Case Report

CORPUS CALLOSAL AGENESIS – AN INCIDENTAL FINDING

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ABSTRACT

Agenesis of the corpus callosum (CC) is an uncommon cerebral malformation that has been reported in 1 in 19,000 unselected autopsies and 2.3% of children with mental retardation. It is associated with a broad range of clinical manifestations, oscillating between the limits of the normal and severe psychomotor delay. Corpus callosal agenesis with asymptomatic presentation is rare. We present here a case of a 40 year old male who was apparently asymptomatic before presented with continuous headache following a minor head injury. CT scan of brain was done and it revealed complete corpus callosal agenesis which was picked up as an incidental finding. Further MRI was done to confirm and to study the entire spectrum of findings.

Keywords: Corpus Callosal Agenesis, Lamina Terminalis, Colpocephaly

INTRODUCTION

The corpus callosum is a white matter structure connecting the cerebral hemispheres and is important in coordinating information and bilateral exchange of sensory stimuli. It is derived from the lamina terminalis in the portion of the neural tube cephalic to the rostral neuropore. Agenesis of the corpus callosum is an uncommon cerebral malformation that has been reported in 1 in 19,000 unselected autopsies and 2.3% of children with mental retardation (Grogono, 1968; Freytag and Lindenberg, 1967). Insults responsible for agenesis of the corpus callosum or varying degrees of hypoplasia of the corpus callosum are not identified. It can present with various spectrum varying from agenesis of the corpus callosum without substantial involvement of other brain areas or with interhemispheric cyst, lipoma, or partial agenesis (hypogenesis) of the corpus callosum. The defect may be complete or partial, depending on the stage at which callosal development is arrested. Until the fourth month of gestation, only the most rostral part of the corpus callosum is formed; the caudal portion develops only after the fifth month (Loeser and Alvord, 1968). An early failure may lead to complete agenesis, whereas a later one will lead to hypoplasia (Loeser and Alvord, 1968).

CASES

A 40 year old male presented to casualty with alleged history of attack by paper roll over his head 2 days back followed by continuous headache. No history of loss of consciousness or ENT bleeds. On examination he was conscious, oriented with normal intelligence. No evidence of motor or sensory deficit Vitals were stable. No focal neurological deficit or meningeal signs. CT scan Brain was done as a workup for headache.

Unenhanced axial sections of CT scan revealed:
- No evidence of intraparenchymal/extra axial bleed
- No evidence of calvarial fracture
- Complete absence of corpus callosum with parallelly oriented ventricles.
- Dilatation of posterior occipital horns - colpocephaly (Figure 3a,b)

Further MRI was done to evaluate brain for complete outline of corpus callosum spectrum and it showed
- Parallel non converging widely separated lateral ventricles on axial sections, which were the most striking feature (Figure 4a, b).
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- Dilated Occipital horn – Colpocephaly
- Pointed or trident shaped frontal horns.
- Non visualization of normal stripe of CC on mid sagittal sections, gyri and sulci directly radiating from roof of third ventricle (Figure 1a, b)
- High riding third ventricle.
- Absent cingulate gyrus which is normally seen parallel and cranial to CC.
- Vertically originated hippocampi with key hole appearance of temporal horns on coronal sections (Figure 2a, b).
- Probst bundles, the abnormal longitudinally orientated band of white matter track medial to lateral ventricles seen on coronal sections.

Figure 1a, b: Saggital t2w mri sections shows non visualization of normal stripe of corpus callosum on mid sagittal sections, absent cingulate gyrus, gyri and sulci directly radiating from roof of third ventricle

Figure 2a, b: T2W MRI Coronal sections showing pointed or trident shaped frontal horns. probst bundles vertically originated hippocampi with key hole appearance of temporal horns
**Figure 3a, b:** Axial sections of unenhanced CTScan brain shows (ring artifact noted)

- Parallel non converging widely separated lateral ventricles
- Dilated occipital horns - colpocephaly

**Figure 4a, b:** Axial sections of t1w and t2w sequences shows colpocephaly and a ‘rabbit ear appearance’ or ‘devils horn appearance’

**DISCUSSION**

Agenesia of the corpus callosum is a rare birth defect (congenital disorder) in which there is a complete or partial absence of the corpus callosum. It occurs when the corpus callosum, the band of white matter connecting the two hemispheres in the brain, fails to develop normally, typically during pregnancy. The development of the fibers that would otherwise form the corpus callosum become longitudinally oriented within each hemisphere and form structures called Probst bundles.
In addition to agenesis of the corpus callosum, other callosal disorders include Hypogenesis (partial formation), dysgenesis (malformation) of the corpus callosum, and hypoplasia (underdevelopment) of the corpus callosum. Callosal disorders can be diagnosed only through a brain scan. They may be diagnosed through an MRI, CT scan, prenatal ultrasound, or prenatal MRI. It can present with various spectrum varying from agenesis of the corpus callosum without substantial involvement of other brain areas or with interhemispheric cyst, lipoma, or partial agenesis (hypogenesis) of the corpus callosum. The differential diagnosis includes Arachnoid cyst, porencephaly, hydrocephaly, and prominent septum cavum pellucidum. In a study by Bedeschi et al., (2006) in 63 young patients, neuromotor skills were impaired in almost all cases (58/63). Mental retardation of different severity was present in 52 cases, whereas 2 patients were borderline and 9 patients had normal intelligence quotient. The study demonstrated that there is no unique prognosis for agenesis of the corpus callosum as this condition is associated with a broad range of clinical manifestations, oscillating between the limits of the normal and severe psychomotor delay.

**Conclusion**

Corpus callosal agenesis is usually associated with broad range of clinical manifestations and is rarely picked up as an incidental finding in CT/MRI scans.

**REFERENCES**


