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Rare Case of Intracranial Calcified Hydatid Cyst

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Abstract
Cerebral hydatid disease is a rare manifestation of echinococcosis, constituting significant fraction of all intracranial mass occupying lesions. We report a case of 12-year-old boy, a shepherd by occupation, who presented to us with headache, vomiting and giddiness. A calcified intracranial cystic mass was found on radiological investigations and it was surgically excised totally, with no post operative complications. Calcification is quite rare in cerebral hydatid cyst. The radiological presentation, differential diagnosis and treatment modalities are discussed in relation to this case.

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INTRODUCTION

Echinococcosis, which is often referred to as hydatid disease or echinococcal disease, is a parasitic disease that affects both humans and other mammals. Cerebral hydatid disease is very rare, representing only 2% of all cerebral space occupying lesions even in the countries where the disease is endemic and calcification is even more rarer [1].

CASE REPORT

A 12 year old boy, shepherd by occupation presented to Department of Neurosurgery with three months history of headache, vomiting and giddiness. There was no history of trauma, surgery, seizures and any history of hydatidosis. The general physical and neurological examinations were normal. except for the bilateral papilloedema. Brain CT revealed 6.5 cm x 5.7 cm rounded complex cystic mass lesion in left temporoparietal lobe with densely calcified walls, thin internal minimally enhancing septations and surrounding gliotic changes (Figure-1). Magnetic resonance imaging shows large well defined spherical mixed intensity, predominantly cystic mass lesion with few solid components within, situated in the left parietal lobe. It appears heterogeneously hyperintense on T2W image with thick hypointense wall. On T1W images cystic component appears hypointense.

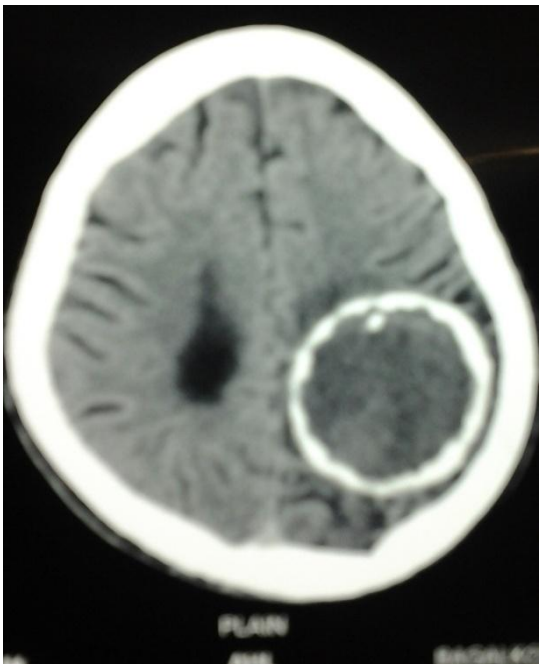

Figure-1: Brain CT scan reveals cystic mass lesion in left temporoparietal lobe with densely calcified walls

minimal perilesional oedema is noted with mass antero-medially compressing posterior part of body and trigone of left lateral ventricle (Figure-2).

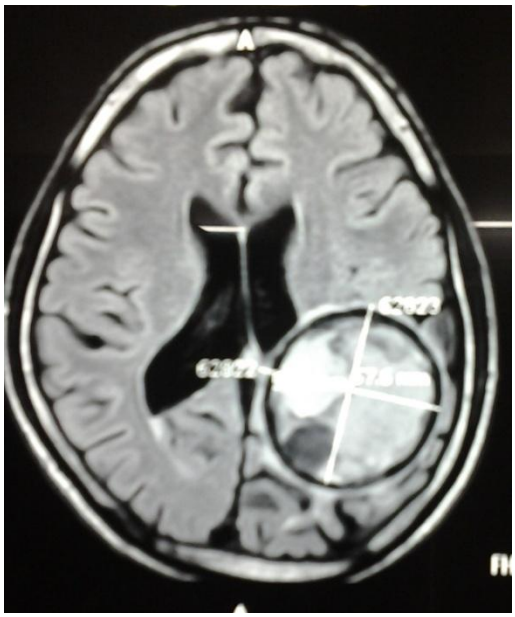

Figure-2: MRI shows large well defined spherical mixed intensity cystic lass lesion

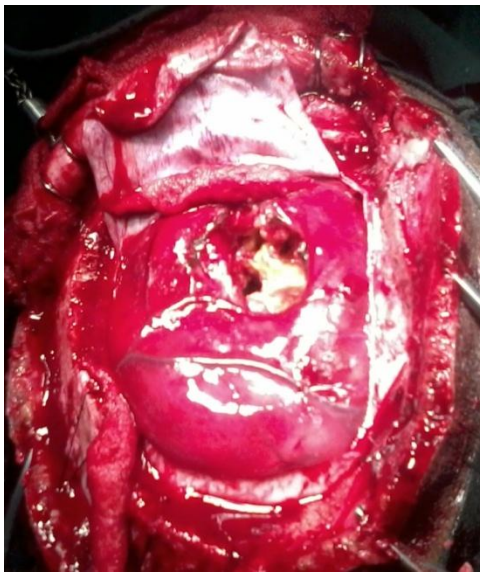

Figure-3: Opened hydatid cyst with yellowish semi solid contents

Radiological investigations of thorax and abdomen disclosed no evidence of hydatid disease. Serologic tests including enzyme-linked immunoabsorbant assay (ELISA) test and indirect hemagglutination (IHA) test for hydatid disease were negative.

A right parieto-occipital craniotomy was performed and on opening the dura, the superficial surface of cyst was adherent to dura and the deeper surface to the ependyma of occipital horn of lateral ventricle. We opened the cyst and the yellowish semisolid contents were sucked (Figure-3). The cyst wall was thick (2mm), calcified and was excised in toto with gentle dissection. The cavity was then irrigated well with chloromycetin solution and hypertonic saline. The postoperative course was uneventful. Histopathological examination confirmed hydatid disease. Patient was treated with oral antiepileptics and Albendazole 400 mg twice a day for 1 month.

DISCUSSION

Hydatid disease or Echinococcosis is a parasitic zoonosis caused by adult or larval stages of cestodes belonging to the genus Echinococcus (family Taeniidae). There are two major species of echinococcosis which are Echinococcus granulosus and Echinococcus multilocularis, which cause cystic echinococcosis (CE) and alveolar echinococcosis (AE), respectively.

The definite hosts of echinococcus are various carnivores, the common being the dog. All mammals (more often being sheep and cattle) are intermittent hosts. Humans get infected through the faeco-oral route by ingestion of food or milk contaminated by dog faeces containing ova of the parasite or by direct contact with dogs.[1]

CE is endemic in areas such as South and Central America, the Middle East, some sub-Saharan African countries, China, and the former Soviet Union. Alveolar echinococcus deserves attention due to the high fatality rate of untreated patients and occurs across the Northern Hemisphere, in parts of central Europe, Russia, western China, areas of North America, and Northern

Africa. In the Indian subcontinent, it is more commonly seen in the Kurnool district of Andhra Pradesh, Madurai district of Tamil Nadu and in Punjab. intracranial hydatids from India have reported its incidence as 0.2% of all intracranial space occupying lesions. [2,3]

The liver (50–77%) and the lung (8.5–43%) are the organs most commonly involved. Involvement of heart, brain, spinal cord, kidneys, bone and eye rare, but can lead to significant morbidity and mortality [4]. Intracranial hydatid disease is rare, with reported incidence of 1-2% of all cases with hydatid disease and are 2-3 times more common in children than in adults [1,3].

Cerebral hydatid cysts are usually supratentorial, the infratentorial lesions are quite rare. The parietal lobe is the most frequently involved region [5]. Intracranial hydatid cysts are commonly solitary. Multiple intracranial cysts are rare and are also be classified as primary or secondary. The primary cysts are formed as a result of direct infestation of the larvae in the brain without demonstrable involvement of other organs; secondary multiple cysts result from spontaneous, traumatic or surgical rupture of the primary intracranial hydatid cyst. Calcification is very rare but seen more commonly in infestation with E. multilocularis[6]

Growth of hydatid cyst in different organs of the body is variable and depends both on patient factors, parasite factors, location of organ of cyst, host reaction and presence of any complications. There is no consensus on the growth rate of the hydatid cyst of the brain and has been variably reported between 1.5-10cm/year[7]. In addition, the presence of patent ductus arteriosus, which might remain for several months after birth, can explain why primary cerebral hydatid disease is more common in childhood [8].

It has been postulated that infection occurs early in childhood. Since brain has a softer tissue, a hydatid cyst in brain grows faster than in other organs. Thus, when hydatid cyst of the brain is large enough to produce symptoms, the cysts in other organs are too small to be detected by clinical and radiological evaluations [9]. Although calcification of all cysts in liver is quite common, this is quite rare in the brain. A calcified cyst in the brain may be called a ‘dead cerebral hydatid cyst’ and it indicates chronicity of the cyst [10].

The patients with intracranial hydatid cysts usually present with focal neurological deficit and features of raised intracranial

pressure; the latter may be due to the large size or due to interference with pathway of CSF flow [11].

A high index of suspicion, radiological investigations as well as histopathological examination is necessary in establishing the diagnosis of hydatid disease at unusual sites in the body.

Headache and vomiting were the most commonly reported symptoms in other series as well as in our patients. Other symptoms such as hemiparesis, seizures, visual field alteration and gait disorders, may vary with the location of the cyst [12].

Diagnosis of hydatid disease is based on the patient's history, clinical findings, serum biochemical profiles, serologic tests and pathologic diagnosis. Serologic tests usually allow hydatid cysts to be distinguished from non-parasitic cysts and abscesses [13].

CT and MR imaging demonstrate a well-defined oval or cystic mass with an attenuation or signal intensity similar to that of cerebrospinal fluid, Fine peripheral rim enhancement in its fibrous capsule is seen. there is no surrounding edema, unlike the abscess or the cystic tumors. The edema may be subtle due to chronic nature of the lesion and may not cause mass effect or midline shift. Although not always seen, contrast enhancement of the lesion has been proposed because of blood-brain barrier disruption caused by the inflammatory reaction.[14] Peripheral ring-like, heterogenous, nodular, and cauliflower-like enhancement patterns have all been reported in different presentations of the infestation by AE [15].

Calcification of cyst wall is rare (less than 1%). Calcification and surrounding edema are common findings of the lesions, as observed in the present case. Differential diagnoses of intracranial hydatid include porencephalic cyst, arachnoid cyst, metastasis, tuberculosis, fungal infections, cerebral abscess and cystic tumors of the brain [16]. The treatment of hydatid cyst is surgical and the aim of the surgery is to excise the cyst in toto without rupture to prevent recurrence and anaphylactic reaction. Various surgical options were summarized by Iniquez, include puncture and aspiration of the cyst fluid through a small hole in the cyst wall, cortical incision over cyst and expulsion of hydatid cyst by insufflation of air in the contralateral ventricle, and the most commonly done procedure designed to give birth to the intact cyst is by irrigating saline in the cyst wall–brain interface [17].

CONCLUSION

The presence of a hydatid cyst in the brain without cysts in other organs has even

now not been fully explained. Our case is therefore also a primary cerebral hydatid cyst, a condition rare in adults. In conclusion, hydatid cyst should be included in the differential diagnosis when a calcified brain lesion is found in patients from an endemic echinococcosis area.

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