Case Report

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Cerebral Venous Sinus Thrombosis (CVST) as a Rare Presentation of a Brain Hydatid Cyst: A Case Report

Maryam Kaeedi [1], Hosein Kaeidi [2], Mojtaba Shahbazi [1], Sanaz Heydari Havadaragh [1], Hamed Amirifard [1]*

1.Iranian Center of Neurological Research, Neuroscience Institute, Tehran University of Medical Sciences, Tehran, Iran.
2 School of Medicine, Shahid Beheshti University of Medical Science, Tehran, Iran.

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Running Title Cerebral Venous Sinus Thrombosis In A Cerebral Hydatid Cyst

ABSTRACT

Cystic hydatidosis is a rare zoonotic condition caused by the larva of the Echinococcus tapeworm. In this disease, brain involvement is a rare condition, accounting for about 2-3% of all cases with hydatid disease. Focal deficits, symptoms of increased intracranial pressure, seizure and mental state changes are most common presentations of CNS cystic echinococcosis. We present a young woman with resistant headache and transient visual obscurations due to increased intra-cranial pressure caused by Cerebral venous sinus thrombosis (CVST) arising from a Hydatid cyst. To the best of our knowledge, this is the first report of hydatid cyst induced CVST.

Our case report highlights the importance of considering CVST as an unavoidable complication of brain's space occupying lesions and especially infective ones as a differential diagnosis.

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*** Corresponding Author:**
Hamed Amirifard, MD.
Address: *Iranian Center of Neurological Research, Neuroscience Institute, Tehran University of Medical Sciences, Tehran, Iran.*
E-mail: *dr.amirifard@gmail.com*

Introduction

Cystic hydatidosis is a rare zoonotic condition caused by the larva of the Echinococcus tapeworm. Due to limited studies, prevalence of echinococcal infection is possibly underestimated worldwide, with most patients reported from South America, the Middle East (including Iran), Africa and Southeast Asia [1]. Liver is the most common affected organ (60-70%) followed by the lungs (20-30%) and other organs involved account less than 10% of cases [1]. Central nervous system (CNS) involvement is extremely unusual (2-3%), leading to significant mortality and morbidity [2, 3]. Focal deficits, symptoms of increased intracranial pressure(ICP) such as headache and visual disturbance, epilepsy, mental state changes and skull deformities are most common presentations of CNS cystic echinococcosis [2, 4].

Cerebral venous sinus thrombosis (CVST) is an infrequent condition, resulting in clotting of blood in the cerebral venous or dural venous sinuses, and in some cases, the cortical veins. Main predisposing conditions associated with CVST include coagulation disorders, pregnancy, puerperium, oral contraceptive pills (OCPs), malignancies, intracranial tumors, direct septic trauma, intracranial infections, regional infections, and general infections. Main clinical sign of CVST are headache, focal deficit, seizure, and altered mental status [4, 5]. There isn't any previous report of CVST caused by a hydatid cyst.

In this study, we reported a woman in her mid-thirties with a brain hydatid cyst who sought help by coming to the hospital with headache and transient visual obscuration caused by intracranial hypertension due to CVST, without any evidence of hydrocephalus or midline displacement in imaging studies

Case Presentation

A previously healthy 35-year-old woman was referred to our hospital complaining of transient visual obscurations and persistent dull headaches in the occipital and bi-temporal regions for the past 25 days. According to the patient history, headache was worst on awakening and the severity of headache progressed over time to the extent that it awakened the patient from sleep. Furthermore, headache was vague, severe, resistant to simple painkillers (NSAIDs), and aggravated by sleeping, bending, and valsalva maneuver. As the patient's headache progressed, her visual impairment also got worse. She did not report any other symptoms such as weight loss, skin lesions, nausea or vomiting, cough, fever, dyspnea, pleuritic chest pain, hemoptysis, sensation of abdominal pain or fullness, and feeling less alert than usual. The patient did not report any recent history of fever or infection, or any specific medication use such as OCPs. No specific family history or habitual history was present.

She was a young, alert female with bilateral papilledema in neurologic cranial exam. Confrontation test was normal and other cranial examinations revealed no abnormal signs. Motor and sensory exam, reflexes (including plantar reflex), coordination and gait were also normal. Other exams were unremarkable.

Investigation and Treatment

With elevated ICP being suspected, brain CT scan was performed which showed a cystic lesion in right occipital lobe. Then, brain magnetic resonance imaging (MRI) with and without contrast were performed (Fig. 1-A). The result indicated a lentiform 33×22 mm extra-axial cystic lesion in the epidural occipital surface with multiple septations next to the junction transverse and sagittal sinuses in the right side with a pressure effect on the inferior part of sagittal sinus (Fig. 1-B). More investigation, including magnetic resonance venography (MRV) was performed to rule out other possible causes of the increased ICP. In MRV sequences, possible evidence of thrombosis was indicated by loss of signal void in the right transverse sinus near to the sagittal sinus (Fig. 2-A). However, it was unclear whether the slow flow was caused by thrombosis or was secondary to the mass effect of cyst. Therefore, digital subtraction angiography (DSA) was requested for the patient. The DSA, performed by a neuro-interventional specialist, indicated that right sigmoid and transverse sinuses and their junction to confluence of sinuses were not visible in the venous phase, suggestive of transverse sinus thrombosis (Fig. 2-B). In further evaluations, chest and abdominal CT scan showed multiple cysts in her liver and lungs.

The patient was diagnosed with a brain cyst, resulting in slow-flow thrombosis due to the mass effect. Therapeutic anticoagulant therapy with heparin (1000 unit/hour) (switched to warfarin (5mg) after 10 days) and anti-parasitic drug, albendazole (400mg bid) was initiated for her, resulting in significant improvement of headache. Thereafter, brain surgery through right sub-occipital craniotomy was performed, which is considered to be a safe method for complete cyst removal since it avoids the intraoperative rupture of the cyst and consequent spillage of its contents, anaphylactic shock, mortality, and recurrence of the disease. No specific complication occurred. This procedure can be done even for very large cysts which are located superficially. After the craniotomy of the occipital and suboccipital areas, the cyst was found in the epidural region, and completely removed from the dura after separation. Pathology specimen was reviewed by a pathologist and the pathologist reported laminated membranes, germinal layers and small cysts that are compatible with hydatid cyst wall and daughter cysts.

Follow-up

The patient's headache didn't improve completely after anticoagulant initiation and surgery while her transient visual obscurations improved completely. Follow up DSA did not disclose further findings. One month later, her headache improved completely.

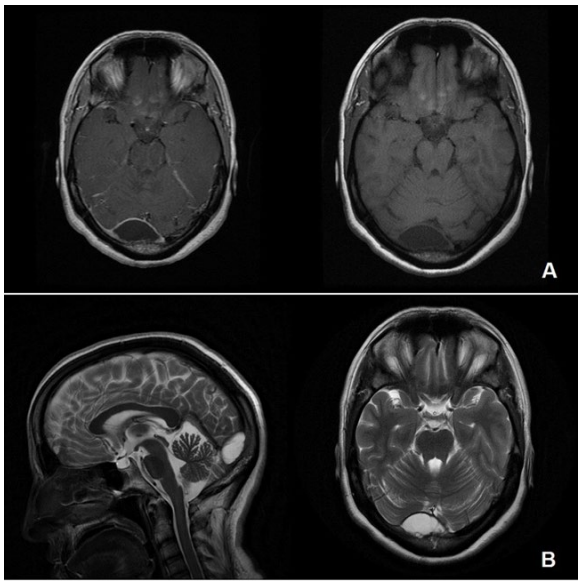

Fig. 1-A: T1 and Contrast Enhanced T1 view of MRI suggested lentiform 33×22 mm extra axial cystic lesion in epidural occipital surface. B: Sagittal and axial T2 MRI view suggested multiple septations in the occipital cyst at the left side of confluence of sinuses and pressure evidence in the inferior part of left sagittal sinus

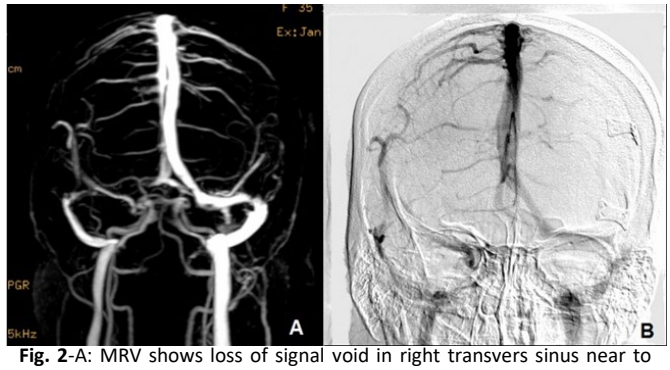

Fig. 2-A: MRV shows loss of signal void in right transvers sinus near to sagittal sinus suggestive of thrombosis B: DSA shows right sigmoid and transverse sinuses and their junction to confluence of sinuses were not visible in the venous phase

Discussion

T Cystic hydatidosis is a rare zoonotic condition caused by the larval forms of echinococcosis through fecal-oral transmission [6]. Parasite eggs, after being eaten, enter the host's intestinal mucosa and go to the liver via portal circulation. Only embryos that pass through the hepatic and pulmonary filtration systems can enter the brain through the systemic circulation. Although hydatid cysts can form everywhere in the body, the liver is the most usually affected organ [6]. Cysts in the brain occurred only in 2-3% of cases [6]. Intracranial hydatid cysts do not cause symptoms until they grow to a significant size (differ by location) [2]. Children and young male adults are the most commonly affected populations [1, 2].

Intracranial infection such as abscess or cyst is known as one of main predisposing conditions that lead to CVST. Headache and papilledema are two main clinical signs of CVST [5]. In this report, we discussed a 35-year-old adult woman who experienced headache and transient visual obscuration. Brain MRI revealed a multiseptated lentiform cyst, without midline shift or hydrocephalus. This patient is unique in several aspects. First and foremost, to the best of our knowledge, this is the first report of hydatid cyst induced CVST. Second, echinococcal invasion to the brain is a rare condition, accounting for less than 2-3% of all cases with hydatid disease [6]. Finally, the occipital lobe is the least common location of intracranial hydatid cysts [6]. In our opinion, the cause of CVST in the reported case is not entirely known and can be related to slow flow due to mass effect of an intracerebral infection and a hypercoagulative state caused by that infection.

Conclusion

CVST was most likely caused by the hydatid cyst's proximity to the venous sinuses. Unlike thrombosis, the proximity of the brain hydatid cyst to the cerebral venous system is common. Based on the findings of this case and previous research, CVST has emerged as an unavoidable complication of brain's space occupying lesions and especially infective ones which should always be considered as a differential diagnosis and as the primary cause of symptoms.

Ethical Considerations

Compliance with ethical guidelines

All authors were involved in the clinical care of the patient and contributed to conception, design, interpretation, and drafting the manuscript and approved the contents of the manuscript.

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Informed consent was obtained from before enrollment

Conflict of interest

None of the authors have any conflict of interest to disclose.

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