Unusual presentation of an intraventricular hydatid cyst as a bleeding cystic tumor: A case report and brief review

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ABSTRACT

Hydatid cysts constitute only 2% of all intracranial masses, commonly involve parenchyma and very rarely ventricles. Here, we report an unusual case of a primary, isolated intraventricular hydatid cyst that mimicked a tumor and presented as intraventricular hemorrhage with hydrocephalus in a boy, causing a diagnostic dilemma. Although, preoperative modalities like computed tomography, magnetic resonance image and serology generally help in establishing the diagnosis, but hydatid cysts with unusual localizations and atypical imaging findings may complicate the diagnosis and need be considered in the differential diagnosis of all cystic masses in all anatomic locations, especially in endemic areas, so as to reduce the patient morbidity and mortality.

Key words: Cystic tumor, echinococcosis, intraventricular hydatid cyst

Introduction

Hydatidosis or echinococcosis, is a zoonotic disease caused by the infestation of the larval form (hydatid cyst) of tapeworm, *Echinococcus*. It is endemic in many countries including India.[1-4] Liver is the most frequently involved organ (75%), followed by lungs (15%).[1,8] Rarely, (1.6-5.2% cases), the central nervous system (CNS) is affected, where the cysts occupy mostly parenchyma and very rarely ventricles.[1,2,6-12] We report a case of isolated, solitary, primary intraventricular hydatid cyst in a boy that mimicked a bleeding cystic tumor and presented as intraventricular hemorrhage with hydrocephalus. None of the earlier reported cases has such atypical presentation.

Case Report

A 24-year-old male patient presented with complaints of tinnitus and decreased hearing for 1-month, headache and vertigo for 15 days, and fever, altered sensorium, nausea, and vomiting for 2 days. On examination, his general condition was poor. He was febrile, drowsy and disoriented; pupils were bilaterally equal and reactive to light. Fundoscopic examination showed bilateral papilledema. Hematological examination revealed raised erythrocyte sedimentation rate (60 mm/h) and leukocyte count of 18,000 mm$^3$. Both computed tomography (CT) scan [Figure 1] and magnetic resonance image (MRI) head [Figures 2 and 3] were of the impression of an intraventricular cystic tumor, with a speckled calcification with intraventricular hemorrhage and hydrocephalus. Radiological examination of the thorax and abdomen did not reveal any other cyst or tumor. In view of the serious neurological condition of the patient, surgery was performed.

A right parieto-occipital craniotomy with posterior parietal transcortical transventricular approach, with near total removal of mass, was done. The mass was yellow-brown, highly vascular, soft, cystic, suctionable, attached to the choroid plexus in the trigone of the lateral ventricle, extending...
in almost whole of the lateral ventricle and third ventricle. Intraoperative crush biopsy revealed only blood admixed with some normal brain tissue, and was inconclusive. The mass was very vascular and bleed profusely during surgery. Hence, the patient required substantial blood transfusions both intraoperatively and postoperatively. The histopathological examination of the resected mass revealed multiple laminated membranous bits of hydatid cyst scattered everywhere, with areas of hemorrhage and normal brain tissue [Figure 4]. There was no evidence of any neoplastic pathology, thus, confirming the diagnosis of an intraventricular hydatid disease.

Postoperatively, the patient showed significant neurological improvement. He was given broad spectrum antibiotics, anticonvulsants and steroids. Later on, albendazole was added for 4 months after histopathology revealed hydatid cyst. The patient is on follow-up for the last 3 months, and there is no evidence of any recurrence.

Discussion

In 50-75% of cases, CNS hydatidosis is found in the pediatric and adolescent age groups. CNS hydatid cysts are commonly parenchymal, located in the supratentorial region, and involve the territory of the middle-cerebral artery, especially, the parietal lobe.[1-3,6,8] Hematogenous spread of the disease can involve unusual sites in the brain.[8] A few cases of intracranial hydatid cysts with unusual locations such as parasellar epidural space, sella, cavernous sinus, cerebellum, thalamus and pons, have been reported.[1] Although involvement of the
ventricular system is found in 15-30% of neurocysticercosis cases, it is extremely rare in hydatidosis.[6,13] Khaldi et al. in a large series of 155 cases of brain hydatosis hospitalized between 1965 and 1998, reported only three cases of intraventricular hydatid cysts.[4]

Hydatid cysts constitute about 2% of all intracranial space occupying lesions even in countries in which the disease is endemic.[2,3,5] The incidence of intracranial hydatid in India is 0.2%.[3] Cysts in the CNS are frequently found in association with systemic dissemination particularly to the liver (65%) and lungs (25%). However, isolated involvement of the brain or spinal cord has also been observed, especially in children.[5] In our patient also, there was no radiological or clinical evidence of hydatid disease elsewhere in the body.

Cerebral hydatid cyst is generally solitary but may be multiple when it ruptures spontaneously or due to trauma or surgery; the cyst is always solitary when the primary site is in the brain.[2-4] Solitary intraventricular hydatid cyst, as seen in our case, was reported by Kamath et al. in a 6-year-old boy.[1] Hydatid cyst may be unilocular caused by Echinococcus granulosus or multilocular caused by Echinococcus multilocularis. E. granulosus is the more common type, whereas E. multilocularis is less common, but more invasive, mimicking a malignancy.[3]

Depending on the size and the site of the hydatid cyst, patients may present with focal neurological deficit and features of raised intracranial pressure.[1] In adults, focal neurological deficit such as hemiparesis, hemianopia, speech disorders or epileptic seizures are usually first to appear, while in children the presentation is dominated by the signs of raised intracranial pressure.[6,13,14] Postural changes may initiate changes in cyst location in the ventricular system, and they may be the cause of hydrocephalus.[6] Our patient presented with features of raised intracranial pressure.

For the planning of appropriate surgical management and for a better prognosis, it is important to be aware of intracranial hydatidosis preoperatively. Imaging modalities like CT and MRI, are not only diagnostically important but also helpful in the treatment. By CT scanning, the most superficial part of the cyst can be identified for a cortical incision, thus minimizing the risk of rupturing the cyst.[15] In our case, the radiological findings were not typical of a unilocular hydatid cyst and the impression of an intraventricular cystic tumor was made preoperatively.

Spontaneous rupture of cerebral cysts into the ventricles or mass effect of cysts on the cerebrospinal fluid pathways may produce hydrocephalus.[2] In our case, occurrence of fever, altered sensorium, nausea and vomiting 2 days before admission, superadded on the initial complaints of headache and vertigo suggests the possibility of rupture of hydatid cyst within the ventricle. Adherence of the cystic mass to choroid plexus and its high vascularity, confirmed intraoperatively, might have caused intraventricular hemorrhage, and hydrocephalus due to raised intracranial pressure following spontaneous cyst rupture.

To the best of our knowledge, this is the first-case report in which an isolated, solitary, primary intraventricular hydatid cyst in a young male, with no relevant past history and no evidence of hydatid cyst elsewhere in the body, mimicked a bleeding cystic tumor and presented as intraventricular hemorrhage with hydrocephalus.

The clinicians, radiologists and pathologists are required to express a high index of suspicion and familiarity with imaging findings while managing all intracranial cystic masses, so that a proper plan of management is formulated, and hydatid cysts can be excised in toto without rupture, avoiding the complications arising, as a result, of cyst rupture like anaphylactic reaction and dissemination of hydatid cysts. Furthermore, such patients should be carefully followed postoperatively for any recurrence.

**Conclusion**

Echinococcosis is a common disease in our country that which usually involves liver and lungs. Although, preoperative modalities like CT, MRI and serology, can generally help establish the diagnosis of hydatid disease, but hydatid cysts with unusual localizations and atypical imaging findings may complicate the diagnosis and need be considered in the differential diagnosis of all cystic masses in all anatomic locations, especially, when they occur in endemic areas so as to reduce the patient morbidity and mortality.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**