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Case Report

Corpus callosum hypogenesis with interhemispheric cyst: a case report

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ABSTRACT

Agenesis or hypogenesis of Corpus callosum with interhemispheric cyst is a rare entity. Origin of interhemispheric cyst is controversial. These cysts may be Arachnoid cyst, neuroepithelial cyst, or extension of lateral or third ventricles. Here we describe a case of Corpus callosal hypogenesis with an interhemispheric cyst. Associated aqueductal stenosis and schizencephaly is also present. Interhemispheric cyst is communicating with lateral ventricle and the cyst has MRI signal intensity similar to CSF. This case supports the theory that the interhemispheric cyst is an ependymal cyst and is consequence of increased intraventricular pressure due to aqueductal stenosis.

Keywords: Corpus callosum, Agenesis, Interhemispheric cyst

INTRODUCTION

Agenesis of corpus callosum is one of the most commonly encountered and probably one of the most genetically diverse brain malformations.

It is associated with many chromosomal abnormalities and malformation syndromes as well as to several inherited metabolic diseases.

ACC or HCC associated with interhemispheric cyst is a rare but well recognized entity. These cysts may be Arachnoid cyst, neuroepithelial cyst, or extension of lateral or third ventricles lined by ventricular epithelium.

Ability of MRI to show gross morphological anatomy of brain clearly in multiple planes allow presumptive diagnosis of cyst histology and origin. Here we present a case of Corpus Callosum Hypogenesis (HCC) with a large interhemispheric cyst communicating with lateral ventricle and associated schizencephaly.

CASE REPORT

A 5 days old male infant with macrocephaly and signs of increased intracranial tension was referred from paediatric department for cranial USG. During ultrasonography we found dilated lateral ventricles and large interhemispheric cystic structure. Corpus callosum was not visualized. Posterior fossa appeared normal. The patient then underwent MRI of brain for further characterization of the cerebral malformation. MRI of brain confirmed hypogenesis of Corpus Callosum (only genu and anterior body was present). Lateral and third ventricles were dilated. A large CSF intensity cyst was present in right paramedian location communicating with the right lateral ventricle without any intervening septa. Near inner table of calvarium cyst was limited by a thin wall. Small open lip schizencephaly was present in right parietal region. White matter volume was reduced in both the cerebral hemisphere. Falx was normal. Fourth ventricle was not dilated and posterior fossa structures were normal. During pregnancy patient did not underwent antenatal sonography check-up.

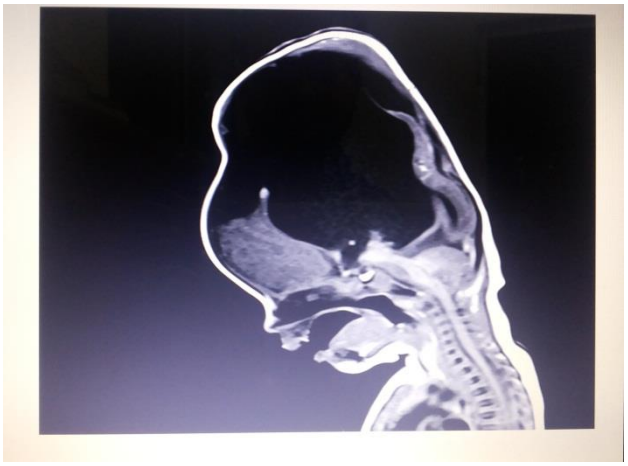

Figure 1: Sagittal T1-weighted image in the midline shows hypogenesis of corpus callosum. Only genu and anterior part of the body is present. A large CSF intensity cystic area is seen dorsal and posterior to the genu of corpus callosum. Skull is tower shaped. Cerebellum and tentorium are compressed inferiorly by the cyst.

DISCUSSION

Corpus callosum is the principal supratentorial cerebral commissure. The development of the corpus callosum begins in the 8th week of fetal life anteriorly at the genu and continues posteriorly. The rostrum develops last, usually folding back under the genu. The mature corpus callosum is developed by the 20th week of gestation.[1,2] It is the most tightly packed bundle of axons in the brain and helps the ventricles to maintain their normal size and shape. It has traditionally been devided into four segments: the rostrum, genu, body and splenium.

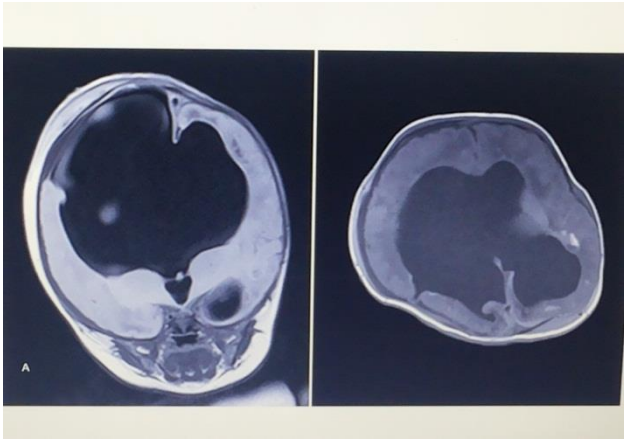

Figure 2: Coronal and axial T1-weighted images show the cyst is in right paramedian location and continuous with the right lateral ventricle. In the region of the cyst cortical mantle is severely thinned out into a membrane. Third ventricle is dilated.

Anomalies of corpus callosum may be agenesis (complete absence of corpus callosum), hypogenesis (later formed segments missing but earlier formed segments present), hypoplasia (all segments are present but small).[3-7]

Interhemispheric cyst associated with Corpus Callosum agenesis or hypogenesis is a special and rare condition that may have a different cause than other types of callosal agenesis.[8] Origin of the interhemispheric cyst in agenesis of corpus callosum is controversial. On the basis of cyst location, morphology, and imaging characteristics, intracranial cysts associated with ACC are most likely arachnoid, neuroepithelial, or ependymal in origin.[7,9] Characteristics of intracranial cysts on sonography and MR imaging can lead to a presumptive diagnosis of cyst histology.[9]

Barkovich et al.[7] divide Agenesis of corpus callosum with Interhemispheric cyst into two major groups. Type-I, in which the interhemispheric cyst is a diverticulum of the ventricular system and thus communicate with the ventricles and type 2, in which multiple cysts are present that donot communicate with the ventricles. Type 1 cysts are isointense to CSF, whereas type 2 cysts are usually slightly hyperintense to CSF on T1 weighted images.[8]

According to Barkovich et al. classification[7] this case is an example of type-I cyst. Here the cyst is communicating with the right lateral ventricle without any intervening septum and cyst fluid is isointense to CSF in all MRI sequences. This suggests that the cyst is an outpouching of right lateral ventricle and it is an ependymal lined cyst. In this case patient presents with macrocephaly, third and lateral ventricles are grossly dilated and fourth ventricle is normal. These also support the theory that type-I cysts are result of increased intraventricular pressure, which in this case is due to blockage of aqueduct of sylvius and the hydrocephalus is noncommunicating. Considering the above facts we categorize the cyst as type-I of Barkovich Classification.

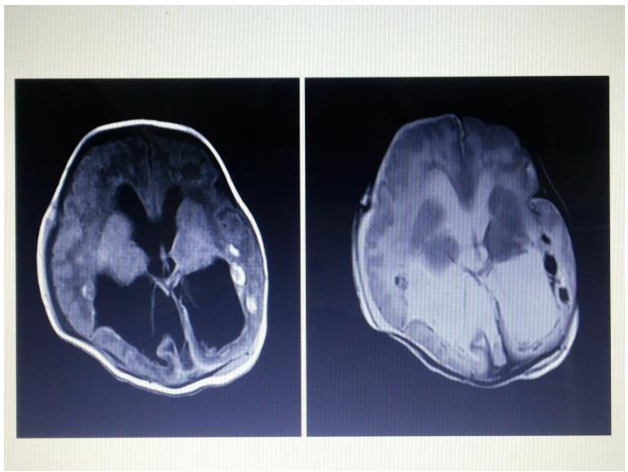

Figure 3: Axial T1 and GRE images show dilated lateral ventricle, mostly the occipital horns. A small schizencephaly is present in right parietal region. Periventricular small hemorrhagic foci are present.

When the corpus callosum is absent, loose white matter surrounds the posterior bodies, trigones, occipital horns and posterior temporal horns of the lateral ventricles.[10]

In corpus callosum hypogenesis, therefore, the posterior portion of the ventricles expand, resulting in dilatation of the trigones and occipital horns of the lateral ventricles. This configuration of the ventricles has been termed colpocephaly.[11]

However, firm caudate and lentiform nuclei keep the size of the frontal horn relatively small in the abscess of corpus callosum, but their lateral borders are convex instead of normal concavity. Other features that are well seen on axial images include lateral convexity of frontal horns, parallel lateral ventricles, upward extension of the third ventricle into interhemispheric fissure between the lateral ventricles, Probst bundles along medial wall of frontal horns.[8]

Hetts et al.[12] in a study of 142 patients with ACC and HCC found that frequency of interhemispheric cysts were similar in ACC and HCC. Among 142 patients 20 patients had interhemispheric cysts. Of which 11 cysts were communicating with ventricles (Type I) and 9 did not (Type-II).

The incidence of Type-I cysts are high in male patients. More careful analysis of the male:female ratio suggests that some types of callosal anomalies with interhemispheric cysts are more common in males whereas others are more common in females. This finding again emphasizes that callosal agenesis with interhemispheric cyst is not a single malformation, but a heterogeneous group of disorders that have in common callosal agenesis or hypogenesis and a midline cyst.[13]

CONCLUSION

ACC or HCC with interhemispheric cyst is a rare condition. MRI plays an important role detecting the condition and helps in the evaluation of morphology and origin of the cyst, hence helps in further management of the condition.

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