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Agenesis of Corpus Callosum in an Elderly Male

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Conflicts of Interest: Nil.

Introduction

Agenesis of Corpus callosum is one of the commonest neurogenic congenital abnormalities reported in the literature. It is known to be associated with various other congenital malformations viz Arnold Chiari, Aicardi syndrome, Fetal alcohol syndrome, schizencephaly and certain trisomies. These patents may manifest with various serious neuropsychiatric symptoms such as mental retardation, seizures, hydrocephalus while others may present with subtle symptoms.

While most of the available literature describes this condition in the children and young adults, we present a case of Agenesis of Corpus Callosum in an elderly male.

Case report:

A 69yr old male patient was brought to our hospital with sudden onset loss of consciousness and unresponsiveness to verbal commands for about 2-3hrs. He was a known hypertensive and diabetic and was on regular medication for the same. On examination the patient was conscious but drowsy and disoriented. His vitals were within normal limits. His GCS was – 9/15 (E 3,M4,V2). His biochemical parameters didn't reveal any significant abnormality. An urgent non contrast computed tomography (NCCT) scan head was done to rule out intracranial hemorrhage followed by contrast enhanced computed tomography (CECT) the following day. The neuroimaging revealed complete absence of corpus callosum associated with parallel configuration of the lateral ventricles, colpocephaly with apparent hemimegalencephaly on the left side (Figure 1), high-riding IIIrd ventricle communicating with the interhemispheric subarachnoid space (Figure 2). On sagittal image the diverging sulci and gyri presented a "Sunray appearance" (Figure 3). No dorsal interhemispheric cyst was noted. No other associated abnormality was noted within the brain. Based on these findings a diagnosis of Agenesis of Corpus Callosum was given.

The patient was managed conservatively in the ICU for the next 10days and then discharged once his physical condition improved.

Discussion:

Corpus callosum is the principal commissure in the brain through which the axonal bundles cross over to the contralateral hemisphere. Embryoligically it develops from lamina reuniens of His in the forebrain starting from6- 8 wks of gestation and completes with the formation of rostrum by 18 – 20 wks of gestation [1,2].

Agenesis of corpus callosum can be partial or complete, depending upon the gestational age at which the insult has occurred. It can be isolated or associated with other neurological and systemic congenital abnormalities like Arnold Chiari malformations, Aicardi syndrome,Acrocallosal syndrome, Andermann syndrome, Shapiro syndrome, Porencephaly, Schizencephaly, Fetal alcohol syndrome and certain trisomies[3,4]. It can manifest in mild form wherein the patients may have normal intelligence and in its severe form the individuals may have seizures, mental retardation developmental

delay, ocular anomalies, gyral abnormalities, hydrocephaly and spasticity [3-5].

Its prevalence is 0.05-0.7% of the general population and 2.3% of children with developmental disabilities. Corpus callosum agenesis (CCA) patients can be asymptomatic with incidental detection of the condition during neuroimaging and prevalence of such asymptomatic CCA is unknown, but based on autopsy series or CT studies, it is estimated to be 0.5/10,000 or 0.13- 0.7%, respectively. With increasing frequency of neuroimaging this entity has become one of the common findings in the present era [6,7].

It can be diagnosed antenatally by sonography or magnetic resonance imaging (MRI) [8]. MRI is considered as the preferred modality of imaging [6]. In a patient with complete agenesis of Corpus calllosum, the neuroimaging reveals absence of corpus callosum with consequent radiating spoke-like or “sun ray appearance” of cortical gyri on the sagittal image, widely separated frontal horns of the lateral ventricles giving a “bat-wing” or “bull-horn appearance” also referred to as “Racing car sign”. Dilated occipital horns also known as colpocephaly associated with narrow frontal horns give the appearance of a “tear drop”. The associated features are a high riding IIIrd ventricle which continues with the interhemispheric fissure with or without the presence of an interhemispheric cyst,. The longitudinal white matter tracts which are otherwise supposed to cross over are seen adjacent to the lateral ventricles paralleling the interhemispheric fissure and are termed as “Probst bundles”. These Probst bundles are seen to protrude into the superomedial walls of the frontal horns best appreciated on coronal MR images and is known as Viking’s hemlet sign. In the coronal section the interhemispheric fissure is in continuity with a dilated and dorsally elevated third ventricle sometimes giving a “Moose head appearance” [9-11].

Majority of the patients are diagnosed with this condition during their childhood as they present with developmental delay, slow mentation or other associated physical deformities like increasing skull size in case of hydrocephalus, microcephaly and cleft lip or palate. However certain individuals go undetected as they remain asymptomatic for most part of their life and our patient belongs to such population[12-14].

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