Musculoskeletal hydatid disease

A report of 13 cases

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ABSTRACT – This is a retrospective study of 13 patients with muscular hydatidosis—i.e., 4% of the 309 cases of hydatid disease treated in our department during 1983–1999. The commonest clinical finding was an asymptomatic and slowly growing mass (7). Puncture or incision of the mass was followed by an infection of the cystic cavity with fistulization in 2 patients. The immunological findings were false negative in 4 patients. MR images were obtained in 4 patients before diagnosis, and were highly suggetive of hydatid disease. The cystic cavities in all 9 patients subjected to radical surgery healed without chemotherapy. Radical surgery was not possible in 4 cases, in 3 of whom the sacrum was involved. Medical treatment of these patients did not eliminate the disease and new operations were necessary.

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Hydatid disease is the commonest disease in humans caused by helminths, but primary skeletal and/or muscular involvement seldom occurs (Briant et al. 1998). It is a zoonotic infection involving larval forms of small tapeworms of Echinococcus granulosus. This parasite inhabits the small intestine of carnivores, such as dogs and wolves, definitive hosts in the cycle (Piédrola-Angulo 1987, Merkle et al. 1997). Humans become infested by ingesting ova from the feces of these carnivorous animals (Piédrola-Angulo 1987). Because of their role as capillary filter stations, the liver and lung are primarily affected in this disease (Piédrola-Angulo 1987, Essadki et al. 1996, Merkle et al. 1997). Musculoskeletal involvement is seen in only 1–4% of cases (Merkle et al. 1997). This disease is ubiquitously distributed with more frequent occurrence in the Mediterranean countries, East Africa, South America, Russia and Australia (Piédrola-Angulo 1987, Martin et al. 1993, Merkle et al. 1997). However, immigration will probably result in the appearance of this condition in unexpected countries (Eckert et al. 1995).

We present a series of 13 cases of muskuloskeletal hydatid disease, and evaluate diagnostic tests and treatment.

Patients and methods

13 patients (8 men) with muscular hydatidosis were diagnosed and treated between 1983 and 1999—i.e., 4% of the 309 cases of hydatid disease treated in our department during this period. Their mean age was 53 (30–82) years (Table), and mean follow-up 6 (1–17) years. Patient no. 4 has been reported elsewhere (García-Alvarez et al. 1999). In 3 patients, bone was affected (1 sacroiliac, 2 sacrums). Involvement of other organs, such as the liver and lung, were ruled out with sonography and chest radiography, respectively. Concomitant hepatic hydatidosis was seen in 1 patient and a reccurence of pulmonary hydatidosis in 1 patient (operated on 17 years previously). The most frequent locations were paravertebral, pelvic-gluteus and the lower extremities.

The serologic test for hydatidosis (indirect hemagglutination test), immunoglobulins G, A, M, E and specific IgE Echinococcus granulosus by ELISA using monoclonal antibodies, were studied

Patients' data

A	B	C	D	E	F	G	H	I	J	K	L	M	N
1	57 m	quadrate muscle left lumbar muscles	–	tumor compr.	–	high [a]	–	CT ++	–	–	–	multi	76
2	46 m	sacrum bone left lumbar muscles	–	tumor sciatica	–	normal	–	CT ++	+	–	+	multi	84
3	52 f	right para-vertebral	–	tumor no pain	–	normal	+	CT +	–	+[b]	pr	uni	46
4[c]	45 m	lumbar muscles longissimus muscle	–	tumor	+	normal	–	CT/MRI ++	–	–	pr	multi	72
5	82 f	abdominal wall muscles	liver [d]	pain tumor	no	no	–	CT –	–	+	–	multi	12
6	69 m	diaphragm left column	lung	vomiting	+	high	–	CT ++	–	–	–	uni	72
7	52 m	right gluteus	–	tumor	–	normal	–	MRI ++	–	–	–	multi	30
8	53 f	sacroiliac bone gluteus	–	fistula	+	normal	–	CT +	++	+	++	multi	79
9	43 m	left gluteus	–	tumor	+	high	–	Echography +	–	–	–	multi	50
10	65 f	right thigh medial vastus	–	tumor fever	+	high [a]	+	Echography/ CT/MRI +	+	+	++	multi	98
11	40 f	left lateral gastrocnemius	–	fever pain	+	high	–	MRI ++	–	–	–	multi	29
12	66 m	right soleus muscle	–	tumor heavy legs	–	normal	–	CT/MRI ++	–	–	–	multi	44
13	30 m	sacrum bone gluteus	–	fistula	+	normal	–	CT/MRI ...	++	+	++	multi	205

[c] Case reported elsewhere (García-Alvarez et al. 1999)

A Patient
B Age, gender
C Localization
D Liver/lung, [d] not operated on
E Symptoms
F Serology
G Total IgE, [a] specific IgE
H Puncture/incision
I Image, methods
  ++ highly suggestive
  + suggestive
  – poorly suggestive
  ... after diagnosis
J Adjuvant treatment
  ++ several
  + yes
  – no
K Infection, [b] septic shock
  + yes
  – no
L 2nd operation
  ++ several
  + yes
  – no
  pr previous
M Uni- or multiocular
N Follow-up, months

in all cases, except in case 5, preoperatively and every 3 months during the first postoperative year and then every year.

Excision with a 3.5-cm safety margin was performed, followed washing with a 20% saline solution and then a 50% povidone solution. The cysts were surrounded by a thick fibrous capsule, the pericyst, and, on incising the cystic cavities outside the operating room, they were found to contain clear fluid and hydatid vesicles with scolices. Microscopic study confirmed the diagnosis of hydatid disease.

In all cases in whom we could not perform radical surgery because of sacral involvement (2, 8, 13) or the proximity of large nerves (10), adjuvant treatment was given. Albendazole 15 mg/kg/day (cases 2 two cycles, 8 six cycles, 10 six cycles, 13 three cycles) or mebendazole 50 mg/kg/day (before albendazole, cases 2 three cycles, 10 for 3 years) was given in cycles of 28 days and 1 month without treatment after every three cycles. The patients were monitored for hepatic toxicity.

Results (Table)

The commonest clinical presentation was an asymptomatic slow growing mass (7 patients). 2 patients

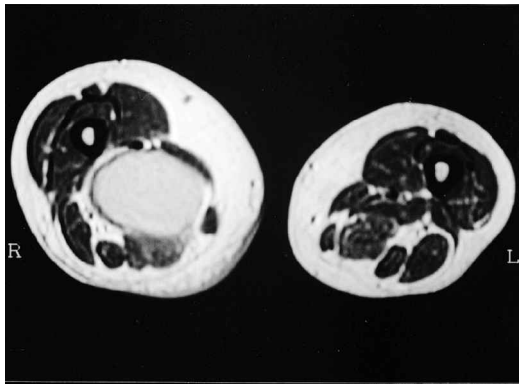
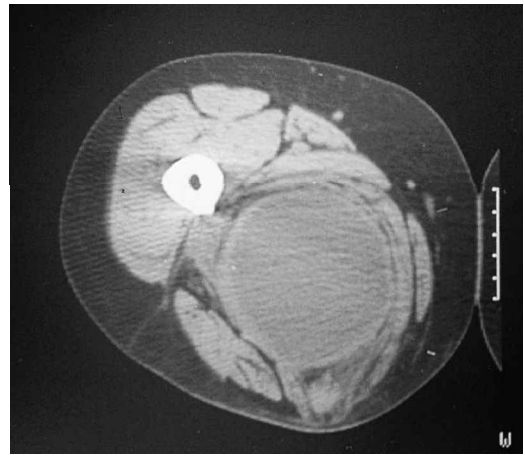


Figure 1. 65-year-old woman (case 10) with a tumor in the medial side of the right thigh.
A. On this axial T2-weighted MR image, the mass has a high signal intensity. The lateral part of the cyst shows the "rim sign"—i.e., a low signal intensity surrounding the cyst and representing the pericyst.



B. CT findings: tumor, 8 cm in diameter, with homogeneous filling and a thin wall of low intensity.

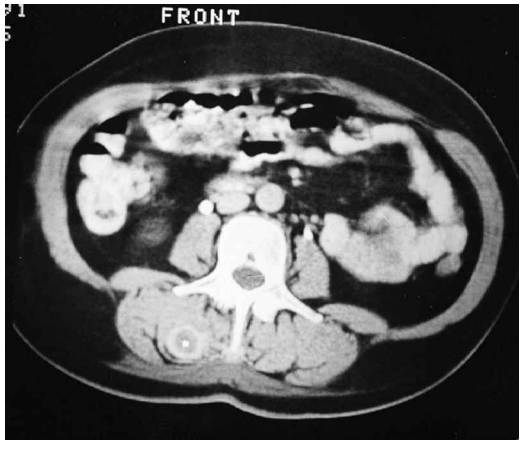


Figure 2. CT image showing a tumor with a high intensity ring in the periphery and a high intensity center in the paravertebral muscles of a 45-year-old man (case 4).

in whom puncture or incision of the mass was done for diagnostic purposes or an incorrect diagnosis had an infection in the cystic cavity with fistulization. 2 patients in whom the disease involved the sacrum (cases 8 and 13) presented with a fistula and pus on admission. 1 patient, who had been operated on because of an erroneous diagnosis of lipoma in another center 1 week before, was in septic shock. In a patient with disease involving the abdominal muscles, a purulent mass was found around the hydatid cyst. However, radical surgery with a good safety margin was performed.

In 5 patients, the indirect hemagglutination test was negative. IgE and specific IgE were negative in 7 patients (Table). The indirect hemagglutination test and ELISA were not decisive in the diagnosis. In case 6, total IgE decreased from 3290 UI/mL at the time of radical surgery, to 361 UI/mL 1 year later. However, specific IgE increased from 9.3 ku/L to 19 ku/L with no new clinical or radiological signs of hydatidic infection in the 5 years after surgery. Apart from 2 patients (6: 2480 eosinophils/mL; 11: 3540 eosinophils/mL) eosinophil counts were normal in all patients.

MR images, taken in 4 patients before diagnosis (4, 7, 11 and 12), and CT in 5/9 patients were very suggestive of hydatid disease.

The cystic cavities in all 9 patients who underwent radical surgery healed without postoperative complications. These patients were not given chemotherapy. Radical surgery could not be performed in 4 cases, in whom treatment with albendazole or mebendazole treatment did not cure the disease and new surgical procedures were necessary without definitive healing. In case 8, treatment with albendazole coincided with the closure of 2 fistulae. However, 4 years later, surgery was necessary. In case 10, the patient received mebendazole for 3 years after surgery. When treatment was interrupted, the symptoms recurred and surgery was necessary again. In patient 13, chemotherapy may have provided 9 asymptomatic years. In no case was it necessary to stop albendazole or mebendazole treatment due to hepatic toxicity.

Discussion

Like others (Essadki et al. 1996, Merkle et al. 1997), we found that musculoskeletal lesions of cystic echinococcosis usually occurred as isolated findings and without concomitant hepatic or pulmonary involvement. Nevertheless, the involvement of other organs should be ruled out. Most cases of muscular hydatid disease have been associated with skeletal lesions (Merkle et al. 1997), but this was not true of our series. Daali and Hssaida (2000) reported 15 patients with muscular hydatidosis. However, the diaphragm or psoas muscle was affected in most of them, and this entailed different criteria for diagnosis and treatment.

Several patterns of disease have been recognized using various imaging techniques. These include the unilocular cyst, the multilocular lesion and the atypical complex or solid lesion (Martin et al. 1993, Essadki et al. 1996). The multilocular lesion with several daughter cysts inside the mother cyst is characteristic, but not pathognomonic of hydatid disease (Martin et al. 1993, Essadki et al. 1996). Endovesicular daughter cysts, present in most of our cases, are regarded as unusual in musculoskeletal cystic echinococcosis by some authors (Von Sinner et al. 1995, Guthrie et al. 1996, Merkle et al. 1997). MR images were obtained in 4 of our patients before diagnosis, and were highly suggestive of hydatid disease. The MRI signal intensity pattern of the daughter cysts reflects their contents and may vary in cysts that are dead or alive. The production of hydatid fluid stops when they disintegrate at death (Martin et al. 1993, Merkle et al. 1997). The presence of a bacterial infection or abundant intracystic debris and inflammatory changes may affect the typical cystic morphology, transforming it into a complex or solid lesion mimicking a tumor (Martin et al. 1993). A T1-weighted low intensity signal pattern may appear in daughter cysts (Guthrie et al. 1996). The "rim sign" on the MR images appears as a low signal intensity surrounding the cyst and it may help to distinguish hydatid cysts from other lesions (García-Alvarez et al. 1999). However, this sign is not specific for hydatid disease and may be seen in other lesions that have a fibrous capsule (Martin et al. 1993, Guthrie et al. 1996). CT image findings are similar to MR T1-weighted ones.

Immunologic tests are useful in the diagnosis of hepatic hydatidosis (Navarro et al. 1983, Torcal et al. 1996), and infected hepatic hydatidosis (Salinas et al. 2000). However, in other reports (Martin et al. 1993, Essadki et al. 1996), as in our series, serological tests and the eosinophil count were not positive for muscular echinococcosis in all cases, therefore only a positive test is helpful. However, seronegativity may develop because unknown host and/or parasite factors cause inadequate T-helper-2 cell activation, thus reducing production of T-helper-2 cytokines implicated in immunoglobulin expression in cystic echinococcosis (Rigano et al. 1998). Bonifacino et al. (2000) found increased circulating antigen and specific antibodies, especially total specific immunoglobulin, of prognostic value in some patients with severe bone or secondary disseminated echinococcosis where the levels remained high and/or increased. However, while the level of total IgE decreased in our patient no. 6 one year after radical surgery, as described in hepatic hydatidosis (Navarro et al. 1983, Torcal et al. 1996), that of specific IgE, which remains increased in hepatic hydatidosis for the rest of the patient's life (Torcal et al. 1996), increased with no new clinical or radiological signs of hydatidic infection in the 5 years after surgery. In chronic cases (10, 13), IgE did not increase during the follow-up. This response in our series may have been due to albendazole/mebendazole treatment. Indeed, Bonifacino et al. (2000) found that antigen-specific in vitro lymphocyte transformation assays show a significantly lower response than controls during albendazole treatment, but return to normal levels in healed patients due to parasite-induced suppression of cellular responses.

Adjuvant administration of benzimidazole derivatives preoperatively and for about 3 months postoperatively is advocated by some authors (Guthrie et al. 1996). Like others (Martin et al. 1993), we consider surgery with a broad safety margin is the best treatment. The cysts in our series that were not subjected to radical surgery (3 of them with involvement of bone) failed to heal and had to be operated on again regardless of chemotherapy. It is difficult to perform a radical resection on the sacrum (Durán et al. 1978). The way echinococcosis spreads in bone has not yet been fully deter-

mined (Sapkas et al. 1998), therefore it is very hard to establish safety margins in osseous hydatidosis. Nevertheless, we think chemotherapy may have helped to close fistulae and delay symptoms in our patients with skeletal involvement.

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