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Case Report

Camalote sign in intraventricular hydatid cyst: A rare presentation of uncommon disease

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ABSTRACT

Background: Hydatid cyst is a common zoonotic condition in endemic areas. Intraventricular hydatid cyst is a rare entity with less than 50 cases published in literature. *Floating water lily sign* (also called *Camalote sign*) is very rare in intracranial hydatid cysts. Only a single case report of this sign exists in literature in intraventricular hydatidosis suggesting rupture of hydatid cyst leading to CSF dissemination and frequent poor outcome.

Case Description: This case report describes the successful management of a 5-year-old child who presented with signs and symptoms of raised intracranial pressure due to large intraventricular hydatid cyst in the right frontal horn, and magnetic resonanace imaging of the brain showed hydatid cyst with typical camalote sign.

Conclusion: Although very rare, the presence of camalote sign in intraventricular cysts is very categorical in establishing preoperative diagnosis of hydatid cyst, especially in endemic areas.

Keywords: Camalote sign, Floating water lily sign, Hydatid cyst, Intraventricular cyst

INTRODUCTION

Cerebral hydatid cysts involve <2% of intracranial space-occupying lesions, usually located in supratentorial white matter.[5] It was first described in 1550 by Paranoli in the corpus callosum.[6] Other rare sites described include ventricles, cisterns, brainstem, basal ganglion, and extradural sites. Literature review suggests that less than 40 cases are described for intraventricular location of hydatid cyst but only one case report has revealed *floating water lily sign* (also called *Camalote sign*) in an intraventricular cyst.[4,9,10] Here's description of one such rare presentation of intraventricular hydatid cyst with evident *Camalote sign* in radiological image. It's appropriate management including surgical approach and resection is also elaborated.

CASE REPORT

A 5-year-old male child was brought to our department with chief complaints of holocranial headache, irritability, occasional vomiting for 3 months, and altered sensorium for 3 days. On examination, patient was in altered sensorium with GCS of E3V4M6. Magnetic resonanace

imaging (MRI) brain were suggestive of a large ruptured intraventricular cyst with irregular floating cyst wall in the right frontal horn giving a typical *Camalote sign* appearance [Figure 1]. It was causing obstruction of foramen of Monro with gross hydrocephalus. On the basis of MRI findings working diagnosis of ruptured hydatid cyst was made. Under steroid cover, Patient was planned for transcortical approach as the lesion was present in the frontal horn of the right lateral ventricle causing its ballooning and thinning out of the overlying cortex. Patient underwent right frontal craniotomy and ventricle was opened up through transcortical approach to access the cyst, taking special care to prevent spillage of cyst contents. The cyst wall was free-floating in clear CSF. Cyst was deroofed and cyst contents were aspirated followed by instillation of hypertonic saline in the cyst wall and aspiration again. After completely emptying the cyst of it's contents, its wall was dissected out [Figure 2]. Ultrasonogram of the abdomen and X-ray of the chest were done to evaluate for any other primary lesion, but these didn't reveal any pathology.

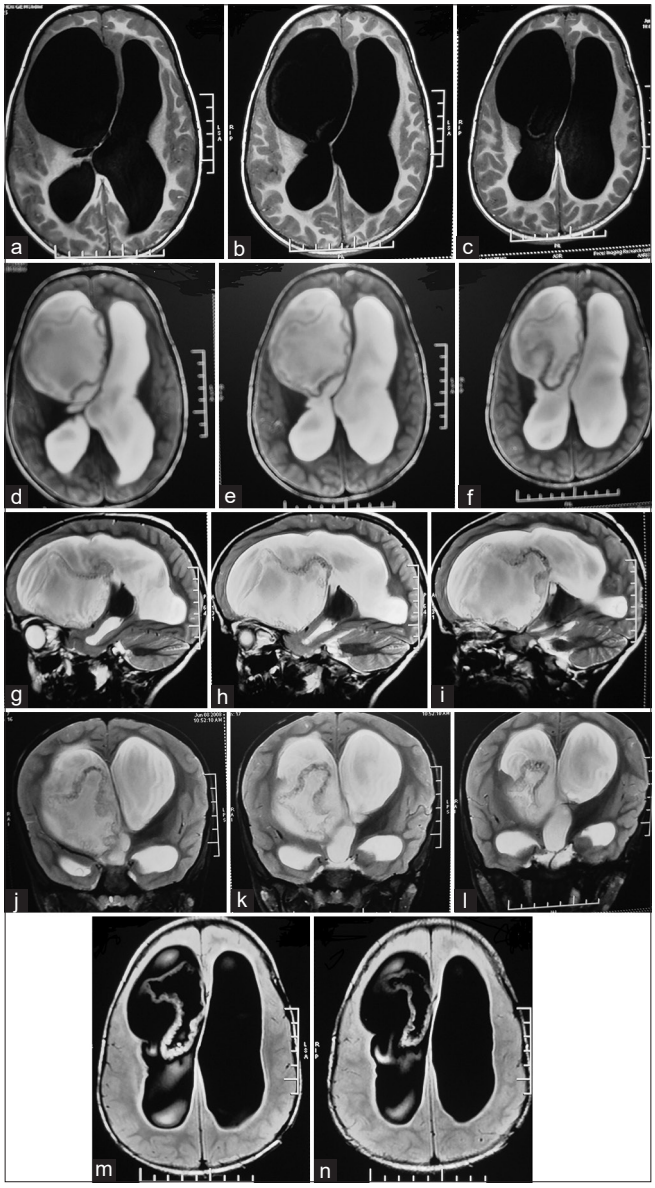

Figure 1: (a-c) T1 MRI axial image sequences showing hypointense intraventricular cyst 1 (d-f) T2 MRI axial image sequences showing hyperintense intraventricular cyst with ruptured hypointense wall in the right lateral ventricle (Camalote sign) 1 (g-i) T2 MRI Saggital image sequences again showing the ruptured cyst and cyst wall sedimenting down 1 (j-l) T2 MRI Coronal image sequences confirming the ruptured hydatid cyst1. (m and n) MRI FLAIR images in axial cuts distinctly defining the cyst wall.

Histopathology also confirmed the diagnosis [Figure 3]. Patient recovered well after the surgery and there was no need of CSF diversion. He was given three cycles of albendazole therapy of 1 month duration after intervals of 14 days between each cycle. Patient symptoms also regressed and there was no recurrence of the cyst during 3 years of follow-up.

DISCUSSION

Hydatidosis is common endemic zoonotic condition in grazing areas of world caused by flatworm cystodes: *Echinococcus*

Figure 2: Excised ruptured hydatid cyst wall.

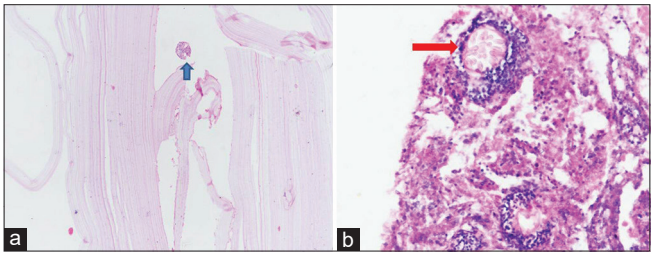

Figure 3: (a) Lamellated membranes of hydatid cyst with scolex (blue arrow) (H and E, ×100) 3 (b) High power photomicrograph of cross-section of a scolex at the level of invaginated head bearing the hooklets (red arrow).

granulosus (unilocular) and *Echinococcus multilocularis* (alveolar form). Humans as accidental hosts, ingest larval form of *Echinococcus* with contaminated food. From the intestine, they are taken to liver through portal circulation, from where they may spread to different parts of body through systemic circulation. Common site of involvement is intraparenchymal especially MCA territory distribution (most common in parietal lobes) where it reaches as embolic spread.[9] Case reports including other uncommon sites (ventricles, cisterns, skull, brainstem, cerebellum, orbits, and extradural sites) exist in literature.[4] On extensive literature search, we could find two important review articles summarising the previously reported intraventricular hydatid cyst which comprise total of 30 cases in large series and 12 cases as individual case reports.[4,9] Echinococcus oncospheres reach ventricles by embolic spread to choroid plexus.[1] Hydatid cysts in any part of the body have three layers. (i) Outer pericyst: layer made from fibrous tissue deposited by host due to granulomatous reaction around the cyst. (ii) Ectocyst: the acellular layer which allows passage of nutrients. (iii) Endocyst: layer comprising the thin and translucent germinal layer and the laminated membrane which produces scolices.[2] Endocyst secretes clear fluid into the cystic cavity. This fluid is rich in various electrolytes, proteins, lipids, and polysaccharides and is highly antigenic which may cause anaphylactic reaction if it comes in contact with normal host tissues.

World Health Organisation has classified pericyst being made up of host tissues without surrounding edema. The cyst wall do not enhance with contrast imaging except in rare cases having secondary infection in cyst. The WHO classifies hydatid cyst in the six stages on basis of its evolution [Table 1].[8,12]

Computed tomography (CT) and MRI features of hydatid cyst are of a spherical well defined, thin walled, homogenous cystic lesion with CSF like fluid density. These imaging features mandates ruling out differential diagnosis of arachnoid cyst, dermoid or epidermoid cyst. Calcification or septations may be present rarely. Presence of daughter cyst is pathognomonic for hydatid cyst but uncommon in radiology.

Table 1: WHO classification of hydatid cyst, based on six stages of it's evolution

Stage	Nature of cyst	Imaging feature
CL	Active	Unilocular cyst, anechoic, no wall visible
CE1	Active	Univesicular anechoic cystic lesion with double line sign, hydatid sand
CE2	Active	Multiseptated cyst (honeycomb/ rosette-like)
CE3a	Transitional	Multiseptated cyst (honeycomb/ rosette-like)
CE3b	Transitional	Cyst with detached membranes, water lily sign
CE4	Inactive	Cyst with daughter vesicles in solid matrix
CE5	Inactive	Cyst with heterogeneous degenerative content (hypo/hiperechoic) without daughter vesicles
CE6	Inactive	CE 4 with calcified wall

Conventional water lily sign is present on ultrasound, X-rays, CT scan or MRI in stage 3A when the endocyst wall is detached from the pericyst and floats freely into the cystic fluid.[3] This sign has been well described in hepatic and pulmonary hydatid cyst but very rare in CNS hydatidosis. Sadashiva *et al*. described this sign in intraventricular hydatid cyst as a floating water lily sign.[10] It was described in postintervention MRI when the cyst got ruptured iatrogenically during CSF diversion. In other case report by Thakur *et al*. in 2017, there was spontaneous rupture of the cyst and multiple daughter cysts were seen in temporal and occipital horns.[13] Although typical Camalote sign was not described however imaging in that case report appeared as classical water lily sign. In our case, there was rupture and complete separation of endocyst wall giving a typical floating water lily sign with intact outer layers. Clinical presentaton of hydatid cyst is mainly due to the mass effect caused by the cyst on surrounding structures commonly leading to raised ICP

The conventional Dowling's technique has been described with an aim to take out the intact cyst by hydrostatic expulsion holds good for parenchymal cysts. However, intraoperative rupture has been described in around 25% of cases even with the use of this technique.[11] Especially for large and deep-seated cysts including intraventricular one, there are more complications like intraventricular/parenchymal hemorrhage, subdural effusions, and meningitis. Few authors have described opening/puncturing the cyst and aspiration of cyst contents followed by removal of the shrunken cyst wall to avoid such complications which can be considered as modification of PAIR technique (percutaneous aspiration, injection, and respiration) which is widely accepted for hepatic hydatid cysts.[7] Albendazole is always recommended in all such cases for several months with 1 month cycle of albendazole followed by 14 days of therapy free intervals.[7]

CONCLUSION

Intraventricular location of hydatid cyst is very uncommon but it should be kept as a differential diagnosis especially in endemic areas. The presence of camalote sign on imaging in such rare sites is very helpful in confirming the diagnosis preoperatively for appropriate surgical planning in such cases.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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