lissencephaly, pachygyria, heterotopia, schizencephaly, hemimegalencephaly, and cortical dysplasia.

Genotypic variations in the DCX gene result in a spectrum of neuronal migration disorders depending on the nature of the mutation and the patient's sex. Hemizygous mutations in males often cause more severe phenotypes such as classic lissencephaly, while heterozygous females typically present with subcortical band heterotopia due to X-linked mosaicism [1]. This variability accounts for the broad range of clinical presentations observed, from mild cognitive impairment and controllable seizures to profound developmental delay and drug-resistant

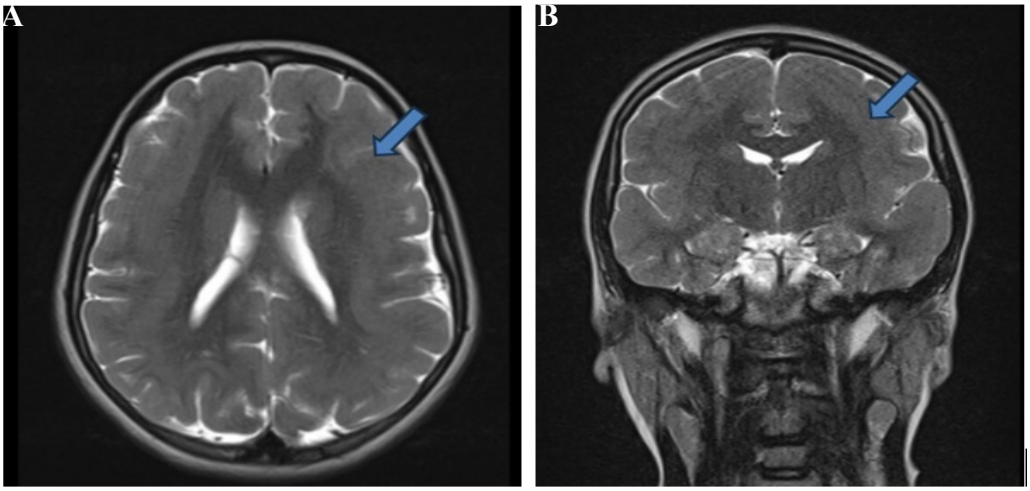

Figure 1: Axial and coronal sections in T2 sequence of a brain MRI **(A and B)** showing thickening of the frontoparietal cortex organized into two parallel bands of gray matter separated by a thin layer of white matter, creating a "double cortex" appearance.

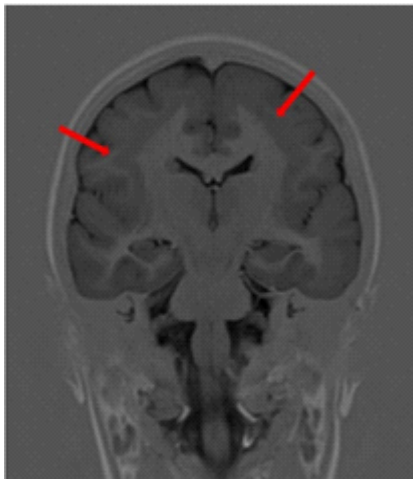

Figure 2: Coronal section in TIR (Turbo Inversion Recovery) sequence showing thickening of the frontoparietal cortex organized into two parallel bands of gray matter separated by a thin layer of white matter, creating a "double cortex" appearance.

epilepsy. Importantly, cases involving familial inheritance have been documented, underscoring the value of genetic screening and counseling for affected families [1].

There are three types of gray matter heterotopia: subependymal, subcortical, and band-like [1]. MRI is the diagnostic tool of choice. Band heterotopia presents as a continuous subcortical heterotopic band that is isointense to gray matter in all sequences [1]. The band can vary in thickness and size. The overlying cortical mantle may appear normal on MRI or may show abnormalities ranging from agyria to pachygyria [1]. The severity of the cortical abnormality may be related to the thickness of the heterotopic band. The thickness of the heterotopic gray matter band is associated with the shallow depth of sulci in the overlying cortex [1]. Differential diagnoses to consider include pachygyria and lissencephaly, which may appear similar but lack the distinct intermediate layer of white matter separating the cortical and subcortical gray matter bands. Additionally, focal cortical dysplasia or tuberous sclerosis may show cortical thickening or signal changes but are usually asymmetric and localized rather than diffuse. Recognizing these imaging patterns is crucial for accurate diagnosis and guiding genetic testing [2].

Early diagnosis is essential for implementing appropriate therapeutic strategies and providing genetic counseling. In terms of treatment, managing epilepsy is a primary concern. While antiepileptic drugs are commonly used, their effectiveness varies among individuals. Non-pharmacological interventions, such as deep brain stimulation of the anterior nuclei of the thalamus, have shown promise in reducing seizure frequency in some patients with subcortical band heteretopia.

Declarations

Consent statement: Written informed consent must be obtained from the patient (or legal guardian) for publication, including images.

Conflict of interest statement: The authors declare that they have no known competing financial interestsor personal relationships that could have appeared to influence the work reported in this paper.

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Ibtissam El Ouali: Contributed to the conception of the case report, acquiring and interpreting the data and undertaking the literature review

Yahya El Harass: Contributed to drafting the manuscript, acquiring and interpreting the data and undertaking the literature review

Lina Belkouchi: Contributed to undertaking the literature review and revising the article critically for important intellectual content.

Siham El Haddad: Contributed to undertaking the literature review and revising the article critically for important intellectual content.

Nazik Allali: Contributed to undertaking the literature review and revising the article critically for important intellectual content.

Latifa Chat: Contributed to patient care, undertaking the literature review and revising the article critically for important intellectual content.

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