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Case Report

Cavum septum pellucidum and vergae cyst: A symptomatic case with intracranial hypertension and multiple nerve involvement

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Received : 07 September 2022
Accepted : 21 November 2022
Published : 02 December 2022

DOI
10.25259/SNI_822_2022

Video available on:
https://doi.org/10.25259/SNI_822_2022

Quick Response Code:

ABSTRACT

Background: Cavum septum pellucidum (CSP) and cavum vergae (CV) are normal anatomical variations present in some children, adolescents, and adults. When the cavity is larger than normal, it is called a cyst. Symptomatic cases of CSP and CV cyst are rare, and the clinical presentation is varied. A case with multiple nerve involvement is described.

Case Description: A healthy 17-year-old female presented a sudden headache, right cervicobrachialgia, right ptosis, visual changes, and left facial paralysis over 10 days. Head magnetic resonance imaging revealed CSP and CV cyst. Inflammation, infection, and vascular disorders were ruled out. We decided to perform a right transfrontal endoscopic intraventricular septostomy and a right Monro foraminoplasty, which were successful. One month after surgery, the patient had no more signs or symptoms. She has remained asymptomatic for the past year.

Conclusion: Multiple nerve involvement was directly related to CSP and CV cyst. The cyst bilaterally occluded the foramen of Monro causing intracranial hypertension. It was possible to obtain complete resolution of the clinical features through neuroendoscopic fenestration and foraminoplasty.

Keywords: Cavum septum pellucidum, Cavum vergae, Cranial nerves palsy, Intracranial hypertension, Septum pellucidum

INTRODUCTION

The septum pellucidum (SP) is a thin, translucent, and triangular double membrane which separates the right and left frontal horns from the lateral ventricles of the brain. It extends between the anterior portion of the corpus callosum, and the body of the fornix and its width varies from 1.5 to 3.0 mm.[3]

The cavum septum pellucidum (CSP) and cavum vergae (CV) are persistent middle line structures of the adult brain that is formed when the proper closure of the membranous leaves

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of the SP does not occur completely. CV is the term used to designate the back extension of the CSP.[9]

It is considered a normal anatomical variation present in all premature babies and in 85% of full-term newborns, which usually disappears by 3–6 months of postnatal life. Nonetheless, 12% of children between 6 months and 16 years of age may continue to have it.[3,6,8] According to Chen *et al*., 19,031 patients (from 0 to 99 years old and averaging 52.6 years old) were studied and 177 (0.93%) had CSP and/or CV. CSP with CV was found in 169 (95.5%) of these.[2]

When the cavity is larger than normal, it can be called a cyst. It is defined as a structure containing cerebrospinal fluid (CSF) between the lateral ventricles, whose walls are laterally arched rather than parallel and are 10 mm or more apart.[12,14] Cyst size is directly associated with neuropsychic dysfunction. In this sense, the larger the cyst, the worse the patient's clinical condition, suggesting more attention in these cases.[7]

In general, it is an incidental finding of imaging examinations (0.04%). However, when symptoms such as headache, behavioral disorders, cognitive impairment, convulsive syndrome, dizziness, hydrocephalus, nausea, and vomiting occur due to its presence, it is even rarer.[3,6,12]

The aim of this case report is to show exquisite clinical features of a symptomatic CSP and CV cyst in a young patient, successfully treated with neuroendoscopic fenestration and foraminoplasty.

CASE DESCRIPTION

A healthy 17-year-old female developed a holocranial headache, photophobia, nausea, vomiting, hypertension, and pain radiating from the right trapezius muscle to the ipsilateral wrist for 10 days. Three days later, she was hospitalized due to right eyelid ptosis, unilateral blurred vision, and worsening headache intensity. We found no abnormalities in her physiological, familial, and social history. The use of any drugs or medications, comorbidities, surgeries, traumas, and allergies was ruled out.

Physical examination revealed palsy of the right third nerve with the right eyelid ptosis, paralysis of the right medial rectus muscle, and ipsilateral mydriasis. She also had a right sixth nerve palsy with the right lateral rectus paresis and a left seventh nerve with the left peripheral facial paralysis. Fundoscopy showed bilateral papilledema [Figure 1]. Other cranial nerves were unchanged, as well as her balance, coordination, strength, muscle tone, and amplitude, reflexes, and sensory examination. Cardiovascular, pulmonary, and osteoarticular systems were normal.

Lumbar puncture performed after computed tomography (CT scan) that ruled out an absolute contraindication for this procedure, showed high opening pressure, and measured in more than 30 cm of water, but no findings of CSF infection were found. Other laboratory tests were negative for autoimmune and infectious diseases.

Cranial magnetic resonance imaging (MRI) demonstrated a CSP and CV cyst measuring 2.5 cm × 2.8 cm × 6.3 cm (longitudinal × transverse × anteroposterior) and an estimated volume of 22.93 cm^3, which was causing bilateral obstruction of the foramen of Monro and consequent hydrocephalus [Figures 2a-c]. There was no evidence of increased contrast uptake in cranial nerves and compression of cervical structures. Inflammation and vascular abnormalities were not seen on additional magnetic resonance angiography.

On the 10th day after admission, a right transfrontal endoscopic intraventricular septostomy was successfully performed, communicating both lateral ventricles with the cyst, as well as a right endoscopic dilatation of the foramen of Monro (foraminoplasty) (EMF) to ensure communication between the lateral and third ventricles [Video 1].

During the following 3 days, the patient showed improvement in headaches, eyelid ptosis, and facial paralysis. On postsurgical CT scan, the cyst was smaller and there were no bleeding or other significant abnormalities [Figures 2d-f]. She was discharged on the 3rd postoperative day with analgesics and scheduled for outpatient follow-up.

At the 1-month follow-up appointment, she no longer had headaches, cranial nerves abnormalities, or any other neurological disorders. After 1 year, she remained asymptomatic and with normal fundoscopy.

DISCUSSION

The clinical evolution time for each patient with CSP and/or CV cyst is not specified in many articles; however, most of the cases progress from 1 month to 3 years.[12] The present study demonstrates a 13-day course, with no previous similar signs or symptoms, which characterizes a very acute presentation.

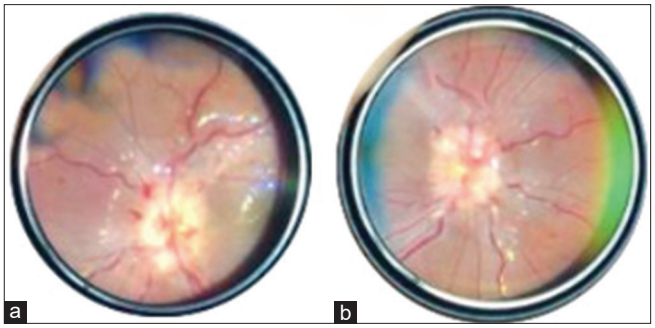


Figure 1: Bilateral papilledema on fundoscopy. (a) To the right eye and (b) to the left eye.

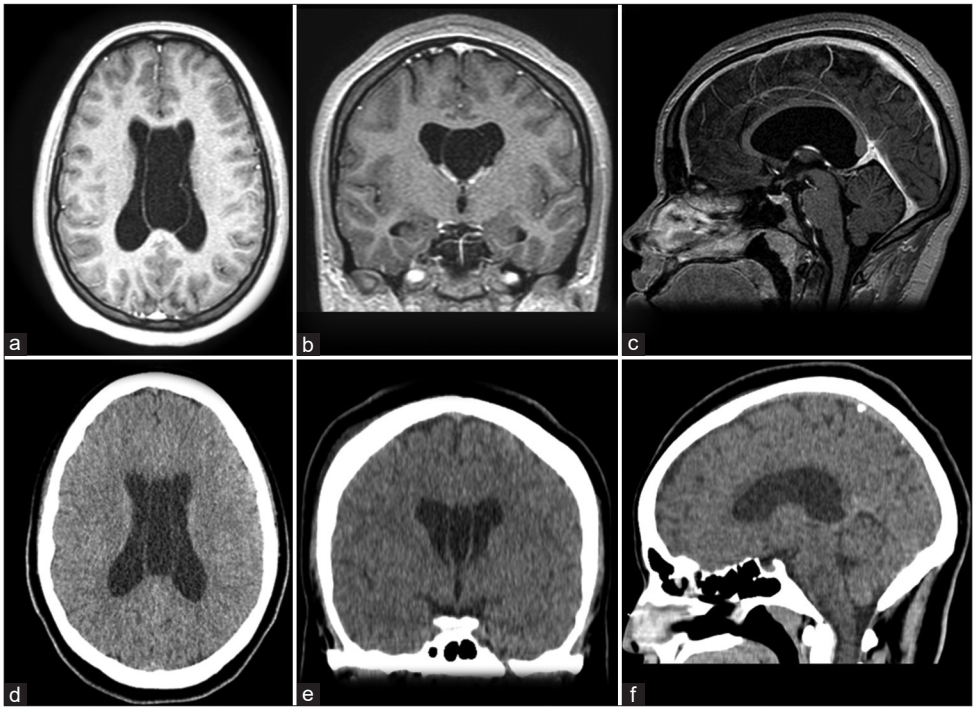
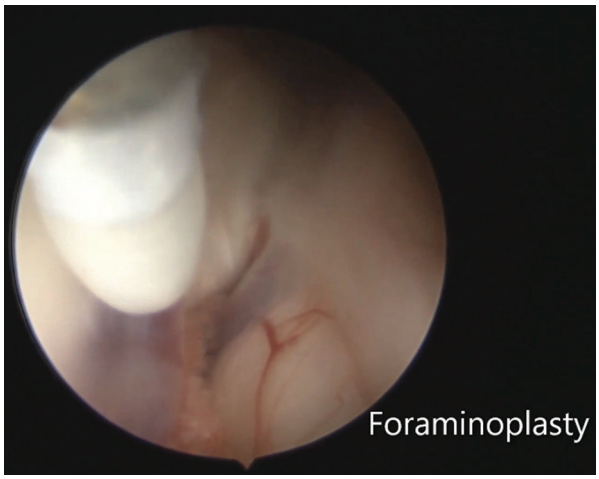


Figure 2: Pre- and post-surgical cavum septum pellucidum (CSP) and cavum vergae (CV). (a and b) show the CSP and CV on presurgical magnetic resonance imaging. (b) shows the septate cyst. (c) it is not possible to see the cyst on presurgical sagittal plane. (d and e), from the postsurgical CT, demonstrate the cyst in smaller volume. (f) it is not possible to see the cyst on postsurgical sagittal plane.



Video 1: Neuroendoscopic fenestration and foraminoplasty.

The patient and her family also had no prior knowledge about the cyst.

CSP and CV cysts are usually considered an incidental finding, although some of them may be pathological. It can be justified by some mechanisms, such as obstruction of the interventricular foramen, resulting in hydrocephalus and/or increased intracranial pressure; compression of the hypothalamic-septal triangle, leading to neuropsychiatric symptoms and/or compression of the optic chiasm and its pathways; and chronic deep venous involvement which can cause progressive focal deficits.[3] Despite being a congenital cyst, we did not find any factor that could explain its clinical presentation in late adolescence.

In a meta-analysis performed with 368 patients, the following signs and symptoms were analyzed: headache (n = 184; 50%) and convulsive syndrome (n = 87; 23.6%); reduced intelligence/delayed psychomotor development (n = 74; 20.1%); mental disorders (n = 58;15.8%); dizziness, nausea, and vomiting (n = 40; 10.9%); impaired consciousness (n = 36; 9.8%); gait disorders (n = 33; 9%); visual impairment (n = 31; 8.4%); optic nerve swelling (n = 17; 4.6%); cranial nerve dysfunction (n = 15; 4%); and hydrocephalus (n = 61; 16.6%).[6] The main clinical findings (headache, signs and symptoms related to hydrocephalus, nausea, and vomiting) were observed in the present study, which may demonstrate a typical course of the disease. In contrast, the involvement of cranial nerves (III, VI, and VII) of our patient is evidenced as a rare manifestation, according to Kryukov *et al.* (2020). Despite the complaint of cervicobrachial pain, findings in the MRI that could justify this, such as downward displacement of the cerebellum or compression of the cervical root, were not observed.

According to Reid *et al*. and Sahin *et al*., in addition to papilledema, the sixth nerve palsy is the most common neurological disorder associated with intracranial hypertension (IH). This occurs when the brainstem moves downward, stretching the VI pair as it crosses over the petrous ridge and enters Dorello's canal.[10,11] The involvement of the third nerve could be explained by its compression anywhere along its course from the fascicle to the orbit.[1] On the other hand, paralysis of the VII nerve could be justified by the traction on the nerve caused by IH before entering the temporal bone.[5]

The surgical approach is indicated when there is obstruction of the CSF flow in the foramen of Monro, direct compression of surrounding tissues, changes in mental status, or focal neurological deficits.[3] In a systematic review with 54 patients with symptomatic CSP, the most common procedures performed were endoscopic fenestration (n = 39; 72.2%), shutting (n = 10; 18.5%), open surgery (n = 3; 5.5%), and stereotactic fenestration (n = 1; 1.8%). It was concluded that open surgery and shutting have poor clinical outcomes when compared to endoscopic or stereotactic procedures (frontal, parietal, and transcavum fenestration). In addition, half of the patients treated by shutting still had recurrence of the condition.[12,13]

Endoscopic fenestration involves a burr hole craniotomy, commonly performed in the right frontal region, to communicate the cyst with the lateral ventricles. It can also be executed by parietal cystostomy or direct transcavum interforniceal endoscopic fenestration. There was no statistically significant difference between these techniques, but they provide a reduction of the cyst size, with a less invasive approach, less recurrence, and better clinical outcome.[3,12]

Regarding EMF, Elkheshin and Zohdi reported three different types: dilatation (for congenital, idiopathic, or posthemorrhagic/meningitis stenosis), restoration (treating cyst wall obstruction or a membrane partially covering a nonstenotic foramen), and excision (for lesions of solid or mixed cystic components). In their study, 55 patients, who were submitted to EMF, had collapsible cyst walls.[4] In our case, the foraminoplasty was performed to restore its size and certify good communication between the right lateral ventricle and third ventricle.

Our patient showed partial improvement of signs and symptoms days after endoscopic surgery and complete improvement 1 month after medical discharge, remaining asymptomatic since then.

CONCLUSION

This study provided a rare case of symptomatic enlargement of CSP and CV with IH and multiple nerves involvement. Cranial MRI was essential for the diagnosis. Neuroendoscopic fenestration and foraminoplasty provided complete resolution of the clinical features.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Ferreira DB, Medeiros JW, Cwajg E, Ferreira-Pinto PH, de Carvalho LU, Nigri F. Cavum septum pellucidum and vergae cyst: A symptomatic case with intracranial hypertension and multiple nerve involvement. Surg Neurol Int 2022;13:564.

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