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ABSTRACT

The veli interpositi represents an embryologic membrane which form following fusion or superposition of two layers of pia mater (tela choroidea) of the third ventricle during embryologic development of the corpus callosum.[1-3] This thus becomes a potential space which can harbour CSF. Kruse in 1930 first described dilatation of this potential space and defined such condition as "cavum veli interpositi".[1] A cavum veli interpositi (CVI) has also been called "cisterna interventricularis", "ventriculi tertii", "transverse fissure", and "subtrigonal fissure".[1] This dilatation is a normal variant in the newborn which spontaneously closes by the end of the first year of life.[3, 4] Only 2-3% persist beyond this period into adulthood.[4] Pathologies involving this region varies, and might range from just a benign cystic dilatation of this space, to pathologies involving true cysts such as arachnoid and epidermoid cysts, and tumours such as meningiomas.

Keywords
cavum veli interpositi, endoscopic fenestration, cavum septum pellucidum, cavum vergae interpositi, neuropsychiatric

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INTRODUCTION

Cavum veli interpositi must be differentiated from other potentially confusing anterior midline intracranial cystic dilatations within this region. Cavum septum pellucidum (CSP) is dilatation of the potential cavity between the membranous leaves of the anterior septum pellucidum, separated by at least 1 mm.[5] This is similarly a normal variant, seen in neonates and expected to close by early childhood. A cavum vergae (CV), another normal anatomical variant, is the potential cavity in the posterior aspect of the septum pellucidum, and only separated from the more anterior cavum septum pellucidum by the anterior column of the fornix. From six months of intrauterine life, fusion of these potential spaces or cavities into a single septum pellucidum begin to occur in most individuals from posterior to anterior. Thus, a cavum vergae obliterates first before its cavum septum pellucidum counterpart. As such, a cavum vergae is rarely an isolated finding and invariably is associated with a cavum septum pellucidum, in which case it is called a cavum septum pellucidum et vergae.[6, 7]

These normal variants differ from true cerebral ventricles in that they lack linings with ependymal cells or choroid plexus.

Radiologically, CVI lie inferior to the fornix, above the third ventricle and posterior to the foramen of Monro. They assume a triangular shape with the apex pointed anteriorly, at times as far as the foramen of Monro. They also displace the internal cerebral veins inferiorly and enclose them laterally. Conversely, CSP lie between the frontal horns of the lateral ventricles and anterior to the foramen of Monro but above the fornix and assume a triangular shape with apex pointed posteriorly. CV lie above

the internal cerebral veins (and are usually distinct from them rather than enclose them as seen with CVI) and transverse part of the fornix, and assume a rectangular shape. They are mostly seen in communication with the more anteriorly lying CSP.[4]

A cavum (cavum= cave-like) can undergo cystic dilatation. A cystic dilatation of the CVI is said to occur when there is dilatation ≥ 10 mm in its widest width, in the presence of lateral bowing of the walls.[8] Thus, a cavum may expand to form a cyst. Furthermore, differentiating a "cystic dilatation of the CVI" from a "cyst of the veli interpositi" is imperative. Most cystic dilatation of the cavum veli are not associated with mass effects such as hydrocephalus because they mostly communicate with adjacent ventricles. A cyst of the veli interpositi can include pathologies such as arachnoid or dermoid cysts while CVI cystic dilatations are simply CSF-containing dilatations which follow failure of fusion of adjacent leaves of the tela choroidea during development.[3, 9, 10]

A cyst of the veli interpositi may be communicating (with adjacent ventricles) or non-communicating. It can also be classed as symptomatic or asymptomatic. Each of these classes are important in treatment decisions.

CVI are usually asymptomatic and are thus mostly incidental findings on cranial MRI or CT scans done for other indications. However, when present, symptoms and complications can include recurrent headaches, hydrocephalus and macrocephaly, developmental delays, mental retardation, and neuropsychiatric behaviours including psychogenic seizures.[2, 11-14] Coexistence with multiple cranial neuropathies, gait disturbances, and progressive memory impairment have also been reported in literature.[9, 13, 15]

The treatment of symptomatic cysts of the veli interpositi (CVI) is a subject of ongoing debate, with available options including conservative management, endoscopic fenestration, and open surgical intervention. Given the debates surrounding the optimal treatment strategy, particularly for symptomatic CVI cysts without hydrocephalus, there is a critical need for studies that explore the rationale behind the choice of surgical versus conservative management in terms of symptoms regression and cyst resolution. This case report provides valuable insights into this decision-making process, offering evidence to support clinical judgments on whether to operate or not.

Case presentation

We report a nine-year-old boy who presented with a one-week history of seizures, recurrent headaches, and neuropsychiatric symptoms. His mother reported a history of seizures which were characterized by several episodes per day of myoclonic-like jerks involving the head and shoulders. The episodes were reported to be spontaneously triggered, lasting a few minutes before spontaneously ceasing. Attacks were not associated with loss of awareness and occasionally occurred while sleeping. He had a history of intermittent dull, pancranial, moderately severe headaches of similar duration, but had no history of febrile illness or other features suggestive of raised intracranial pressure. The pregnancy and birth histories were unremarkable. He also attained adequate developmental milestones for his age and had been apparently well until the incident. He was the last of six children in a monogamous family setting, and there was no history of similar problem with any of his siblings or in his family. He was a primary four pupil, and his academic performance was reported as satisfactory.

A brain magnetic resonance imaging (MRI) revealed a cyst within the region of the velum interpositum, lying posterior to the foramen of Monro and inferior to the fornix, with upward displacement of the posterior aspect of the corpus callosum. The cyst measured 4.5 cm in its widest dimension on axial slice with bowing of its lateral walls (figure 1). The intensity of the cyst fluid was similar to cerebrospinal fluid (CSF) on all MRI sequences. EEG showed no abnormal electrical activity within the brain. He was placed on antiseizure medications and analgesics with no relief of symptoms, including the seizure-like events.

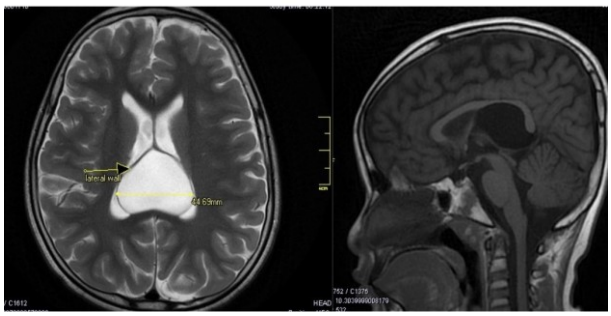

Figure 1. Index patient with a cyst of the veli interpositi. The cyst measured 4.5 cm in its widest diameter. Note the bowing of the lateral walls.

Following due consent, he was worked up for endoscopic fenestration of the cyst. A decision was made to utilize a right parietal transcortical approach because this non-dominant area was most accessible to the cyst with potentially insignificant breach of cortical tissue. Possible trajectories or corridors of entry for a cystoventriculostomy in the index patient are shown in Figure 2.

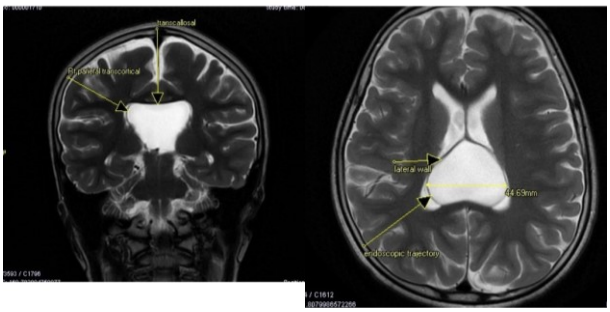

Figure 2. Possible corridors for endoscopic fenestration in the patient. A right parietal transcortical approach was utilized in this case.

A single burrhole was made about 2 cm inferior to the right Keen's point (figure 3), and the underlying dura was electrocoagulated and incised in cruciate fashion. Its overhanging leaflets were further cauterized to widen access for the endoscope. Under endoscopic guidance, the cyst wall was fenestrated into the ipsilateral lateral ventricle at multiple points, and these fenestrations were widened with the aid of biopsy forceps.

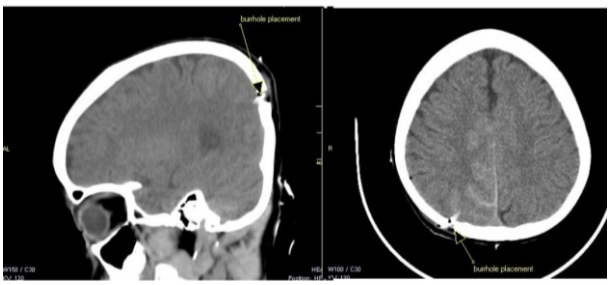

Figure 3. Burrhole placement.

Inspection of both cyst and ventricles were done, and biopsy of the cyst wall was obtained for histology. An external ventricular drain was further inserted into the cyst. Post-operatively, he was nursed in the ward and post-operative recovery was uneventful.

Post-operative CT scan done showed satisfactory communication between the cyst and the lateral ventricle, and some resolution of the cyst size and its lateral bowing.

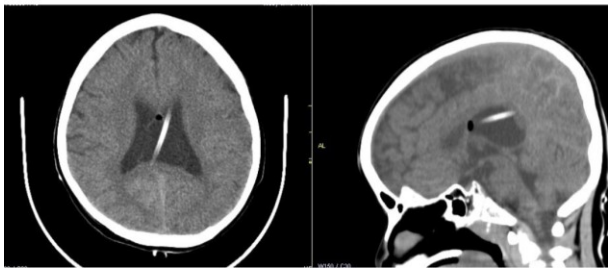

Figure 4. Immediate post-operative CT scan with EVD.

The external drain was removed on the 6th day and the patient was discharged on the 7th day post-surgery and followed-up in the clinic on an outpatient basis. Three-month follow-up post-surgery consistently demonstrated no return of symptoms.

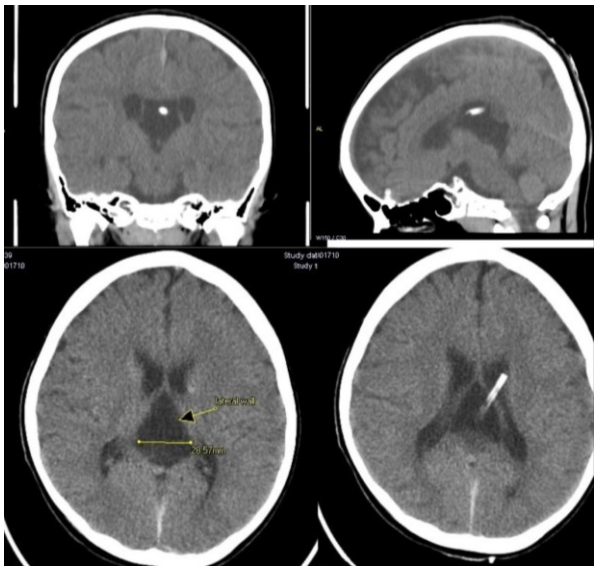

Figure 5. 5th day post operation. Significant resolution of cyst can be seen with loss of bowing of the lateral walls.

Discussion

Cysts of the velum interpositum are rare findings in our environment. Treatment modality for asymptomatic cysts is typically straight forward – utilizing conservative approaches such as observation and follow-up with serial clinical evaluation to detect emergence of new symptoms, as well as serial intracranial scans to evaluate for increase in size of the cyst. These developments might consequently warrant abandonment of conservative approach in favour of surgery.

Furthermore, patients who present with symptomatic velum interpositum cyst in the presence of hydrocephalus or features of raised intracranial pressure are of necessity treated surgically. However, in patients who present with symptomatic velum interpositum cyst with no features of hydrocephalus or raised intracranial pressure, the debate arises on what treatment modality should be employed between surgical and conservative approaches. Moreover, if surgical approach is adopted, further controversy arises on which surgical modality offers best outcome in terms of symptoms resolution and cyst regression. This case report examines the outcome of endoscopic fenestration of a symptomatic velum interpositum cyst in a nine-year-old boy with vivid neuropsychiatric symptoms and no evidence of hydrocephalus and EEG-based seizure disorder. The outcome presented here is also compared with those observed in literature.

Surgical options of treatment of such symptomatic cysts include open craniotomy, endoscopic fenestration into a ventricle (or a cystoventriculostomy), stereotactic procedures and shunting procedures including cyst-peritoneal and cyst-atrial shunts. A meta-analysis of 368 cases of cavum cysts from 72 manuscripts was carried out to evaluate the epidemiology, pathogenesis, clinical symptoms, and results of surgical treatment of these cysts. This study concluded that operative treatment yields better regression of symptoms and low complications than conservative approach.[15]

Endoscopic fenestration of a CVI has been demonstrated in literature to be effective in obviating the need for open craniotomy or formal shunting procedures and yielding better outcomes than open surgery or shunting.[14-18] A systematic review study of 37 articles evaluating the surgical management of 54 patients with symptomatic cysts was done. Thirty-five patients (71%) had endoscopic fenestration while 3 had open craniotomies (8%) and 10 had shunting procedures (15%) done. Endoscopic fenestrations included 33 right frontal approaches and 3 right parietal approaches. Craniotomies were two transcortical and one transcallosal approaches. Outcome measures were both clinical (improvement in symptoms) and radiologic (resolution of cyst size). Endoscopic fenestration resulted in symptom improvement in 37 out of 39 patients, and radiologic improvement in cyst size in all 39 patients (100%). Two of the 39 patients remained the same symptomatically. There were no cases of recurrence. Conversely, the craniotomy arm resulted in both clinical and radiologic improvement in only 2 out of 3 patients. Recurrence and consequent re-operation occurred in one patient.[16] Endoscopic approaches described for a cystoventriculostomy include a frontal transcortical, parietal/occipital transcortical and transcavum interforniceal approaches. There was no statistical difference in outcome with respect to recurrence rates or complications between any of the approaches used.[16]

Conversely, employment of conservative approaches for such symptomatic cyst with no hydrocephalus have been described. This approach entails serial clinical and radiologic follow-up to monitor for worsening of symptoms as well as increase in the size of the cysts, as well as symptomatic treatment of the condition. With the conservative approach, there is no attendant risk of surgery. However, its practicability within our environment is doubtful. There is increased probability of defaulting on outpatient follow-up, especially if the patient is stable symptomatically with no signs of raised intracranial pressure. Moreover, the financial feasibility of doing serial MRI scans in our environment might be a further propellant for defaulting. Additionally, studies have demonstrated that utilizing conservative approach, symptom resolution takes longer time when compared to surgical alternative.[19-22] In many cases, treatment failure results.[14, 15, 19] There is thus the attendant risk of sudden deterioration during conservative management and eventual need for surgery.

Conclusion

Cysts of the veli interpositi are rare midline intracranial cysts. Most are asymptomatic but they can present with neuropsychiatric manifestations including psychogenic seizures. Treatment for such symptomatic cysts in the absence of hydrocephalus has been debatable. Endoscopic cystoventriculostomy provides an effective treatment option with complete resolution of symptoms.

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