

Case report

Dual Cortex: Subcortical Band Heterotopia.

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Abstract:

Subcortical band heterotopia (SBH) is a disorder of neural migration. Also called as double cortex syndrome due to it's appearance. Patient presents with mental retardation and epilepsy. Usually seizures start in first decade of life and may vary between focal seizures to generalized seizures. MRI is the diagnostic investigation of choice which reveals the characteristic findings. It's a rare disorder with only a few hundred cases reported till date.

Key words: Subcortical band heterotopia, Epilepsy, Mental retardation

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Introduction: SBH is a syndrome of cortical malformation. It is characterized by presence of bilaterally symmetrical, heterotopic grey matter which is located between the ventricles and the cortex. Females are predominantly affected.

Patients typically present with mental retardation and epilepsy. It is a rare disorder with only a few hundred cases reported in literature till date.

Case report: A 19 years female reported to our out-patient department with history suggestive of partial

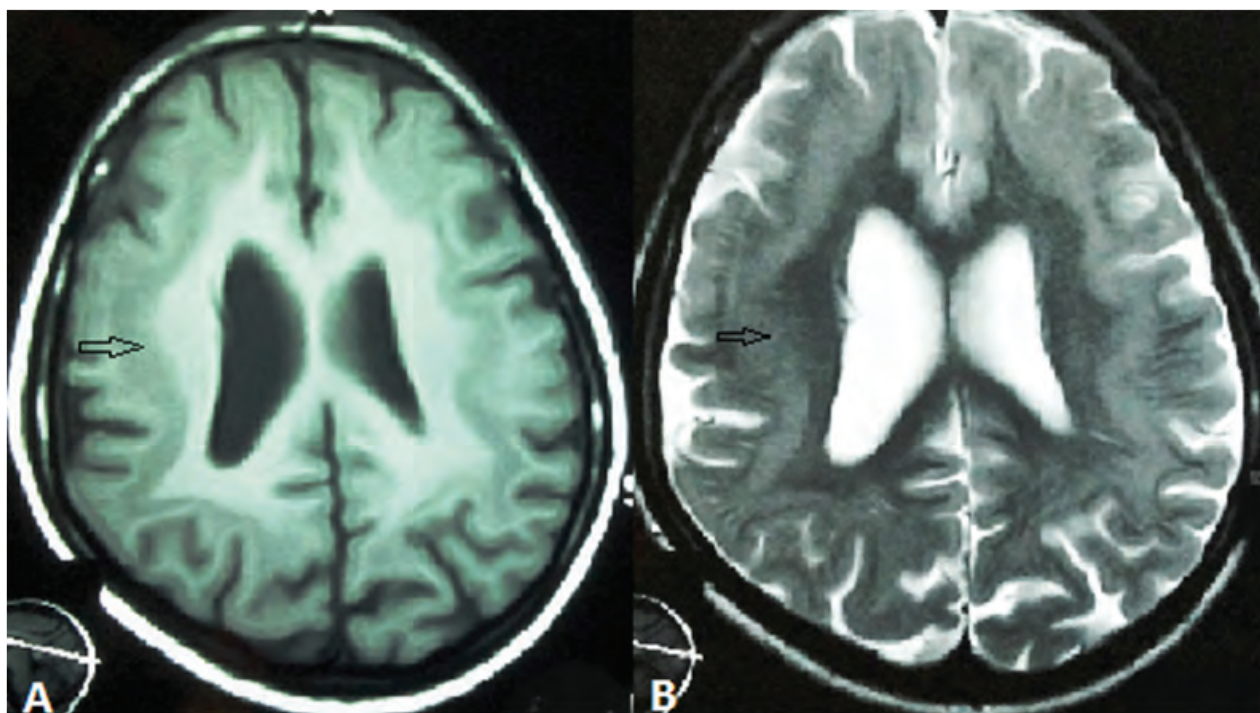


Figure 1 (A,B): MRI Brain(T1 and T2 images) –Arrow showing sub-cortical gray matter separated from the ventricles and cerebral cortex by layer of normal appearing white matter

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seizures, complex partial seizures and generalized tonic-clonic seizures since 6 months of age. She was 2nd in birth order and was born out of non-consanguineous marriage. She was borne out of full term normal vaginal delivery. There was no history suggestive of peri-natal distress or complications. She had delayed development milestones with poor scholastic performance. Her usual frequency of seizures was 6-7 episodes per month. No medical attention was sought for the same till the age of 19 years when she first presented to us. On examination she had low IQ. Electroencephalogram showed multifocal spike discharges. Cranial MRI showed bilaterally symmetrical thinning of the outer cerebral cortex with formation of sub cortical gray matter separated from the ventricles and cerebral cortex by a layer of normal appearing white matter (Figure.1). Patient was started on antiepileptic drugs. Her seizure frequency has decreased and is on our follow up for drug dose titration.

Figure 1 (A,B): MRI Brain(T1 and T2 images) – Arrow showing sub-cortical gray matter separated from the ventricles and cerebral cortex by layer of normal appearing white matter

Discussion:

Sub cortical heterotopias were first differentiated into two types (a) Nodular and (b) Laminar type heterotopia by Jacob¹. The third type of heterotopia termed as subcortical band heterotopias(SBH)

was first coined by Barkovich et al in 1989². The presence of gray matter as diffuse circumferential band in the subcortical area on MRI differentiated it from nodular and laminar type of heterotopia. Later on this MRI pattern was described as “double cortex syndrome” by Palmini et al³. MRI pattern of our patient was consistent with laminar heterotopias or ‘double cortex syndrome’. Till date only a few hundred cases of SBH has been reported in literature and most of the patients were females. Majority of the cases are sporadic in nature. Few cases of X-linked familial SBH too have been reported⁴

The genes which have been reported to be involved in SBH are

I. DCX-doublecortin gene, located on chromosome Xq22.3-q23. Nearly 80% of sporadic cases with SBH are associated with DCX mutation⁵
II. LIS1 mutation on chromosome 17p13.3⁶

DCX mutations are associated with predominant heterotopias over anterior region where as it is predominantly in occipital and parietal regions in cases of LIS1 mutations.^{7,8}.

Patients typically present with mental retardation and epilepsy. Our patient was a female and she too had history of mental retardation and epilepsy. Her Cranial MRI revealed SBH. It is a rare disorder which is now a day’s being picked up frequently due to wide availability of MRI.

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