Volume : 4 | Issue : 7 | July 2015 • ISSN No 2277 - 8179

Research Paper

Hydatid Disease of the Central Nervous System; Retrospective Study of 20 Cases.

Medical Science

KEYWORDS : brain, spine, computed tomography, echinococcosis, magnetic resonance imaging, orbit, central nervous system, proptosis, albendazole

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ABSTRACT

Objective: Hydatid disease primarily affects the liver; the secondary involvement due to hematogenous spread may be seen in almost any locations e.g.., lung, kidney, spleen, bone, and central nervous system. We aimed to assess clinical findings, radiological investigations and treatment of cases with central nervous system (CNS) hydatid cyst disease.

Materials and method: the present study included 20 patients who were operated for CNS hydatid cysts disease from 1984- 2014. The clinical features, radiological investigations and treatment of cases are retrospectively analyzed and the pertinent literature is reviewed and compared the results with those reported in literature.

Results: fourteen patients were male and six were female .The mean age of the study population was 20 years (range 10-50 years). All the patients came from rural areas .Headache and motor deficits were the most common clinical symptoms in patients with intracranial hydatidosis where as the back pain and spinal compression syndrome were most frequent clinical presentation in case of spinal hydatidosis. The lesion was demonstrated by computed tomography (CT) and magnetic resonance imaging (MRI) as large smooth thin walled spherical shaped cystic homogeneous lesions with same density as cerebrospinal fluid (CSF) which showed no edema or contrast uptake in intracranial hydatid cyst disease. The location of cysts was intracranial in 11 cases (55%), orbital 2(4%) and spinal in 7(14%) cases. Two patients were infected cysts. In all patients with intracranial, the cysts were removed completely with Dowling's technique. All the spinal hydatid cysts are excised by doing laminectomy at the level of spinal involvement. Eight patients had ruptured cysts during operations. Recurrent or systemic hydatid cysts were treated medically. Pathological findings were consistent with hydatid cysts in all cases. During the follow up period which ranged from 5 months and 16 years, 8 had recurrence (40%).

Conclusion

Despite all the advances in imaging techniques and therapeutic methods ,CNS hydatidosis remains difficult to cure and the patient outcomes are not satisfactory especially in case of spinal involvement due to high incidence of recurrence.

Introduction

Hydatid disease is a parasitic disease that affects both humans and other mammals, such as sheep, dogs, rodents and horses. The cystic echinococcosis caused by E. granulosus is much more common than alveolar echinococcosis caused by E. multilocularis. Most common form found in humans is caused by larval stages of tape worm Echinococcosis Granulosus. The definitive hosts are carnivores such as dogs, while intermediate hosts are sheep and cattle. Humans are accidental intermediate hosts, are usually the dead end for the parasitic cycle. The adult worm resides in the small intestine of a definitive host. The eggs are passed in faeces of the definitive host. The egg is then ingested by the intermediate host. The egg hatches in the small intestine of the intermediate host and penetrate the small intestine and moves through the circulatory system into different organs commonly liver and lungs, where the larvae get entrapped and encysted and some pass through the capillary filter of liver and lungs and get into systemic circulation and reach the brain, bone and other organs[1, 2]

Methods

This study retrospectively assesses medical data of 20 patients diagnosed with CNS hydatid cysts with pathological confirmation and treated during hospital admission at Government General Hospital, Department of Neurosurgery, Kurnool, between 1984and 2014.The assessment included age, clinical symptoms, examination findings ,laboratory, radiological results, medical and surgical therapies and follow –up data. To reveal extra CNS involvement all patients underwent chest radiography and abdominal ultrasonography or computed tomography.

Results

Fourteen were male and six were female .Mean age of the study population was 20 years. All patients came from rural areas. The presenting symptoms were headache in 11 cases, nausea and vomiting in 8 cases, motor deficits in 5 cases, and visual disturbances in 2 cases. Papilloadema was observed in 6 cases (Table 1). Time from symptom onset to the admission was 30 days to 9 months. Ten cases underwent CT and 10 cases underwent MRI. The lesion size varied between 1cm and 6 cm. Eleven cases were intracranial, 2 are orbital and seven are spinal. The most common location was parietal region (40%). Three cases had multiple lesion localizations. The most common lesion sites in cases with multiple lesions were parietal and temporal lesions. Seven solitary lesions were located at the left hemisphere and four were located at the right hemisphere (Table2).

Table I: symptoms and signs

Symptoms and signs	Number of patients (%)
Headache	11(55%)
Nausea and vomiting	8(40%)
Papilledema	6(30%)
Hemiparesis	5(25%)
Seizures	3(15%)
Speech disurbance	2(10%)
Low backache	7(35%)

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Paraplegia	3(15%)
Proptosis	2(10%)
Decreased vision	2(10%)

Table II: Localization of cyst

Localization	Number of patients (%)
Parietal	9(45%)
Orbital	2(10%)
Temporal	2(10%)
Spinal	7(38.5%)

Table III: Distribution of spinal hydatid cysts

Level of the lesion	Number of patients (%)
Thoracic	2(28.6%)
Lumbar	3(42.8%)
Sacral	2 (28.6%)

Four patients were also detected to have extra cerebral organ involvement. CT demonstrated shift of midline structures in four cases. CT shows large smooth thin walled spherical shaped, cystic homogeneous lesions with no perifocal edema and no contrast uptake, which as the same density as CSF (figure; 1). MRI showed well defined round hypo intense lesions on T1 weighted images (fig; 2) and hyperintense lesions on T2 weighted images. Cysts walls were hypointense on T1 and T2 weighted images. Two cysts were complicated or infected showed perifocal oedema and contrast uptake. A large craniotomy and wide cortical incision were used to reach the brain tissue in most of the patients and the cysts were delivered by Dowling's method (fig; 3 and 4). In cases of spinal cysts the presenting symptoms were low backache in seven cases and paraplegia in two patients. Out of seven cases in spinal cysts two are dorsal (fig; 5), three are lumbar and two (Table III) are sacral (Figure 6). Plain X-ray showed multiple, well-defined, osteolytic expansile cavitatory areas without periosteal reaction or sclerosis. Computed tomography scan showed multiple cystic, osteolytic expansile lesions in vertebral bodies without enhancement of the lesion or the margins on the intravenous (IV) contrast study. Magnetic resonance imaging showed multiple cystic fluid-filled lesions with thin walls and irregular branching resembling a grape bunch on the axial, sagittal, and coronal images of the spine. The spinal cysts are removed by doing laminectomy at the level of spinal involvement. At the time of surgery multiple pearly, shiny, grape-like cysts were seen bulging from the spinal canal with multi-level paraspinal muscle involvement, which required extensive decompression and debridement through wide laminectomy. The two intra orbital cysts (fig; 7) are removed by orbitotomy. The proptosis decreased and vision improved (fig; 8) There was no mortality. The mean follow-up period was 5 months and 16 years .Three cases were drug controlled seizures, five had hemi paresis, two had speech disturbance. Intra-operative cyst rupture occurred in eight cases (40%) which were slightly higher compared to other series in literature. Eight cases (40%) had cyst recurrence. The average time for cyst recurrence was 14 months. Pathological findings were consistent with hydatid cysts in all cases. Albendazole was administered post operatively for a period of four months in all patients.

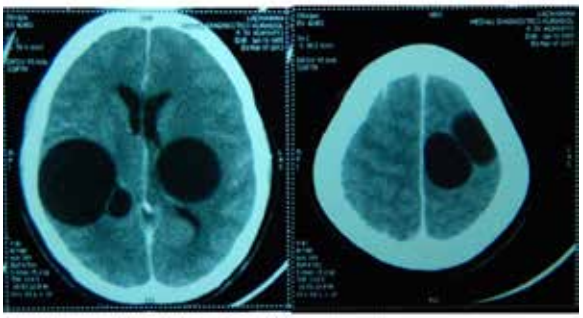

Figure 1; CT image shows multiple cysts (5) in different locations of brain in one patient

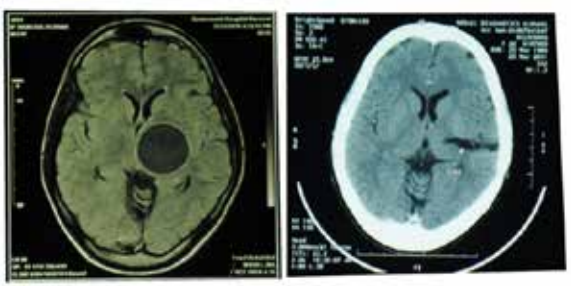

Figure 2; MRI image on left panel showing single unilocular cystic lesion in left parietal lobe of cerebral hemisphere and on the right panel post operative CT scan showing complete excision

Figure 3; per operative picture showing hydatid cyst

Figure 4; shows in total removal of five hydatid cysts.

Figure 5; MRI T1-weighted image of the thoracic spine showing intradural nonenhancing cystic lesion

Figure 6; MRI of lumbo sacral spine showing multiple hydatid cysts in sacrum

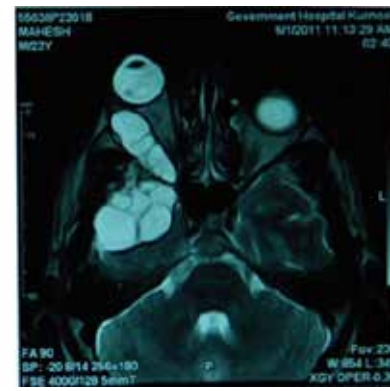

Figure 7; MRI shows multiple cystic lesions in right temporal lobe of brain and right orbit causing proptosis

Figure 8; shows intra orbital hydatid presenting with proptosis of right eye on the left panel and postoperative picture showing proptosis resolved on the right panel

Discussion

Intracranial hydatid cysts are rare, accounting for only 1-2% of all intracranial space occupying lesions[3]. Hydatid cyst of the orbit are rare ,is responsible for nearly 1% of all orbital growths[4]. The disease is endemic in Middle east ,Mediterranean countries, South America, North America and Australia. In India the hydatid disease is more commonly seen in the Kurnool district of Andhra Pradesh, Madurai district of Tamilnadu, and in Panjab[1, 5, 6, 7]. The incidence in India is 0.2%.Cerebral hydatid cyst is more common in paediatric population[6]. Cerebral hydatid cysts are usually supra tentorial and often involve middle cerebral artery territory because of the embolic nature of the infestation[5].The common location in parietal lobe has been reported by Gupta .S [8] and Abu-Eshy SA[9] in their series. Cerebral hydatid cysts commonly occur in children and young adults.

The human brain can be involved primarily via haematogenous route or by metastatic spread when a cyst ruptures in the heart or lung. The cysts may be single (primary) or multiple (secondary) [10, 11].The primary solitary cysts are formed as a result of direct infestation of 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 they originate from multiple larvae that affect brain after crossing the gastrointestinal tract, liver, lungs and right side of the heart without affecting them. Primary cysts are fertile as they contain scolices and brood capsule, hence the rupture of the primary cyst can result in recurrence. Multiple hydatid cysts usually results from cardiac embolus. Secondary multiple cysts results from spontaneous, traumatic or surgical rupture of the primary intracranial hydatid cyst and they lack brood capsule and scolices[2] .The secondary intracranial cysts are therefore infertile (acephaloceles)and the resultant risk of recurrence after

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their rupture is negligible. The cranial extradural variety is extremely rare as the physiological flow of blood to the brain is mainly through the internal carotid system, so the likelihood of the larvae travelling through the external carotid system is very low[9].The growth rate of hydatid cysts of the brain is higher than in other organs ,has been variably reported between 1.5-10 c.m/ year[12].The immune system can inhibit the growth of hydatid cyst in other organs ,except the central nervous system due to limited access to the immune system. Multiple intracranial cysts are rare. Onal *et al*[3, 6, 7] found only three cases in their series. Patients with intracranial hydatid cysts usually presents with focal neurological deficits and features of raised intracranial pressures, the later may be due to the large size or due to interference with pathway of C.S.F flow[13].Hematogenous spread is the most common route of dissemination of the ova to CNS. Most cysts are acquired during the childhood .Spinal hydatid disease is the most common form of the bone involvement .Although hydatid disease may be located anywhere in the brain ,it is most frequently located in the hemispheres ,particularly in the territory of middle cerebral artery mostly in the post rolandic part of the hemispheres .The orbit, hypothalamus , pons , the subarachnoid space of the cerebro pontine angle,, ventricular system, aqueduct of Sylvius and intracranial epidural space comprise the rare locations for growth of the hydatid cyst.

Radiologically the best diagnostic clue of a hydatid cyst is a single large thin walled, spherical non enhancing cyst, perilesional edema is usually absent. The two visible imaging components are the cyst and the pericyst. The pericyst is a peripheral capsule of the cyst. MR imaging is more sensitive in demonstrating the pericyst. CT is more sensitive in depicting the calcification. [14,]The diagnostic feature hydatid cyst on MRI is low signal intensity of cyst wall on T2 weighted image[15].The radiological differential diagnosis is from other cystic lesions such as cystic astrcytoma,cerebral abscesses , arachnoid cysts or epidermoid. Cystic astrocytomas shows higher attenuation values than hydatid or arachnoid cysts, due to higher protein content. The presence of surrounding edema and homogeneous enhancing solid component (mural nodule) of the tumor distinguishes it from a hydatid cyst. Abscesses show perifocal edema and peripheral enhancement. Arachnoid cyst has similar appearance as that of hydatid cyst but they are said to have an irregular border and are not spherical shape. Epidermoid can be differentiated usually by their lobulated, vessel engulfing, self moulding behavior.

The diagnosis of hydatid cysts is based upon clinical suspicion particularly in endemic areas. The Casoni and Weinberg tests, indirect haemagglutination, eosinophilia and ELISA are used in diagnosing hydatid cysts, but as brain tissue evokes minimal response many results tend to be false negatives[2, 3]. ELISA is more specific, but less sensitive than most imaging modalities and has up to 84 percent sensitivity.

Nur Altinors *et al*[16] reported that out of 330 patients with hydatidosis who underwent operation, 207 cysts were executed successfully. In 86 (25.6%) the cysts ruptured inadvertently during surgical manipulation. In 35 cases (10.42%), the cyst was punctured, its contents were aspirated, the cystic cavity irrigated with a hypertonic saline and the cyst shrunk. This method is known as PAIR .Recurrence rate was reported in 56 (16.6%) of patients. A variety of post operative complications are reported in 32 patients ((9.52%). There were 34 deaths in patients with intracranial hydatid cysts, accounting for the mortality rate of 10.12% (34 out of 336). The surgical mortality rate was 8.48 %(28 of 330)

The aim of surgery is to excise the cyst without rupture to prevent recurrence and anaphylactic reaction. The pericystic hydraulic method (Dowling-Orlando technique) gives better results in removing these intact, in which normal saline irrigation is used with mild force between the cyst wall and brain interface in order to deliver the cyst intact[17].It is important not to rupture the cyst, even minimal spillage can cause anaphylactic shock and recurrence.1 ml of hydatid sand contains 4 00,000 of scolices. Entire operative field should be irrigated with 10% hypertonic saline which is scolicidal. Antihelminthic therapy is beneficial and indicated in patients with spillage. Albendazole is a broad spectrum antihelmenthic drug, daily dose of 10 mg/kg taken three times a day for four months. Albendazole 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[18].Adverse effects of albendazole include hepatotoxicity, severe leucopoenia, alopecia, embryo toxicity and teratogenicity. Erashin *et al* reported better effectiveness of the drug therapy in recurrent cases and cases with rupture at surgery[13].

Hydatidosis of spine was first described by Chaussier in 1807. Infection of the CNS is unusual and only between 2-4% of patients with hydatid disease present brain cysts. Bone involvement occurs in only 1% of the patients, and approximately half of them show spinal involvement. Primary vertebral disease without any other systemic involvement can occur with direct portovertebral shunt[19]. Any segment of the spine can be affected including the thoracic (50%), lumbar (20%), sacral (20%) and cervical (10%) regions[20]. These segments can be affected by an isolated or by multiple cysts infestation may occur at any location primarily or by metastatic seedling of protoscolices released from another location. The clinical presentations of spinal hydatidosis is variable ,including paraparesis (62%) or paraplegia(26%), back pain or radicular pain(55%) ,numbness or sensitivity loss (36%) and bladder and bowel disturbance(30%).Morphologically spinal involvement has been classified by Braithwaite and Lees[21] into five types;1)primary intrmedullary hydatid cysts ;2)intradural extramedullary hydatd cysts;3) extradural intraspinal hydatid cysts;4) vertebral hydatidosis; and 5)paravertebral hydatidosis. Some imaging characteristics are typical for spinal hydatid disease. These include lack of osteoporosis and sclerosis in the host bone ,absence of damage to the disc space and spread of the disease either via hematogenous route to the most vascular areas of the vertebra ,or via a subperiosteal and subligamentous path to the paraspinal region that results in involving a contiguous rib and destructing the adjacent costo-chondral junction. CT allows a precise assessment of osseous lesions and can clearly depict hydatid cysts or the microvesicular polycystic vertebra. MR findings are characteristic, showing the cysts which are usually alveolar and /or a diffuse form typically compressing the spinal cord posteriorly or posterolaterally. MRI is the preferred imaging modality when spinal hydatid disease is suspected and characteristically shows a lesion resembling a bunch of grapes. This unique feature can help in distinguishing hydatid infestation from spinal tuberculosis. The cyst walls are thin and regular without septations. The presence of a markedly hypointense cyst wall on T1- and T2-weighted images and the absence of wall enhancement with gadolinium are also characteristic of hydatid disease. Pure spinal hydatid cysts can be extradural, subdural, subarachnoid or intramedullary. Nur Altinors *et al*[19, 22] reported intraoperative rupture of spinal hydatid cysts in 44% of cases. Operative procedures are the treatment for the spinal cord compression caused by hydatid cysts. Laminectomy with simple decompression is the most frequent procedure[23, 24].

Orbital hydatidosis occur in 2-3% of cases CNS involvement, but hydatid manifestation of orbit comprises < 1%. Orbital hydatid cysts are solitary in majority of cases. But there are reports in the literature of multiple intraorbital cysts occurring in 55 of the patients with orbital hydatid cysts. From the literature, orbital hydatid cysts are situated in the superolateral and superomedial angle of the orbit, lying in or close to the muscle cone. The most common symptoms in orbital hydatidosis are slowly progressive

unilateral proptosis with or without pain, diplopia, headache, visual deterioration and peri orbital pain. Orbital MRI is proved to be the preferred diagnostic tool to rule out other lesion and understanding of the microanatomy of the orbit and proper surgical approaches are very important in preventing surgical complications .During intra orbital surgical hydatid cysts excision cyst rupture is common and may result in severe anaphylactic reaction, incomplete removal and secondary implantation.

Human hydatid disease though preventable, continues to be endemic in sheep- raising countries worldwide including India and China. The best strategy of any public health policy should be prevention and control of the disease. Killing of the stray dogs humanely and reducing the dog population, surveillance of dogs and periodic test of stools and treating them if infected and preventing from access to raw offal at slaughter houses and farms will be effective to control the menace of hydatidosis. Many countries such as Newzealand, Australia and China have been successful in this regard and India is yet to catch up[11, 25]

Conclusions

High index of suspicion is required despite the availability of advanced neuro-imaging. It is the benign lesion and the removal of the cyst without rupture is the most important to avoid spillage and recurrence of secondary multiple cysts. Of clinical importance is that a great liver hydatid cyst may be asymptomatic and discovered incidentally by sonography or CT scanning .The great majority of them will undergo spontaneous abortion and do not need any active intervention. This is not the case for the hydatid cysts of the CNS. Because of the unpredictable growth pattern of CNS cysts, until a proper medical therapy is discovered, they must be surgically removed at almost any stage they are diagnosed. Despite all the advances in imaging techniques and therapeutic methods ,CNS hydatidosis remains difficult to cure and the patient outcomes are not satisfactory especially in case of spinal involvement due to high incidence of recurrence.

9. Acknowledgements

I thank professor of neurosurgery Dr. W. Seetharam. M.Ch., Dr. Joji Reddy professor of radiology , Dr.G.Rama Prasad., M.D., Principal, Kurnool Medical College and Dr.J.Veera swamy .,M.Ch., Superintendent Government General Hospital Kurnool for their valuable suggestions.

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