Intracranial hydatid cyst: a report of five cases and review of literature

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Abstract

The authors present five cases of intracranial hydatid cysts managed at the department of Neurosurgery, King Edward Memorial Hospital, Mumbai, between 1984-1997. The mean age of presentation was 13.4 years. Four patients (80%) were in the first decade of life. All patients presented with focal neurological deficit and clinical features of raised intracranial pressure. Radiological investigations included computerised tomography (CT) scan in three cases, CT and magnetic resonance (MR) scan in one case and accidental cystogram in one case. Two patients had multiple intracranial cysts. One patient had a solitary cyst in the lateral ventricle. Commonest location was in the parietal lobe (3 cases). Total excision of the cyst was done in all five cases. Recurrence was seen in two cases, probably as a result of rupture of the cyst during first surgery. The features of this rare disease are retrospectively analyzed in this presentation and the literature is reviewed.

Introduction

Hydatid disease is caused by the infestation of the larvae of taenia echinococcus. 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. The eggs loose their enveloping layer in the stomach, releasing the embryos. The embryos pass through the wall of the gut into the portal system and are carried to the liver where most larvae get entrapped and encysted. Some may reach the lungs and occasionally, some may pass through the capillary filter of the liver and lungs and get entry into the systemic circulation. These may even reach the brain. In India, the hydatid disease is more commonly seen in the Kurnool district of Andhra Pradesh, Madurai district of Tamil Nadu and in Punjab. We have analyzed five cases of the hydatid cysts and discussed their mode of presentation, radiological features and outcome. The relevant literature is reviewed.

Material and methods

We retrospectively analysed five cases of intracranial hydatid cysts managed at K.E.M. Hospital, Mumbai between 1984-1997. A detailed recording was made of the presenting features, neurological signs, investigations, type of surgery, recurrence of hydatid disease and other therapeutic modalities.

Results

Five cases of intracranial hydatids treated during the period of 1984-1997 at K.E.M. Hospital Mumbai represented an incidence of 0.05% of all space occupying lesions operated during this period. The mean age of presentation was 13.4 years. Four patients (80%) were between the age group of 8-10 years and remaining one case was thirty years old. The male to female ratio was 3:2. In two cases, history of contact with pet dogs was present. Three patients came from rural areas while two came from urban areas. The duration of symptoms varied from 1 month to 2 years. The clinical features are described in [Table I]. These varied depending on the location of cyst in the brain. Hemiparesis was the commonest finding present in all the cases. Papilloedema was seen in four cases. Seizures were present in two cases. In three
patients, radiological investigations included CT scan. In one case, both CT and MR scans were done. Accidental cystogram was diagnostic in one case. In the latter case, following the features of raised intracranial pressure, an attempted ventriculogram resulted in the inadvertent puncture of the cyst with resultant demonstration of smooth walled spherical frontal cyst [Figure 1]. One patient had solitary cyst in the lateral ventricle [Figure 2] and one had large solitary cyst in parietal lobe [Figure 3]. Multiple cysts were found in the remaining two cases (3 cysts in the parietal lobe and 8 cysts in the temporoparietal lobe) [Figure 4]. X-ray chest was done in last four cases. These investigations failed to reveal any associated hydatid cyst in lungs and abdomen.

The hydatid cysts were totally excised at first surgery in all the five cases. During surgical resection, rupture of cysts occurred in three cases (60%). Two had multiple cysts and one case had solitary frontal cyst. Diagnosis of hydatid was suspected preoperatively in two of these cases and all necessary precautions to prevent rupture and dissemination of hydatid were taken at the surgery. In one case with rupture, hydatid cyst was not suspected prior to surgery. Following rupture, anaphylactic reaction was not seen in any of these cases. Two patients, one with solitary cyst in the frontal lobe and second with multiple cysts in temporoparietal lobe had recurrence of hydatid cysts and presented with multiple cysts after 6-12 months of first surgery. Both of these cases were reoperated and total removal of all cysts was done at second surgery. There were extensive adhesions between the cyst wall and dura, due to which rupture occurred at second surgery as well. Albendazole (10mg/kg) was given for one month in both patients with recurrence. All five patients have been followed up from six months to eight years with mean follow up of three years. There was no further recurrence. All patients have shown good recovery from neurological dysfunction.

> Discussion

Intracranial hydatid disease is rare, with reported incidence of 1-2% of all cases with hydatid disease.[4] Hydatid disease is endemic in the middle east, Mediterranean countries, South America, North Africa and Australia.[5] Cerebral hydatid disease is more common in paediatric population[1][6] and 80% of patients in the present series were children. This high incidence in children is probably related to patent ductus arteriosus.[7] None of the patients in the present series had a patent ductus arteriosus. History of direct contact with dogs is not available in all the cases, as infection can be acquired by eating contaminated food and milk. In the present series, history of contact with pet dogs was available in two cases.

Intracranial hydatid cysts are more frequently located in the supratentorial compartment. The parietal lobe is the commonest site and was seen in three cases in the present series. All four cases reported by Dharker et al[8] and three out of five cases of intracranial hydatid cysts reported by Balasubramaniam et al[9] had parietal lobe involvement. The other less common sites reported are skull[5] cavernous sinus,[10] eye ball,[11] pons,[12] skull,[9] extradural,[13] cerebellum and ventricles.[6] Solitary hydatid cyst in the lateral ventricle, as seen in one case in the group of patients being reported, is relatively rare site for intracranial hydatid cyst. The cerebral hydatid cysts are slow growing and present late when they increase in size and become large. There is no consensus on the growth rate of the hydatid cysts of the brain and has been variably reported between 1.5-10 cm/year.[1][14] Formation of the large multiple hydatid cysts within six to twelve months of the first surgery following total excision in two cases in this report suggests that probably the growth rate is higher.

Intracranial hydatid cysts are commonly solitary. Multiple intracranial cysts are rare. Oral et al[8] found only three cases of multiple cysts in their series of 33 cases and Lunardi et al[7] found 2 cases in their series of 12 cases. We observed fairly high incidence of 40% (two cases) of multiple cysts in the present series. Both of our patients with recurrence had multiple cysts.

Intracranial hydatid cyst may 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. In primary multiple cysts, each cyst has a separate pericyst with brood capsule scolices and these originate from multiple larvae affecting brain after crossing the gastrointestinal tract, liver, lungs and right side of heart without affecting them. The primary cysts are fertile as they contain scolices and brood capsules, hence rupture of primary cyst can result in recurrence. The secondary multiple cysts results from spontaneous, traumatic or surgical rupture of the primary intracranial hydatid cyst and they lack brood capsule and scolices. The secondary intracranial hydatid cysts are therefore, infertile and the resultant risk of recurrence after their rupture is negligible. Primary multiple cysts are uncommon and isolated case reports of primary multiple hydatid cysts have appeared in the literature.[15][16] Nurchi et al[15] while reviewing the literature found only eleven reported cases of primary multiple hydatid cysts. In the present series, case number 3 [Table I] was a case of primary multiple hydatid cysts and its rupture at surgery caused recurrence of multiple intracranial hydatid cysts.

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. Erasini et al[4] observed that 18 out of 19 cases presented with raised intracranial pressure. Four cases had seizures. All patients in the present series had focal neurological deficits and features of raised intracranial pressure. Two patients had seizures. MR and CT scans characteristically show hydatid cyst as a spherical, well defined, non enhancing cystic lesion without peripheral oedema.[17][18] The fluid density is generally equal to that of CSF on both CT and MR scan. A fine rim of peripheral enhancement with perilesional oedema may be seen in the presence of active inflammation.[18] MR scan may show a low density cyst wall[17] and relations with surrounding structures is better delineated than on CT scan. We could not identify scolices on MR scan. Other reports on MR findings[15][17] showed similar findings. Kohli et al[19] performed in vivo and in vitro MR spectroscopy (MRS) studies in a patient of intracranial hydatid cyst. Besides lactate, alanine and acetate, a large resonance for pyruvate was observed. MRS pattern appeared different from the other cystic lesions of brain and they suggested MRS as an adjunct to imaging in the differential diagnosis of intracranial hydatid. Role of MRS in monitoring drug therapy was also highlighted.

The treatment of hydatid cyst is surgical and the aim of surgery is to excise the cyst in toto without rupture to prevent recurrence and anaphylactic reaction. Various surgical options as summarized by Arana-Iniquez[20] 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 contra lateral ventricle and the most commonly done procedure designed to give birth to the intact cyst by irrigating saline between cyst wall-brain interface. This is possible because of minimal adhesions around the cyst wall.

Only a few reports are available mentioning the efficacy of drug therapy. Isolated case reports[21][22] showed complete disappearance of multiple intracranial hydatid cysts with Albendazole therapy in a daily dose of 10 mg/kg, taken three times a day for four months. Albendazole is a broad spectrum oral anthelmintic drug, which act by blocking glucose uptake of the larvae and the adult worm. The glycogen storage is depleted and thereby decreasing the ATP formation resulting in the death of the parasite. Golematis et al[23] analyzed 44 patients who were treated with albendazole and found that the large cysts decreased in the size, while the smaller ones disappeared.
Herein, we report a 66 year old woman who was diagnosed to have intrabiliary rupture of liver hydatid cyst with demonstrative computed tomography, magnetic resonance imaging, and magnetic resonance cholangiopancreatography findings, with a review of the literature.

Keywords. Common Bile Duct.  Anaphylactic shock, cyst infection of the biliary tree, and rupture into the peritoneum are the most common complications. Intrabiliary rupture is reported to be seen in a range of 6,1-17% [2][4]. In a study which was done when the ultrasonography (US) and computed tomography (CT) were not awailable, this range was reported to be 41% [4]. There are 2 types of intrabiliary rupture: frank and occult (silent) [5]. (1982). Æ¢Primary retroperitoneal hydatid cyst (a report of 3 cases and review of the literature).Æ¢ J Postgrad Med 28(2): 112-114B. PubMed. Akbulut, S., A. Senol, et al. (2010). Æ¢Primary retroperitoneal hydatid cyst: report of 2 cases and review of 41 published cases.Æ¢ Int Surg 95(3): 189-196. PubMed. Sirus, M., M. Zhiannpour, et al. (2006). Æ¢Omental and Retroperitoneal Hydatid Cyst: A Case Report.Æ¢ Iran J Radiol 3(4): 217-219. Homeida, M., W. Leahy, et al.Â® Sign up today for articles, videos, conference highlights and abstracts from peer-review publications by disease and condition delivered to your inbox and read on the go. Subscribe. About UroToday.