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Corpus Callosum Agenesis presenting in an Adult

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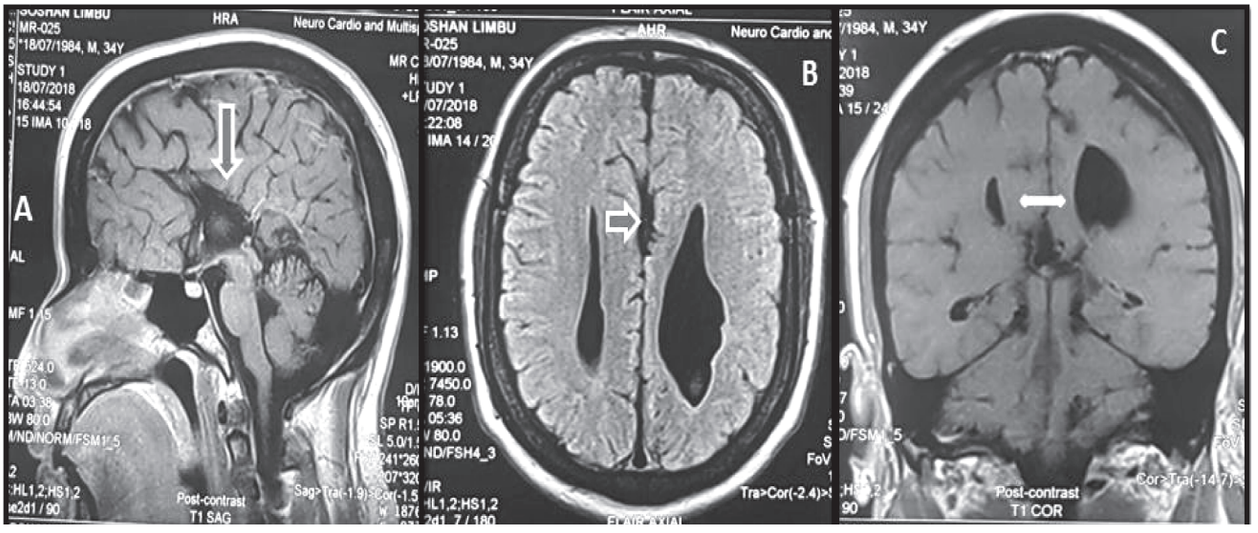

Figure 1. *MRI showing Complete CCA with high riding third ventricle (arrow) on sagittal section (A), parallel asymmetrical ventricles with Probst fibres in Axial section (B) and bull horn appearance of ventricle in coronal section with Probst fibres (arrow) (C).*

Corpus callosum is the interhemispheric fiber bundle which starts forming at 6 weeks of gestation and assumes its shape at 20 weeks. Corpus callosum agenesis (CCA) is one of the common congenital brain malformation (prevalence of 0.3-0.7 %) which can be associated with alcohol use during pregnancy, phenylketonuria and monogenic or complex chromosomal syndromes (del (4) (p16), del (6) (q23), dup (8) (p21p23), dup (11) (q23qter)). It can be pure CCA or partial dysgenesis along with Mowat-Wilson syndrome or Walker-Warburg syndrome. It can be either type 1 (forming large aberrant bundles that do not cross the midline known as Probst bundle or type 2 (the axons fail to form and hence no Probst bundles).[1,2,4] clinically these patients present with mental retardation,seizure, speech or learning disability, visual abnormalities, social and behavioral problems and feeding problems.Magnetic Resonance Imaging (MRI) remains the diagnostic modality which will show CCA or dysgenesis along with other congenital lesions like, Probst bundles, polymicro gyria,schizencephaly,lissenencephaly, heterotopia, Chiari II or Dandy –Walker malformation.[3]

A 34-year old male presented with history of seizure disorder since few years with irregular anti-epileptic treatment. He had 3 episode of seizure 3 days ago lasting for 5 to 6 minutes with up rolling of eyes and tightening of body. Post-ictal headache was present for 3 hours. He had been prescribed ani-epileptic medication which was left few months back. Examination showed conscious male with no motor or sensory deficit. He had no social-psychological problems, except for emotional liability and was married with one normal6-year old child. Except for occasional alcohol intake he had no co-morbid ailment. Routine hematological examination was normal and in view of his long standing seizure disorder he was advised MRI which showed gross cerebral atrophy with type 1 CCA

and asymmetrical lateral ventricle (Figure 1). Inter ictal Electroencephalogram was normal .He has been prescribed Levetiracetam 1000mg /Day and is seizure free till now.

CCA is a relatively common congenital disorder and can be associated with other brain anomalies.The patients can present with either, Type 1. Severe neuro-psychiatric disorders including growth, visual, hearing, verbal and mental retardation, Type 2. Moderate with autism, mental retardationand limited neuro-psychiatric deficits or Type 3. Mild with near normal range of neuro-psychiatric functions.[5] Our patient was the mild type which is very rare and is explained by the brain plasticity to function and perform task in the absence of some anatomic part. In the other twotype's family counseling and rehabilitation along with seizure control is advised. Children with mental retardation with or without seizure are advised to undergo MRI and if abnormal, VEP or BAER is recommended to assess other neurological deficits. Neuropsychological counseling can help in the management of these cases as most do not do well and suffer from developmental delay, intellectual disability, and neurologic deficit.[6]

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