



AN UNUSUAL CASE OF THIRD VENTRICULAR ARACHNOID CYST PRESENTING WITH INTENTIONAL TREMORS IN A CHILD

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ABSTRACT

Intraventricular arachnoid cysts are rare with intentional tremors being an unusual presentation of this cyst in a child. We report such a case of a large third ventricular arachnoid cyst in a 5-year old girl who was pre operatively diagnosed on magnetic resonance imaging of brain and underwent endoscopic fenestration of the cyst. 6 weeks post-surgery patient is doing well with significant reduction in tremors.

KEYWORDS : Arachnoid cyst - Intentional tremors - Hydrocephalus - Endoscopic fenestration

INTRODUCTION

Arachnoid cysts are common benign cerebrospinal fluid (CSF) containing cystic lesions lined by a layer of flattened arachnoid cells. Majority of these are considered to be a result of abnormal meningeal development where failure of endomeninges to merge during its embryogenesis results in accumulation of CSF in between the duplicated meningeal layers. Most arachnoid cysts are supratentorial and off midline, most common location being anteromedial to temporal lobe. Midline supratentorial arachnoid cysts are very rare with only a few cases of intraventricular midline supratentorial arachnoid cysts reported in literature. [1]

Arachnoid cysts can occur in any age group, however are most commonly found in children and young adults. These cysts can be incidentally detected in asymptomatic patients or symptoms may vary depending on location and size. [2]

Case Report

A 5-year-old girl presented to paediatrics OPD with complaints of intentional tremors progressively increasing with time over last 1 year and with new onset episodes of intermittent falling while walking. She was born at term, immediately cried at birth and had no significant perinatal history. She had attained normal milestones with age and shows good cognitive functions. On examination child had ataxic gait with dysidiadochokinesia, past pointing and intentional tremors. Deep tendon reflexes were graded as 3+ and plantar jerk was upgoing bilaterally. Nystagmus, Romberg sign and pendular knee jerk were absent on examination. No papilledema noted on ocular examination. Routine blood investigations were within normal limits. Based on complaints and examination findings, clinician suspected a cerebellar pathology. Further Magnetic Resonance Imaging (MRI) brain was advised to localise the cause of complaints and identify the pathology.

Plain MRI study of brain without contrast was done for the patient. The scan revealed a large well defined thin walled T2 hyperintense lesion (approx. 5 x 4.3 x 5.2 cm) with signal intensity following CSF attenuation on all the sequences, suprasellar in location with epicentre and involvement of 3rd ventricle (Figure 1). No septations, solid or haemorrhagic component was noted associated with the cyst. The lesion was

superiorly extending to cause near complete obliteration of bilateral foramen of Monro with suspicious small patency seen on left side as against right side which appeared completely obliterated. It was also causing mass effect by displacing optic chiasma and infundibulum anteriorly and midbrain posteriorly {Figure 2(C)} with insinuation into interpeduncular cistern causing splaying of bilateral cerebral peduncles {Figure 1(B)}.

Upstream dilatation of bilateral lateral ventricles was noted with transpendymal seepage of CSF. Third ventricle was distended by the cyst. Fourth ventricle appeared normal in size however aqueduct of Sylvius at its proximal opening was seen to be obliterated partly by the cyst wall forming the floor of the cyst. Rest of the aqueduct appeared uniform and normal in caliber. Based on MRI findings, diagnosis of arachnoid cyst suprasellar or third ventricular in location was made.

After initial identification of arachnoid cyst leading to upstream hydrocephalus on imaging, patient underwent endoscopic surgery that confirmed the imaging findings followed by endoscopic fenestration of the cyst to establish communication between the cyst and ventricular system in order to reduce the size of the cyst and release obstruction.

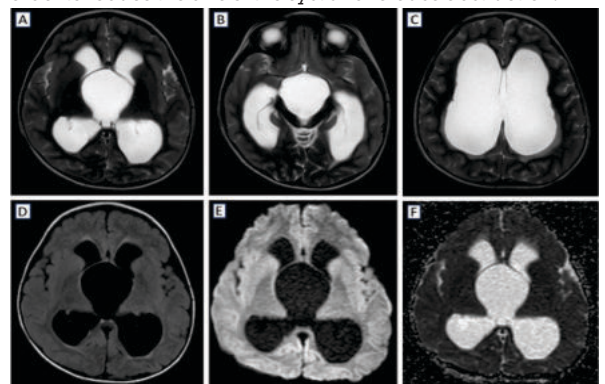


Figure 1. T2WI of axial sections of brain (A and B) shows T2 hyperintense lesion distending third ventricle with signal intensity similar to CSF and associated dilatation of bilateral lateral ventricles (A) and splaying of cerebral peduncles (B). T2WI axial sections at the level of lateral ventricle superior to

the lesion shows dilated lateral ventricles with periventricular hyperintensity s/o transpendymal seepage/ooze. FLAIR (D) shows complete signal suppression within the lesion similar to CSF. DWI (E) and corresponding ADC (F) shows no diffusion restriction with bright signal on ADC similar to CSF.

Patient is stable post operatively and shows improvement in the symptoms. 6 weeks post-surgery, patient had substantially reduced intentional tremors. Follow up CT showed significant reduction in the size of the cyst and the hydrocephalus with near complete resolution of periventricular changes suggesting reversible transpendymal seepage of CSF. This confirmed good surgical outcome of the fenestration procedure.

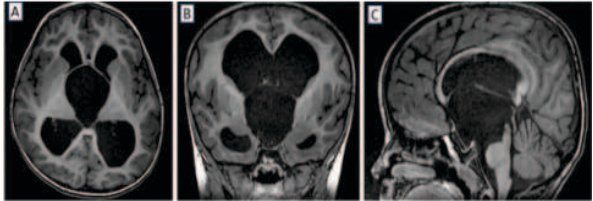


Figure 2. T1WI of brain shows dilated third and bilateral lateral ventricles with obliteration of bilateral foramen of Monro (A, B) and aqueduct of Sylvius (C). Optic chiasma in midline along with pituitary stalk are displaced anteriorly with flattened pituitary and intact posterior bright spot (C).

DISCUSSION

Arachnoid cysts can be either primary or secondary i.e. they can be congenital or acquired. More common are the congenital type however arachnoid cysts have also been known to develop post-surgery or post trauma. [1] Many arachnoid cysts remain asymptomatic but third ventricular cysts may become symptomatic either due to compression of adjacent structures or due to obstruction of foramen of monro or aqueduct of sylvius usually resulting in obstructive hydrocephalus.[3] Clinical manifestations due to compression may result in visual abnormalities and/or hypothalamic/pituitary dysfunction and drop attacks/head bobbing due to obstructive hydrocephalus or intermittent foraminal obstruction secondary to pendulous nature of cyst. [4]

Third ventricular cyst on imaging may be misdiagnosed as ventriculomegaly/dilatation especially if cyst wall is indiscernible. Aqueductal stenosis (by web/diaphragm) is a close imaging differential of such a midline arachnoid cyst in these scenarios. However, in case of arachnoid cyst 3rd

ventricle is more rounded with significantly large mass effect seen as anterior displacement of optic chiasma, compression of pituitary, elongation of pituitary stalk, splaying of the cerebral peduncles, flattening of pontine belly as was seen in this case. In case of aqueductal stenosis, 3rd ventricle is more rectangular/ ovoid anteroposteriorly with relatively less displacement of chiasma/pituitary. Also the splaying of cerebral peduncle will be less conspicuous as against 3rd ventricular arachnoid cyst. Cerebellum appeared normal.

For confirmation, MRI brain can be done using High-resolution thin-sections of T2WI, FIESTA, or CISS sequences which helps in delineating the CSF spaces and may also demonstrate subtle abnormalities not detected on standard sequences. CSF flow dynamics in the aqueduct and around the foramen magnum can also be depicted using 2D cine-phase contrast imaging. [2]

COMPLICATIONS

A large 3rd ventricular cyst as in our case may result in upstream intraventricular obstructive hydrocephalus (IVOH). In severe cases of IVOH, there may be compression of corpus callosum against the free inferior margin of falx with resultant pressure necrosis and loss of callosal axons. This condition can be labelled as corpus callosal impingement syndrome (CCIS). In acute CCIS, there is T2/FLAIR hyperintensity of corpus callosum. In subacute and chronic long-standing cases encephalomalacic changes with thinned atrophic corpus callosum is seen. These changes may persist post decompression especially in long standing pathologies. Massive enlargement of ventricle may also result in CSF diverticulum extruding through the inferomedial medial wall of atrium of the lateral ventricle secondary to weakening of the lateral ventricular medial wall. These diverticula may cause mass effect on the posterior third ventricle, tectal plate and aqueduct. Large diverticulum can also herniate posteriorly compressing vermis and fourth ventricle. Rarely, there may be ependymal rupture resulting in ventricular disruption and subsequent spill of CSF into the adjacent periventricular white matter creating a fluid filled cleft like appearance. [2]

In order to improve patients symptomatically and avoid aforementioned complications of long-standing disease, early surgical intervention is indicated. Various surgical procedures have been used in the past. Table 1 shows the cases of third ventricular cyst reported in literature till date with symptoms, radiological findings and operative techniques.

Table (1): Third Ventricular Arachnoid Cysts Reported In Literature.

Various studies	Age (in years)/ Gender	Symptoms	Radiological findings	Operative technique
Faris et al. (1971) [5]	16/M	Headache, precocious puberty	Triventricular hydro-cephalus	Frontal craniotomy and fenestration
Ericson et al. (1986)[6]	5/M	Headache, nausea, somnolence	Enlarged 3rd ventricle	Stereotactic puncture
Tamburus et al. (1987) [7]	20/M	Nausea, vomiting, horizontal nystagmus	Triventricular hydro-cephalus	Omayya reservoir, parietal craniotomy, cyst excision
Tamburus et al. (1987) [7]	46/M	Intracranial hypertension, optic atrophy	Aqueductal stenosis	Ventricular drainage, frontoparietal craniotomy, fenestration
Shiba et al. (2010) [1]	35/M	Epilepsy, mental retardation	Triventricular hydro-cephalus	Endoscopic fenestration
Jeltema et al. (2014)[8]	2.5/-	Altered sensorium	Triventricular hydro-cephalus	Endoscopic fenestration, partial cyst removal, ventricular cisternostomy, endoscopic fenestration
Ho et al. (2015)[9]	33/F	Headache, blurred vision, gala-ctorrhoea	Hydro-cephalus with mildly enlarged third ventricle	Frontal craniotomy with gross total cyst excision
Akpınar et al. (2017) [3]	57/M	Loss of cons-ciousness	Triventricular hydro-cephalus	VP shunt
Ong et al. (2019) [4]	19/M	Antero-grade amnesia	Third ventricular dilatation with obstructive hydro-cephalus	Endoscopic fenestration

Current case (2024)	5/F	Intentional tremors	Obstructive hydro-cephalus by third ventricular cyst	Endoscopic fenestration
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CONCLUSION

In conclusion, this is a rare case of intraventricular arachnoid cyst in a child with a rare clinical presentation of intentional tremors pointing clinically towards a cerebellar pathology. A comprehensive history followed by cross sectional imaging of brain is indicated for appropriate assessment with use of adequately thin sliced sections for identification of the cyst wall in case of suspicion.

Pre operative identification of arachnoid cyst and its differentiation from other possible imaging differentials of obstructive hydrocephalus is important for early intervention and adequate treatment planning for the patient. Early surgical intervention may help evading the chronic irreversible damage to the brain parenchyma with excellent outcome.

Declarations

Ethical Approval: It was carried out under declaration of Helsinki and patient's informed consent was obtained for participation and publishing the report.

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