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Case Report

Outcome of Surgery in a Patient with Blindness and Inter-Hemispheric Cyst: A Case Report

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Abstract

Interhemispheric cysts are rare congenital condition which is also associated with corpus callosum agenesis. Most patients present at neonatal age or early childhood with some neurological deficiency. Some patients may present at older age. Here we present a patient with acute blindness of the right eye and dimness of vision of the left eye. He had no mental retardation or other neurological deficit. His fundus showed bilateral papilloedema. His MRI showed an interhemispheric cyst with agenesis of the corpus callosum. He was urgently operated in this hospital. The cyst was approached through interhemispheric fissure. It was excised and communicated with the third ventricle. His post-operative period was uneventful. Immediately after surgery he had improvement of his vision over a period of two week. He could return to normal life following discharge. Therefore, prompt surgical intervention is important for saving vision of these patients.

Keywords: *Interhemispheric cyst; Agenesis of corpus callosum; interhemispheric approach; ventriculostomy, papilloedema.*

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Introduction:

Interhemispheric cyst is a benign condition. Cerebrospinal-fluid-filled interhemispheric cysts associated with corpus callosal agenesis are relatively rare lesions[1]. It is congenitally formed and is associated with agenesis of corpus callosum. The major findings are absence of the corpus callosum, a large interhemispheric cyst, and ventricular dilatation[2]. Many patients are reported to have mental retardation with this condition, but our patient was free from such condition.

The origin of the interhemispheric cyst in agenesis of the corpus callosum (ACC) is controversial. Neuroenteric, arachnoid, and ependymal cysts have all been suggested as possible causes[3]. There are reports that this occurs with Dandy-Walker Malformation[3]. Barkovich et al had classified the interhemispheric cysts. According to him, these are type I and type II, which were further subdivided Figure I[4].

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Subtype	Cyst Characteristics	Communication
Type 1a: Presumed communicating hydrocephalus	Unilocular	With lateral ventricles
Type 1b: Hydrocephalus secondary to diencephalic anomaly	Unilocular	With and obstruction of third ventricle
Type 1c: Small head size and cerebral hypoplasia	Unilocular	With lateral and third ventricles
Type 2a: No abnormality apart from ACC	Multilocular	No communication with lateral or third ventricles
Type 2b: Aicardi Syndrome	Hyperattenuation (CT), hyperintense (T1W MR), multilocular	No communication with lateral or third ventricles
Type 2c: Subcortical heterotopia	Multilocular	No communication with lateral or third ventricles

Figure 1: *Classification of ACC with interhemispheric cyst - Barkovich et al[4]:*

Common abnormalities in ACC patients included: mental retardation (MR), 85%; seizures, 42%; ocular anomalies, 42%; gyral abnormalities, 32%; hydrocephalus, 23%; other central nervous system (CNS) lesions, 29%; costovertebral defects, 24% [5]. But in our patient only dimness of vision in the left eye and blindness in the right eye was observed.

Here we report a single case of interhemispheric cyst with agenesis of the corpus callosum presented with blindness, and rapidly treated with surgical excision of the cyst.

Case Report

A 16 years old man hailing from Dhaka was brought to the Department of Neurosurgery at National Institute of Neurosciences & Hospital, Dhaka, Bangladesh with acute dimness of vision. The patient stated that he was completely well 7 days back. Then the patient developed pain and dimness of vision of both eyes. But for the last three days the patient was unable to see in his right eye. The patient also complained of dull headache for the last three days. The patient had no history of fever, trauma and loss of consciousness or seizure. He was seen by ophthalmologists and was sent to the neurosurgeons. On examination he was well alert. His GCS was 15, and his speech and gait was normal. His higher psychic function was normal. All his cranial nerves were intact except II Cranial nerve. His right eye was blind and at left eye visual acuity was finger count at two feet. On fundoscopic examination there was bilateral papilloedema (Figure 3).

Motor and sensory examination was normal, cerebellar signs were absent and there was no sign of meningeal irritation. His MRI revealed agenesis of corpus callosum with a large interhemispheric cyst compressing both the optic nerves (Figure II).

The patient was immediately admitted and was prepared for surgery. The cyst was approached through interhemispheric approach in the right fronto-parietal area. Dura was opened with base to the sinus. The right cerebral hemisphere was gently retracted. the operating microscope was brought in. The distal anterior cerebral artery was preserved. Then wall of the arachnoid cyst came into view. It was incised. Clear CSF had escaped. The cyst was totally excised from lateral walls. Then the deep part of the cyst was reached. The deep part of arachnoid was excised. A small ventriculostomy was done at the floor of the third ventricle in front of the mamillary bodies.

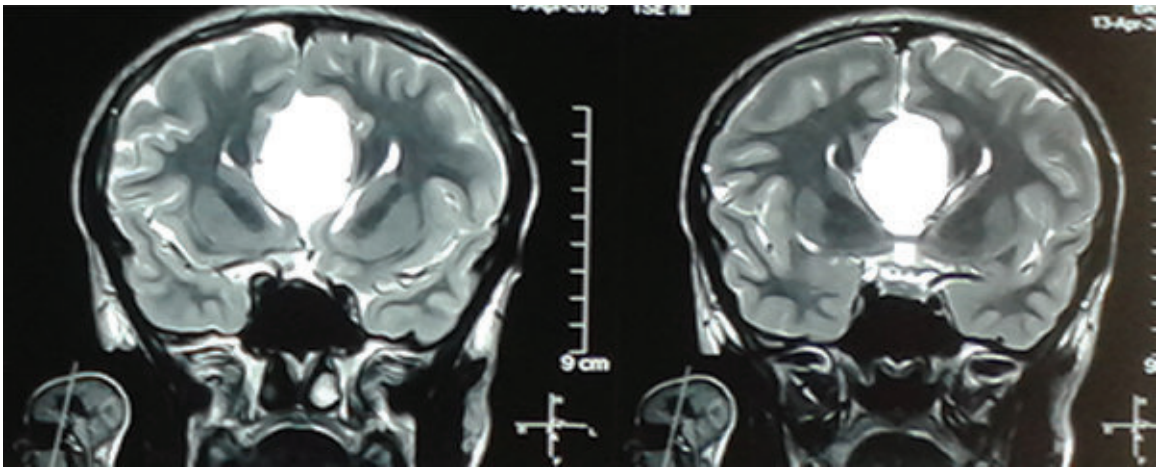

Figure 2: *T2WI coronal section shows interhemispheric cyst with agenesis of Corpus Callosum*

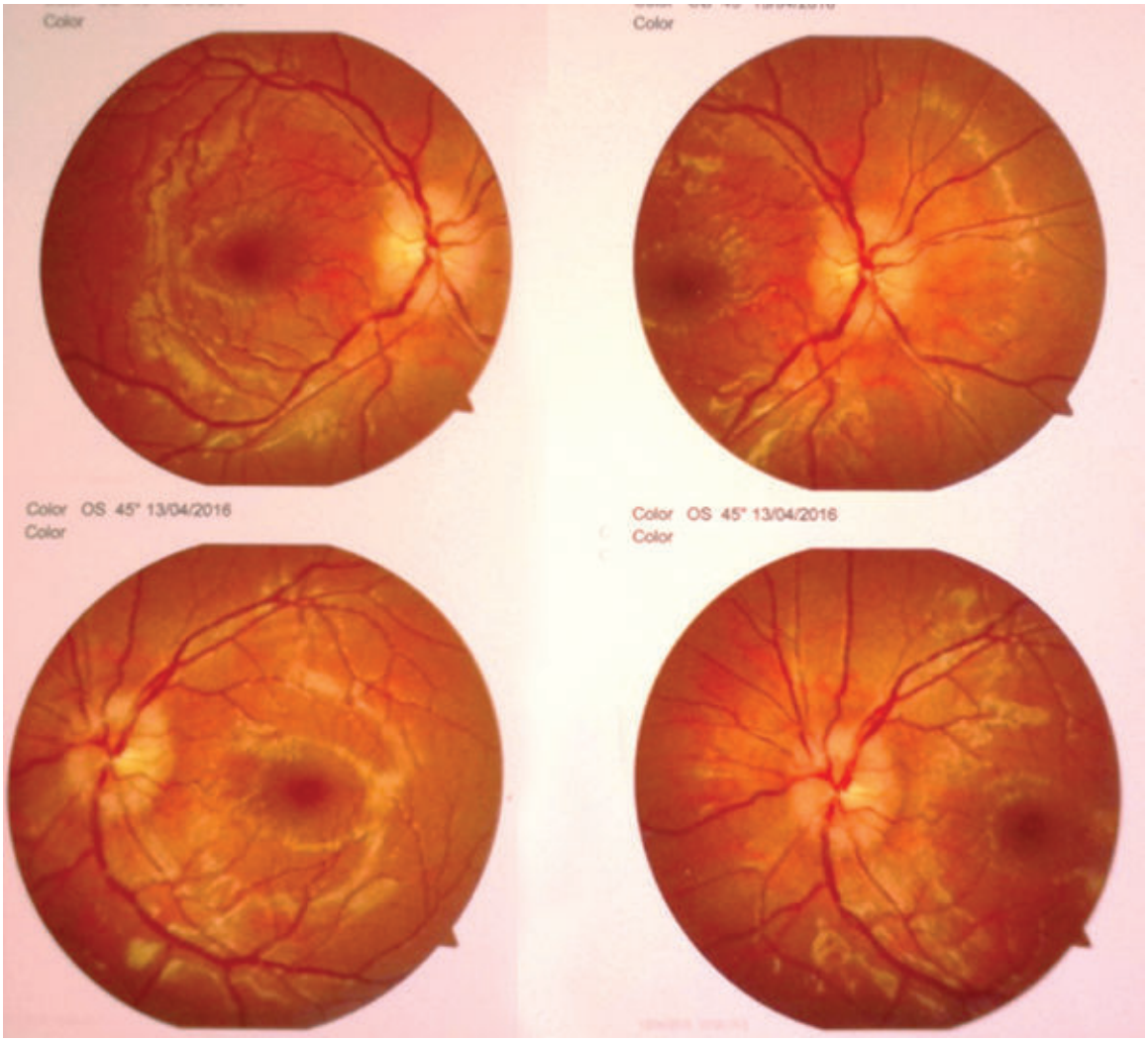

Figure 3: *Pre-operative Fundal Photograph showing Papilloedema*

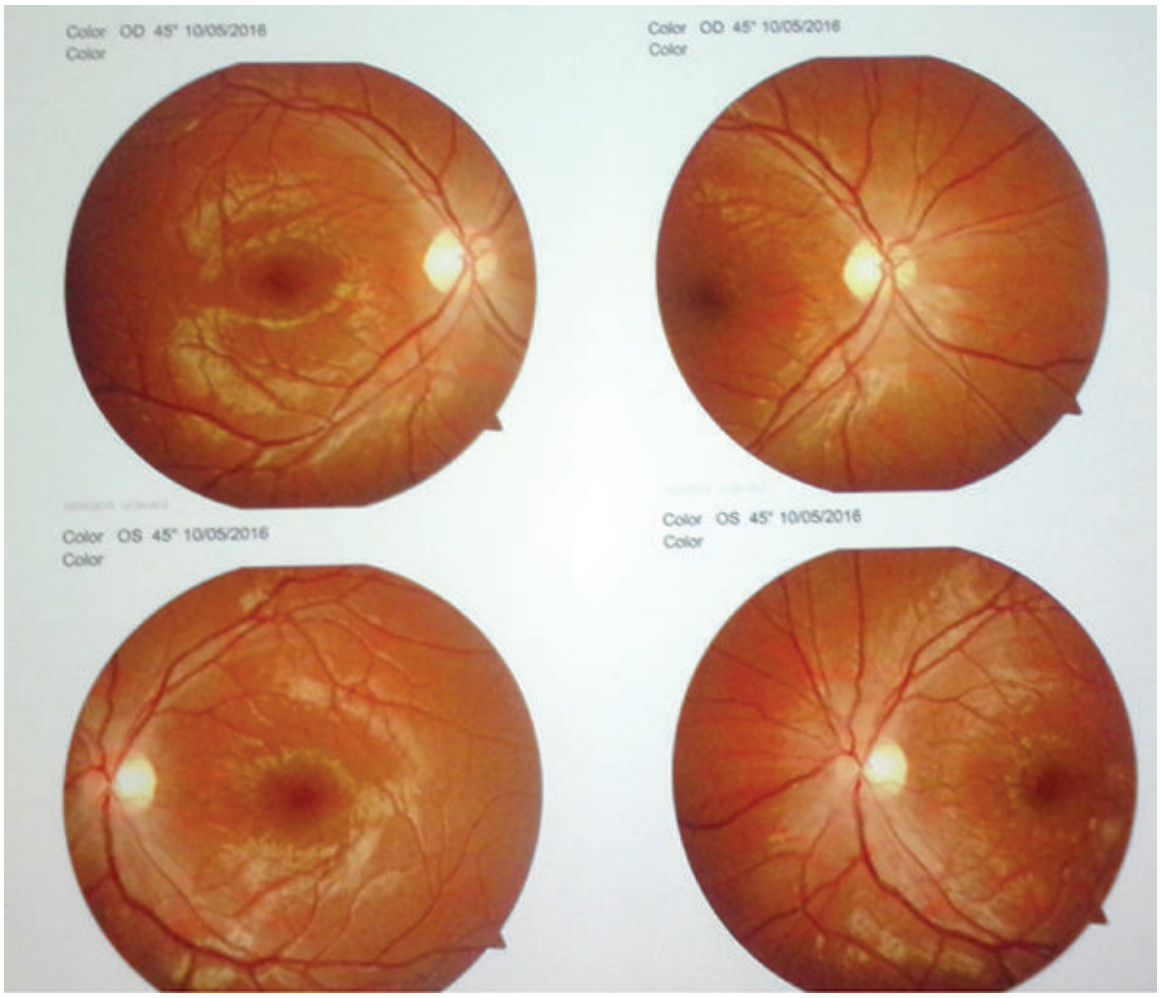

Figure 4: *Post-operative Fundal photograph showing improved optic disc*

Thus, the cyst cavity communicated with the third ventricle. Haemostasis was secured and dura was closed. Bone flap was replaced. A sub-galeal drain was placed and wound was closed in layers. His post-operative period was uneventful. His drain was removed on the third post-operative day. His visual acuity improved significantly. Just after surgery he had better vision in the left eye and perception of light in the right eye. After two days his visual acuity improved further with 6/6 on the left eye and blurred vision in the right. After 4th day of surgery, he had 6/6 vision of both the eyes. On fundoscopic examination, papilloedema of the patient had disappeared (Figure IV). The histopathologic examination commented that it was an arachnoid cyst. His stitches were removed on the 8th POD and he was discharged on the 19th. He came for follow up after six weeks and he had full recovery of his vision.

Discussion:

The inter-hemispheric cysts are usually presents in children. But this patient presented at adolescent age. Lena et al reported a series of 16 cases all were children[1]. However, Hirohata et al reported two cases in adults[6]. Neonatal diagnosis after one year was also reported[7]

All patients with interhemispheric cysts have agenesis of corpus callosum. Our patient also had agenesis of corpus callosum. But this posed no neurological problem to the patient. Visual disturbances are not the usual presentation. Solt et al have reported use of CT scan for the diagnosis in 1980[8]. But MRI is most sensitive due its ability to take thin slices and different phases[4]. This patient's interhemispheric cyst was approached through right interhemispheric approach and it was completely excised. In contrast Lena et al have reported treatment of sixteen patients with cystoperitoneal shunt[1]. In our patient, we had operated through the interhemispheric space and removed the cyst as much as possible.

Arachnoid cysts are thought to develop secondary to splitting or duplication of normal arachnoid membrane

during development. Most are diagnosed in childhood, with 50% reported before the age of 5 years[3]. But our patient had presented at the age of sixteen years.

According to Barkovich et al, Type 2a (multiloculated cysts) were associated with no abnormalities other than callosal agenesis/ hypogenesis[4]. In our case it was not multilocuted and there were no other abnormalities present other than agenesis of corpus callosum. So, we had concluded it was type IIa.

Interhemispheric cysts are unusually large and noncommunicating, and are often associated with agenesis of the corpus callosum Associated neurological deficits are minimal. Neuroradiological images mimic those of porencephalic cyst, holoprosencephaly, and agenesis of the corpus callosum. These anomalies should be differentiated clinically[9]. Our patient was similar in the sense that other than visual problems he had no neurological deficits.

This patient's interhemispheric cyst was approached through right interhemispheric approach and it was completely excised. In contrast Lena et al have reported treatment of sixteen patients with cystoperitoneal shunt[1]. In our patient, we had operated through the interhemispheric space and removed the cyst as much as possible.

Conclusion:

Interhemispheric cyst usually does not produce blindness. But our patient had presented in adolescence and with visual disturbance. Prompt surgery relieved all the visual symptoms and saved his vision. Therefore, early surgery and the interhemispheric approach is a safe and comfortable option for the patient.

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