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GIANT INTERHEMISPHERIC FISSURE CYST WITH MULTIPLE CEREBRAL ANOMALIES: A CASE REPORT

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ABSTRACT
Interhemispheric fissure Arachnoid cysts are rare findings and their association with corpus callosal agenesis is quite exceptional. The origin of interhemispheric cyst with corpus callosal agenesis is controversial. We report a very rare case of giant interhemispheric arachnoid cyst associated with complete agenesis of corpus callosum, dysgenesis of falx cerebri, diencephalic malformations and significant cerebral atrophy in a newborn who presented with macrocrania. To the best of our knowledge, this is the first case report in literature, showing giant interhemispheric fissure cyst with multiple cerebral malformations.

INTRODUCTION

Interhemispheric fissure cysts are rare lesions, often associated with complex brain malformations such as corpus callosal agenesis and hydrocephalus [1,2]. It is defined as, cystic collection in the interhemispheric fissure with or without communication with the ventricular system. The prevalence of interhemispheric fissure cyst in association with corpus callosal dysgenesis is not known [3,4]. Fifteen cases have been described between 1937 & 1992, among which most of them were young children. Very few cases have been reported in foetuses and newborns. Although exact aetiopathogenesis is unclear but, it may result from X-linked recessive syndrome, trisomy 13,15 &18, Aicardi syndrome as well as vascular, traumatic, infectious or toxic insults [5,6]. The superior extension of the roof of third ventricle in interhemispheric fissure gives the origin to interhemispheric fissure cysts, which may or may not have its communication with third ventricle. The presence of choroid plexus tissue (originating from third ventricle) is responsible for CSF content of the cyst. Apart from dysgenesis/agenesis of corpus callosum, the interhemi spheric cysts may be associated with Dandy-Walker malformation, gray matter heterotopias, encephalocoeles, lipomas and midline facial anomalies. The children with interhemispheric cysts and corpus callosal agenesis have poor prognosis, with gross mental and motor deficits.

CASE REPORT

A 4 days old male baby presented with abnormally large head size as well as signs and symptoms of raised intracranial tension. It was a male term baby, born vaginally to a 30 years old multigravida mother, with no complications in 37th week of gestation. His head circumference was 38 cm, length 52 cm and weight 2400 grams. Anterior fontanelle was tense, bulging and GCS score 9 ($E_2V_2M_5$). Non-contrast CT scan head showed midline hypodense mass lesion, producing mass effect and cortical mantum is thinned out. Cyst is not communicating with ventricular system with mild dilatation of frontal horns and third ventricle visible. Falx cerebri is shifted to left side due to mass effect. Contrast MRI brain showed large cystic lesion in midline occupying almost entire

cerebral hemisphere. It was associated with complete agenesis of corpus callosum, partial dysgenesis of falx cerebri, diencephalic malformations and marked thinning of cerebral mantum.

Bilateral frontal horns and third ventricle is separately visible from the mass and mildly dilated. Fourth ventricle, posterior fossa and brainstem structures are compressed from mass effect of interhemispheric cyst. MRI findings were consistent with Type 2 interhemi spheric cyst. Surgical intervention to decompress the cyst was planned. Right sided Cyst-Peritoneal shunting was done. The post-operative course was uneventful and the patient's condition improved. One month after surgery, there was significant reduction in head size and anterior fontanelle was lax. Patient is under regular follow up, improving gradually but milestones are delayed, which signifies underlying cerebral atrophy and mental retardation.

Figure 1. Noncontrast CT Head showing midline hypodense mass lesion, producing mass effect and cortical mantum is thinned out. Cyst is not communicating with ventricular system with mild dilatation of frontal horns and third ventricle visible. Falx cerebri is shifted to left side due to mass effect

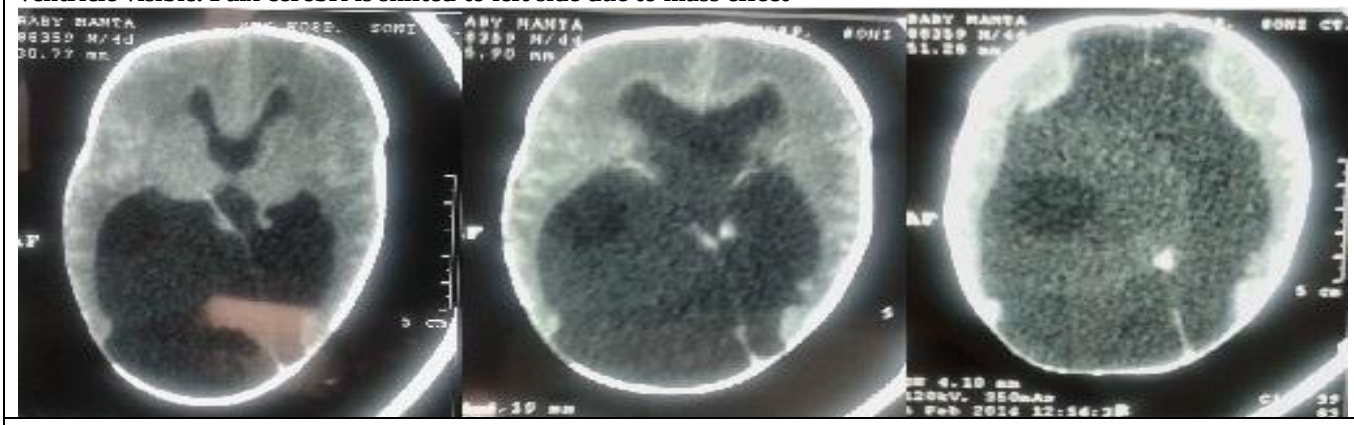
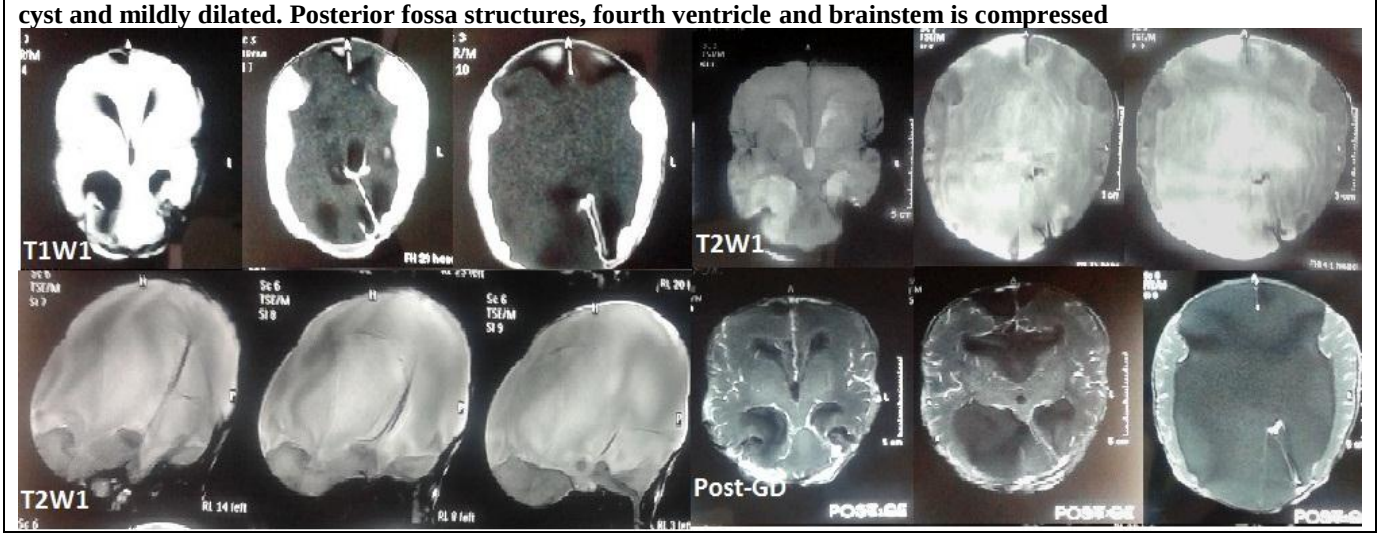


Figure 2. MRI Brain showing cystic midline lesion, occupying whole cerebral hemisphere, which is hypointense on T1W1, hyperintense on T2W1, isointense to CSF with CSF flow artefacts, showing no abnormal contrast enhancement. There is complete agenesis of corpus callosum, dysgenesis of falx cerebri, diencephalic structures are not visualised and cerebral mantum is thinned out. Frontal horns and third ventricle is not communicating with the cyst and mildly dilated. Posterior fossa structures, fourth ventricle and brainstem is compressed



DISCUSSION

Arachnoid cysts are developmental collections of CSF contained within a lining leptomeningeal membrane. The overall incidence of interhemispheric arachnoid cyst is 0.05% of all atraumatic intracranial mass lesions [3] Almost all reported cases of interhemispheric cysts have associated corpus callosal dysgenesis. Our case is unique in the sense that, apart from complete agenesis of corpus callosum, giant interhemispheric cyst is associated with diencephalic malformations, dysgenesis of falx cerebri and cerebral atrophy. Embryologically, Arachnoid cysts probably arise as a result of anomalous splitting and duplication of the endomenix during formation of neural tube fold [1,7]. Corpus callosum develops from commissural plate that lies in close proximity to anterior

neuropore. The first fibres forming the corpus callosum appear anteriorly near the lamina terminalis at around 12 weeks and crossing of the fibres is completed by 22 weeks of geststion [3,5]. Interhemispheric cysts tend to be enormous size because of the accommodation of cyst by the brain and expanding calvarium. The lining of arachnoid cyst is usually hyperplastic arachnoid cells and thick layer of collagen [6]. There is no evidence of choroid plexus tissue, tumour or inflammation. The fluid content of cyst is clear, colourless and similar to CSF but protein content varies [4].

Based on morphology, Barkovich has broadly classified interhemispheric cysts into two subtypes [1]. Type 1 cysts appear to be an extension or, diverticulation of third or lateral ventricles whereas Type 2 cysts are loculated and do not communicate with ventricular system. Therefore Type1 cysts may present with communicating type of hydrocephalus without other cerebral malformations. Various types of associated cerebral malformations as, dysgenesis of falx cerebri, subependymal heterotopias, polymicrogyria, diencephalic malformations, cerebral hemispheric dysplasia/hypoplasia and corpus callosal dysgenesis/agenesis is more common in Type 2 cysts [1,7]. Regarding gender predilection, they are predominant in males. These cysts may be unilocular or multilocular.

Clinical symptomatology of interhemispheric fissure cyst depends on its size, rate of growth and associated cerebral malformations. A very slow growing cyst generally becomes symptomatic in late childhood or adulthood [3]. Children with large cysts associated with corpus callosal dysgenesis present with macrocrania, mental retardation, hydrocephalus, motor weakness, developmental delay and failure to thrive [4,6]. Patients with isolated interhemispheric cysts have good functional recovery and normal intelligence. Interhemispheric fissure cyst associated with corpus callosal dysgenesis have poorer prognosis, usually with gross mental and motor deficits [1,4]. Ultrasonography is useful tool in diagnosis of intracranial malformations in foetuses and neonates [7]. MRI brain is the best tool to diagnose the interhemispheric cyst, to delineate associated cerebral anomalies and to rule out the tumoural pathology [3,4].

Regarding management, when cyst is large, associated with hydrocephalus or macrocrania and discovered before 24 weeks of gestation, the termination of pregnancy should be offered to parents [6]. Antenatal shunting of cyst has not been attempted till date. Incidentally discovered or small sized cysts do not require surgery [1,2]. But when symptoms are significant and severe, surgical decompression of cyst must be warranted. Cyst-Peritoneal shunting is the best surgical treatment [6]. The drawbacks of shunt surgery are sub-dural hygroma, shunt dependency, postop seizures, infection and shunt blockage. Neuroendoscopic treatment in children is a new emerging option of treatment. Endoscopic fenestration of cyst wall (cystoventriculocisternostomy) is minimally invasive technique with reported success rate of 71% to 81% [2,5,7].

In endoscopic treatment patient should be watched for postop hyponatremia, as endoscopic processure is carried out in close proximity to supraopticohypophyseal tract [7]. Other complications are bleeding, subdural hygroma and diabetes insipidus [7]. Children with interhemispheric cysts treated within first 3 months of life have best prognosis [2,3]. Recurrence risk of interhemispheric fissure cyst after treatment is still not known [3,5,7].

Conflicts of interest: None declared.

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