Hydatid disease of the spine: a report on nine patients

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Received: 12 November 2011 / Accepted: 28 December 2011 / Published online: 21 January 2012
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Abstract

Purpose The author presents this prospective study of nine cases of pathologically confirmed spinal hydatid disease.

Method Hydatid disease is a difficult diagnosis in non endemic areas but it should be considered in the differential diagnosis of spinal pathology in endemic areas. Spinal involvement is very unusual. There is nothing typical of spinal involvement. Nine patients presented with hydatid disease of the spine between September 2001 and October 2010. The patients were clinically evaluated as well as by the latest imaging modalities, haematological and serological tests. All had decompressive surgery with or without fixation and the diagnosis was confirmed by histopathological examination. All received albendazole and praziquantel for ten months.

Results MRI was the best diagnostic test, CAT scan was also useful, eosinophilia was a constant finding, and ESR was above normal in five patients. All had decompression laminectomy and clearance; in addition, transpedicular fixation was done to three patients. After surgery one patient had complete recovery with no recurrence, seven patients showed recurrence over time and residual disease was observed, and one patient died within 24 hours of surgery.

Conclusion Diagnosis was easy from the start, but eradication was difficult, and recurrence rate was very high despite the use of chemotherapy.

Introduction

Hydatid is a Greek word meaning water cyst [17]. Hydatid disease is an infestation caused by the larval stage of the tape worm *Echinococcus granulosus*. Although a total of four different organisms can cause echinococcosis in humans, *Echinococcus granulosus*, which causes the cystic echinococcosis, is the usual causative organism.

Hydatid disease is very rare in Europe and America but it is a very significant problem in underdeveloped regions of the world where there is no veterinary control.

The first description of spinal hydatidosis was made by Churrier in 1807 [17, 19]. Hydatid cysts of the spine constitute 1% of all cases of hydatidosis and are most commonly located in the thoracic spine and less so in the lumbar, sacral and cervical regions [4]. Spinal hydatid cysts may occur by direct extension from pulmonary or pelvic foci or, rarely, begin primarily in the vertebral body [4]. Primary intradural extra medullary hydatid disease is extremely rare [1]. Approximately 90% of spinal hydatids are located extradurally, most commonly in the vertebral body [17]. The infection may then spread to the epidural space or the para vertebral soft tissue.

Parasites are considered to reach the highly vascularised centre of the vertebral body through portal vertebral venous shunts [11]. Intramedullary hydatid disease has also been reported [14]. Trauma may draw attention to the site of lesion. Spinal hydatidosis is predominately seen in adults, usually men of age 21–40 years [3]. The usual manifestation is due to pressure by the cyst on the surrounding tissue. There are no specific symptoms or signs, usually the presentation is related to radiculopathy, myelopathy, and/or local pain due to bony destruction, cord compression or pathological fracture.

Diagnosis is easy in endemic areas because of the clinical awareness. Clinically they present with features of spinal

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cord or cauda equine compression similar to any other compressive pathology but, because of the slow growth of the parasite, it may take several years before presenting clinically. Spinal hydatid cysts may grow to a very large size and clinically remain asymptomatic for several years [7]. Eosinophilia and the Casoni test may help in the diagnosis, and computed tomography may also help in making the diagnosis, but magnetic resonance imaging is the preferred imaging modality in the diagnosis of hydatid cyst, and recent use of diffusion-weighted MRI has been shown to help in the differential diagnosis [1]. The best treatment is radical surgical excision which is not easily achieved with spinal hydatidosis. Chemotherapy may help in reducing the recurrence which is very common.

The aim of this study was to describe the clinical features of these nine patients, with this peculiar, perplexing and rare spinal infection, and to evaluate the effect of chemotherapy.

Patients and method

This prospective study was performed over nine years. Nine patients with hydatid disease of the spine were diagnosed and treated.

All of the patients were investigated by routine hematological investigations including blood counts, plain radiographs of the whole spine in antero posterior and lateral views, chest radiograph, abdominal and pelvic ultrasound to rule out chest and visceral hydatid disease.

All patients had magnetic resonance imaging (MRI) and computerised tomography (CT) (Figs. 1, 2, 3, 4). The Casoni test was also performed on all patients.

Results

The series included six men and three women with an age range between 24 and 62 years (Table 1). All presented late for treatment, and the duration between the first symptom and the medical consultation varied between six and 11 months.

Fig. 1 T2 axial images revealing the typical multilobular, multicystic septated mass lesions (arrow) at D1-D2-D3 levels with posterior extradural location causing dural and cord compression and anterior deviation

Only one patient with a dumbbell hydatid cyst of the dorsal spine showed no bony involvement. The sites of the cysts included five in the dorsal region, two in the lumbo-sacral region, one in the lumbar and one in the lower cervical region. In three patients, another cyst was discovered in the liver; one patient had a cyst in the lung and one patient had multiple intramuscular cysts in the left thigh muscles. In only six patients was the cyst a primary cyst in the spine. The site was extradural with bony destruction in severe patients, while in two patients there was intra and extramedullary invasion. All presented with some degree of neurological deficit. One patient presented with quadripareisis because of the lower cervical location, while eight patients presented with paraparesis.

Those with cervical and dorsal location presented with features of upper motor neuron lesions while those with lumbar and lumbo sacral lesion presented with lower motor neuron lesions. Sphincter control was lost in five patients at the time of initial presentation. Only the patient with a dumbbell hydatid cyst in the dorsal region showed complete recovery with no recurrence.

Eight patients showed some degree of recovery of neurological deficit within a month of surgery but later showed gradual deterioration and six of the eight patients needed re-exploration. Histopathological confirmation of the diagnosis was obtained.

One patient with an L4 lesion and a mass adherent to the abdominal aorta developed intra arterial dissemination which led to severe anaphylactic reaction and death within 24 hours of surgery. One patient with lumbo sacral involvement ended up with multiple discharging sinuses in the gluteal region.

Laboratory data

All showed some degree of eosinophilia. The erythrocyte sedimentation rate was higher than normal in five patients (70–80 mm/hour) because of associated bacterial infection.

The Casoni test was positive in seven patients only. In cases with bony involvement plain radiographs

Fig. 2 MRI T2 axial and 2D myelogram images showing a multilocular septated cystic mass (*arrow*) within the cervico-dorsal right paraspinal muscle clearly shown posterolateral to the thecal sac and spinal canal

showed multiple well-defined, osteolytic lesions without periosteal reaction or sclerosis. Plain radiography of the chest showed hydatid cysts in the right lung. Ultrasonography of the abdomen showed involvement of the liver with hydatid cysts in three cases.

All MRI were highly suggestive of hydatid cyst, with typical findings of a lobulated, multilocular, septated cystic mass, connected or separated, containing fluid; and the returned signal varies depending on whether the cyst is complicated or if infection changes the picture. The signal is bright hyperintense in T1 and clear hypointense in T2.

CT scan was also very helpful in demonstrating the multiple cysts, as osteolytic expansile lesions in vertebral bodies.

Surgical intervention

All patients underwent surgery to excise the cyst and to decompress the cord or the cauda equina, or to fix the spine. In all patients the operative findings were highly suggestive of hydatid disease; six had laminectomy performed through the posterior approach for neurological decompression at the level of spinal involvement and transpedicular fixation was done to three patients.

Three had anterior decompression, one for the removal of dumbbell hydatid cyst in the dorsal region, one for anterior decompression in the dorsal region and one for anterior decompression at the level of L4 where a large mass was discovered eroding the abdominal aorta, which was separated with difficulty. The last patient had dissemination of a

Fig. 3 Coronal and sagittal T2 images, showing, multilocular, lobulated intramuscular extradural right sided paraspinal hydatid cyst (*arrow*) at the cervico-dorsal region (C7, D1-D2)

Fig. 4 Sagittal T1 images showing massive bone destruction of the lumbar vertebrae multiple multilocular hydatid cysts (arrow) and soft tissue swelling and angular kyphosis

daughter cyst in both femoral arteries leading to pulseless limbs and death within 24 hours of surgery due to severe anaphylactic reaction.

Due care was taken to prevent rupture while removing the cyst (Figs. 5 and 6), which contains hundreds of protoscolices, which can form a new hydatid cyst; four cysts were found already ruptured. Scolicidal solution was not used because of the proximity of the neural tissue.

Discussion

What made the diagnosis easy at presentation was the author's awareness of this condition because it is a locally

endemic disease. But sadly all patients presented very late for treatment because of low educational standards and poor socio-economic status; this late presentation makes achieving cure almost impossible, and it is the reason behind the high recurrence rate in the cases studied.

The growth of hydatid cysts is very slow; taking several years to show themselves, so probably the lesions start in childhood.

The author agrees with Benzagmout et al. [2] that the Casoni test is useful for the diagnosis of hydatid disease but a negative result does not rule out the diagnosis. In the cases studied two patients showed a negative Casoni test which was probably related to an associated bacterial infection. The erythrocyte sedimentation rate was

Table 1 Presenting symptoms and neurological signs of the patients with spinal hydatid disease

Characteristic	Case								
	1	2	3	4	5	6	7	8	9
Sex	M	M	F	M	F	M	M	M	F
Age (years)	48	62	35	40	26	24	55	60	28
Duration of symptoms(months)	9	6	11	6	5	7	9	10	8
Back pain	+	+	+	+	+	–	+	+	+
Radicular pain	+	+	+	–	+	–	+	+	–
Paraparesis or quadripareses	+	+	+	+	+	Quadriparesis (6)	+	+	+
Urinary hesitancy or incontinence	–	+	+	+	–	+	+	–	–
Systemic hydatid disease	No	Liver	No	No	Lung	No	No	Muscle	No
Level of spinal lesion	D1-D2	D2-D3	D6	D7-D8	D7-D8	C6-C7	L4-L5	S1-S2	L5-S1
Recurrence	+	+	+	+	+	–	+	+	+
Complications	–	–	–	+	–	+	Death	+	+
Secondary infection	+		+	+		+		+	

M male, F female

Fig. 5 Multiple discharging sinuses in the gluteal and upper thigh

high in five patients; this is also related to associated bacterial infection.

Magnetic resonance imaging (MRI) was confirmed to be very useful in the diagnosis of hydatid disease [1, 2, 8, 15, 20]. MRI can be used to precisely locate the site and even to show any daughter cysts.

All patients studied had an MRI of the whole spine because the author feels that the spine is one bone, though multiplicity of spinal involvement was not found in the cases studied. Güneş et al. [9] reported multiple distinct spinal intradural extra medullary hydatid cyst. CAT scan is also useful for analysing the bony involvement. Hydatid cysts were discovered in the liver, lung and muscle. No tissue in the body is immune to hydatid disease involvement, making whole body screening mandatory. Tuberculosis of the spine should be considered in the differential diagnosis, a fact also mentioned by Tabak et al. [19].

Recurrence was faced in eight out of nine patients and was related to many factors, including late presentation, as all presented several months after the initial symptoms. This is related to the lack of awareness of both the patient and the local physicians. Another cause for the high recurrence rate is the massive bony and neural tissue damage which makes wide local resection with safety margin practically impossible. All patients presented with some degree of neurological deficit.

Four patients presented initially with ruptured cyst and intra osseous dissemination, which makes excision impossible. Local scolical was not applied, fearing damage of the nearly neural tissue; this accords with Probhakar et al. [18].

Formalin irrigation led to one death in the cases reported by İşlekel et al. [10], because a dural tear allowed intradural penetration of formalin. Although those authors believe that formalin (10%) can be used for irrigation if the dura is

Fig. 6 Huge number of hydatid cysts coming out of sinuses after pressure

intact, my belief is that it is very unsafe to use formalin for irrigation. Duran et al. [5] used ethacridine lactate [Rivanol] in a concentration of one milligram per milliliter of water for irrigation to clean the operative field and in the post-operative period through a tube inserted in the lesion for several days. Probably the safest solution is 20% hypertonic saline as mentioned by Güneş et al. [9].

In the cases studied neurological deficit was the modus of presentation. Acute bleeding as a rare presentation was documented by Wang [21].

Anaphylactic reaction following rupture of the cyst was mentioned by Gopal et al. [6]. Of the cases studied one patient developed severe anaphylactic reaction leading to death due to intra arterial dissemination of the small daughter cyst and the fluid.

Albendazole as a chemotherapeutic agent was used for several months to one year by Kotil et al. [12]. Lam et al. [13] add praziquantel to albendazole, leading to better control and a reduction of recurrence rate.

Recurrence was not prevented in the cases studied although both albendazole and praziquantel were; this fact was also supported by Probhaker et al. [18].

I consider chemotherapeutic agents to be useful for the soft tissue involvement; so far, to my knowledge there is no study concerning the concentration of albendazole and praziquantel in the bony or neural tissue and the overall efficacy of these drugs requires a further period of observation.

Duran et al. [5] emphasise the value of immunological measures (hydatid antigen desensitisation) in the treatment complementing the surgical procedures used. So far this line of treatment is not widely used.

Surgery on hydatid cysts of the spine is very difficult, in particular when they present very late. Clearance of the site of pathology was not achieved because both neural and bony tissue involvement were present at the same time.

Only one patient without bony involvement showed no recurrence, probably indicating that sparing the bony tissue may carry the best outcome.

The author agrees with Ozdemir et al. [16] who believes that a lasting solution is hard to achieve in primary hydatid disease of the spine.

Pamir et al. [17] consider spinal hydatidosis as an unsolved problem, and the author supports the feeling of İşlekel et al. [10] that this benign disease has a malignant course because of its high rate of recurrence.

Conclusion

Spinal hydatid disease is rare, but a high index of suspicion in endemic areas and should be considered in the differential diagnosis of spinal infection. Spinal hydatid disease will remain an unsolved problem in patients who present late

because surgical extirpation is practically impossible. More studies are required to confirm the concentration of the chemotherapeutic agent required in the bony and neural tissue.

Conflict of interest The author declares no conflict of interest.

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