SUBCORTICAL BAND HETEROTOPIA: CLINICAL AND IMAGENOLOGICAL CHARACTERISTICS

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INTRODUCTION

Subcortical band heterotopia (SBH) is the least common malformation of cortical development and is included in the lissencephaly spectrum. Genetically, it is related to DCX and LIS1 genes. Clinically it presents with drug-resistant epilepsy, intellectual disability and behavioral, learning and language disorders.

OBJECTIVE

To present the clinical and imaging characteristics of 13 patients with a diagnosis of Subcortical band heterotopia.

METHOD

Retrospective, descriptive study based on a review of medical records from 2010 to 2023.

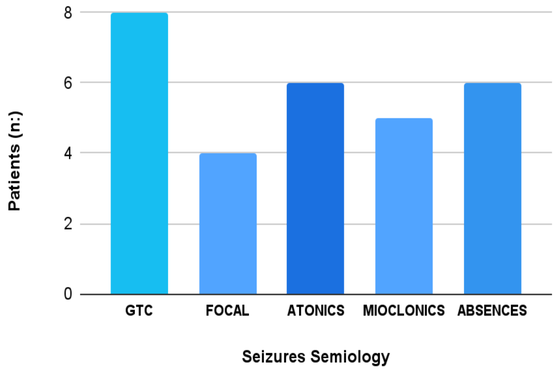
MATERIALS AND METHODS

Patients: n: 13
Age Range: 3 m - 16 y
Female: 76,92% (n:10)

All of them were diagnosed in the context of epileptic debut (average 4 years 7 months). 62% had a pattern of epileptic and developmental encephalopathy, being Lennox Gastaut Syndrome the most frequent (75%). The seizure semiology in descending order was: generalized tonic-clonic seizures, absences, atonic/drop attacks, myoclonus and focal seizures. 62% of the cases evolved into drug resistance. None of the patients has genetic confirmation at the moment (7 out of 13 patients have pending results).

EPILEPSY (n:13)	
EPILEPTIC ENCEPHALOPATHY	61,53% (n:8) n:6 LGS n:2 IESS
DRUG RESISTENT	61,53% (n:8)
AVERAGE OF ASM	5.4 (2 - 10)
VNS	37.5% (n:3)

Table 1. Clinical features of patients diagnosed with epilepsy.

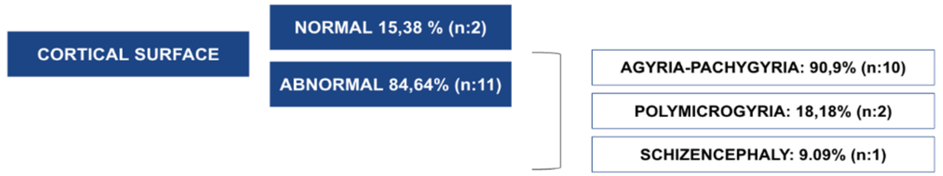


Graphic 1. Semiology of seizures in patients diagnosed w/ epilepsy.

The images showed that the most frequent gradient was anterior or global. 85% presented an abnormal superficial cortex with a predominant pattern of agyria - pachygyria and lissencephaly. Only 2 patients presented polymicrogyria. 92% presented other findings: anomalies in the corpus callosum (92%), ventricular dysmorphia (83%) and cerebellar hypoplasia (42%).

DOMINANT PATTERN	1 = ANTERIOR OR GLOBAL	2 = POSTERIOR	3 = LIMITATED OR UNILATERAL
(n)	**9**	**4**	**-**
(%)	**69,23 %**	**30,7%**	**-**

Table 2. Dominant heterotopic band pattern. Classification taken by Subcortical laminar (band) heterotopia. Chapter 12. Handbook of Clinical Neurology, Vol. 87 (3rd series). Malformations of the Nervous System Teruyuki Tanaka and Joseph G. Gleeson



Graphic 2. Imaging characteristics of the upper cerebral cortex.

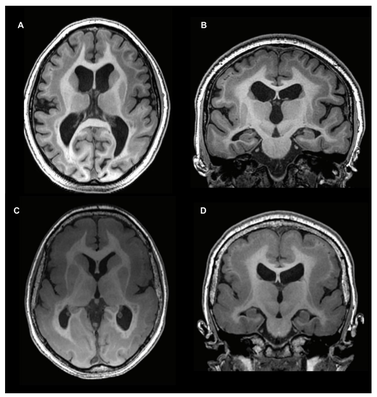


Image 1. Axial and Coronal section T1.
(A & B): Band of gray matter parallel to the cortex, separated from it by a thin layer of white matter in relation to predominant band heterotopia at the frontal and parietal level, associated with poor deepening of sulci.

(C & D): Diffuse abnormal rotation pattern of the cerebral cortex associated with agyria. Extensive thick subcortical band isointense to the gray matter parallel to the cortex showing antero-posterior gradient.

CONCLUSION

Subcortical band heterotopia is a rare malformation of cortical development.

Epilepsy was the reason for consultation and the reason for diagnosis in all cases.

In more than half of the patients, epilepsy progressed to epileptic encephalopathy and was associated with drug resistance.

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