Hydatid disease of bones and joints

8 cases followed for 4–16 years

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Hydatid disease is a rare parasitic disease that seldom involves the skeleton. Treatment is difficult because of problems with the preoperative diagnosis, the invasive nature of the bony involvement and the variable anaphylactic reaction to the cyst fluid antigen. We present 8 cases with osseous hydatidosis who were treated over a period of 11 years. The spine was involved in 2 cases, the ilium in 2, the hip in 2, the tibia in 1 and the humerus in 1. We point out that diagnosis is difficult and the prognosis is often poor.

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Submitted 97-03-01. Accepted 97-07-22

Echinococcus infestation is endemic in the Mediterranean, the Middle East, Central Asia, East Africa and some areas of South America and northern Canada. Because of frequent overseas traveling, it may today appear anywhere in the world (Rao et al. 1991). Hydatid disease from echinococcus infestation is a rare parasitic disease occurring mainly in the liver or the lungs (Agarwal et al. 1992). Echinococcosis involves the skeleton in approximately 1% of the cases (Dorn et al. 1984, Porat et al. 1984, Rong and Nie 1985, Ocete et al. 1986, Charles et al. 1988, Rao et al. 1991).

Bone echinococcosis occurs in adults. Growth is slow and the way it spreads in the bone has not yet been fully determined. The prognosis generally is poor; the disease is difficult to eradicate (Rao et al. 1991). Anaphylactic reactions to the cyst fluid antigen may lead to shock. The efficacy of desensitization and intraoperative scolicides is low. Anthelmintic chemotherapy, with drugs such as mebendazole or albendazole, has been used to treat surgically unfit patients or to reduce the risk of recurrence after intraabdominal spillage. The drugs do not cure bone echinococcosis.

We present our experience in treating 8 patients with osseous echinococcosis.

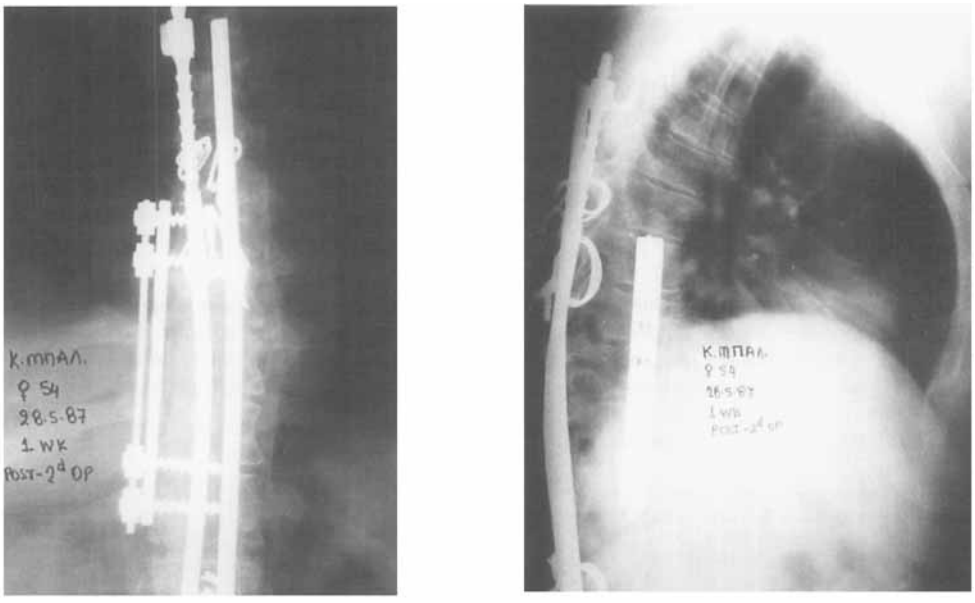
Patients and methods

We studied 8 patients (4 women) who were treated in the Orthopedic Department of the Medical School of Athens University during 1980–1996. Their mean age was 57 (44–68) years. The infected parts were the spine in 2 patients, the ilium in 2, the hip in 2, the tibia in 1 and the humerus in 1. The patients were followed for mean 11 (4–16) years.

Case 1 (Figures)

A 54-year-old woman was admitted to our department with acute paraplegia graded as Frankel B, neurologic deficit down to T9 level and no anal sphincter function. 3 years earlier, the patient had had similar symptoms, for which she had undergone T9–T10 laminectomies in another hospital. At that time, no diagnosis was made and later on the patient was considered fully recovered.

Case 1. Myelography. Block at the T9-T10 level due to intraspinal extradural hydatid cyst. It is not a specific finding.



Case 1. Anterior and posterior stabilization of the spine following extensive posterior decompression and anterior vertebrectomy.

On our examination, laboratory findings were not specific. Casoni intradermal and Weinberg tests were negative. Plain radiographs and CT showed cystic osteolysis involving the T9 and T10 vertebrae. 2 cysts were seen in the spinal canal, compressing the spinal cord anterolaterally on both sides. At that level, Tc-99 bone scan was positive. Myelography revealed a complete block at T9–T10 level.

During the operation the patient first underwent an extension of the old laminectomies through a posterior approach. The cysts were found anterolaterally to the spinal cord, causing extensive spinal canal encroachment. It was at this point that the diagnosis of spinal echinococcosis was established. The cysts were linked with the thoracic cavity through the intervertebral foraminae. Through this approach, only one of the cysts could be excised. The spine was stabilized with Harrington rods, in combination with the Luque system and allografts from frozen femoral heads. 2 weeks later, an anterior approach was made through the thoracotomy and the remaining cyst was excised, along with the T9 vertebral body. The gap was bridged with a massive pelvic bone graft and the spine was stabilized with the VDS system.

Postoperatively, the patient was treated with mebendazole for 3 months. At 5 months postoperatively, she was pain-free and showed a noteworthy neurologic improvement to Frankel C. However, 1 year later she developed complete paraplegia due to recurrence of the disease. She refused further operations or treatment and died 2 years later due to its progression.

Case 2

A 57-year-old woman was admitted to our department with paraplegia Frankel C, and neurologic deficit below the T4 level. The symptoms had started 1 year earlier. The laboratory findings were nonspecific. Casoni intradermal and Weinberg tests were negative. Plain radiographs and CT showed cystic osteolysis involving the posterior arch of the T4 vertebra, but the vertebral body was intact.

The patient underwent T4 laminectomy through a posterior approach. Postoperatively, she was treated with mebendazole for 3 months. 6 months after the operation, she was pain-free and showed much neurologic improvement to Frankel D. 10 years later, she was still asymptomatic.

Case 3 (Figures)

A 58-year-old man was admitted to our department with productive sinuses in the right hip. 1 year before, he had been operated on for echinococcosis of the liver. Casoni intradermal and Weinberg tests were negative. Plain radiographs and CT showed hydatid cysts in the lungs and a cystic osteolysis in the right ilium.

He underwent an operation for echinococcosis of the lung and surgical drainage for echinococcosis of the ilium. Postoperatively, he was treated with mebendazole for 3 months. 1 year later a productive sinus was present in the ilium, but the radiographs were negative for hydatid lesions. He was treated with mebendazole for another 3 months. 2 years later, new plain radiographs and CT scan showed reappearance

Case 3. Lytic lesions of the right ilium due to echinococcus infection.

Case 3. CT. Lytic lesions of the right ilium and sacroiliac joint due to echinococcosis.

of the cystic lesions in the right ilium and also in the right sacral bone. He underwent another surgical drainage, followed by a 3-month mebendazole treatment. 9 years later, hydatid cysts reappeared in the hip muscles. The patient again had surgical drainage and postoperatively was treated with mebendazole. 1 year later he was free of symptoms.

Case 4

A 68-year-old man was admitted to our department suffering for 10 years with productive sinuses of the right humerus. Casoni intradermal and Weinberg tests were negative. Plain radiographs showed total destruction of the upper part of the right humerus due to echinococcosis.

He underwent several surgical drainages and postoperatively was treated with mebendazole. Unfortunately, the results were disappointing with total osteolysis of the humerus head and the upper third of the shaft, with productive sinuses.

Case 5

A 53-year-old man was admitted to our department after suffering for 9 years from pain in the left lower leg. The patient had undergone an operation in another institution 5 years earlier. The lesion was excised and the diagnosis was tuberculosis. Postoperatively, he was given antituberculosis medication. The symptoms reappeared 2 years later. Casoni intradermal and Weinberg tests were negative. Plain radiographs showed a cystic lesion involving the left tibia shaft. During 5 years, he had 2 surgical drainages, followed by mebendazole treatment and today, 3 years later, he is well.

Case 6 (Figure)

A 44-year-old man was admitted to our department with right hip pain, serious walking difficulties and productive sinuses around the right hip. Casoni intradermal and Weinberg tests were negative. Plain radiographs and CT showed destruction of the right hip joint, with cystic lesions in the right ilium, the right sacral bone and the upper part of the right femur.

The patient refused the proposed amputation and finally underwent 3 surgical drainages and an excision of the upper part of the right femur. Postoperatively, he was treated with mebendazole. He continued to have productive sinuses 16 years later and now walks with the help of a walking-stick with difficulty.

Case 7 (Figure)

A 57-year-old woman was admitted to our department to undergo a total hip arthroplasty, due to congenital dislocation of her right hip. During the preoperative

Case 6. Extensive lytic lesions of the right hip due to echinococcus infection.

Patient data

No.	Age	Sex	Location	Main clinical findings	Operation	Follow-up years	Outcome
1	54	F	Spine	Paraplegia	Decompression, vertebrectomy, spinal stabilization	6.5	Recurrence Death
2	57	F	Spine	Paraplegia	Decompression surgical drainage	10	Symptom-free
3	58	M	Ilium,liver, lungs	Hip sinuses	Surgical drainage	10	Symptom-free
4	68	M	Humerus	Destruction of upper humerus, sinuses	Surgical drainage	16	Nonfunctioning arm, productive sinuses
5	53	M	Tibia	Pain in lower leg (tibia)	Surgical drainage, bone grafting	16	Symptom-free
6	44	M	Hip joint	Pain and sinuses	Surgical drainage	16	Hip pain and walking difficulty, sinuses
7	57	F	Ilium	Nonspecific Coexisting hip disease (CDH)	Surgical drainage	6	Symptom-free
8	63	F	Hip joint	Nonspecific coexisting RA	THR, surgical drainage	4	Death

Case 7. Plain tomography: A large echinococcal cyst in the right ilium.

control with plain radiographs, a cystic lesion in the right ilium was discovered. The laboratory findings were not specific. Casoni intradermal and Weinberg tests were negative. A Tc-99 bone scan was positive for that area. She underwent an open biopsy, followed by surgical drainage and mebendazole postoperative treatment, after histologically diagnosed echinococcosis. 6 years later, she has no signs of an infection.

Case 8

A 63-year-old woman was admitted to our department for reconstructive surgery of the hip, due to long-standing rheumatoid arthritis. The medical history was unremarkable, with the exception of rheumatoid arthritis.

At surgery, during osteotomy of the greater trochanter, 2 fluid-containing, white cysts were found under the attachment of the gluteus medius. As we did not understand the significance of the cysts at the time, we completed the Charnley hip arthroplasty. Histologically the cysts proved to be echinococcus disease. Casoni intradermal and Weinberg tests, done postoperatively, were positive. Mebendazole was administered to the patient but, by the seventh postoperative day, local swelling, increasing tenderness and redness appeared at the surgical wound. After removal of some skin sutures, a few milliliters of clear, watery fluid were released and during the next few weeks a sinus had developed. A sinogram showed a connection between the hip joint, through the muscles of the buttock, and an area of combined cystic and sclerotic bone on the posterior wall of the ilium.

She refused a suggested hemipelvectomy. During the next 2 years, her hip function deteriorated, with disabling pain. Hip sepsis and loosening of the prosthesis were obvious. The chronic infection caused a steady deterioration in the patient's health and 4 years after the operation she died of pneumonia.

Postoperatively, all patients were treated with mebendazole, 100 mg 3 times a day, increasing up to 600 mg 3 times a day by the seventh day, for 3 months.

Discussion

Hydatid disease is caused by a cestode, Ecchinococcus, that uses man as an intermediate host to complete its life cycle. The adult worms inhabit the small intestine of the dog. Ova are shed in its excreta. Human infestation occurs when these ova are accidentally ingested. Embryos liberated from the eggs in the intestine penetrate the mucosa and enter the portal blood stream. Bone involvement is rare (Charles et al. 1988)

because they must penetrate the liver and lung filters to enter the arterial circulation (Rao et al. 1991).

The cyst wall insinuates itself between the bone trabeculae and may erode through the cortex into surrounding tissue. Growth is slow because of the resistance of the bone (Rao et al. 1991). The clinical manifestations of bone hydatid disease may take 10–20 years to become obvious (Markakis et al. 1990). In the spine, cysts permeate and destroy the medullary cancellous bone of the vertebral body, later spreading to the pedicles, laminae, heads of ribs and adjacent soft tissue (Charles et al. 1988). Paraplegia or nerve root compression occurs in one fourth to four fifths of the patients with spine involvement (Porat et al. 1984). Pathologic fracture is common, because of weakening of the cortex (Porat et al. 1984, Booz 1993).

On plain radiographs, osteolysis can usually be seen, sometimes associated with bone expansion and thinning of the cortex. These signs are not specific (Bouras et al. 1984, Rong and Nie 1985), but large lesions with soft tissue calcifications are highly suggestive of echinococcosis (Torricelli et al. 1990, Booz 1993). Tomography, especially CT, has proved to be an effective and sensitive method for discovering the cysts, their spread in adjacent soft tissues and their location in other parts of the body. Moreover, CT is valuable in preoperative planning (Charles et al. 1988, Torricelli et al. 1990). Postoperatively, CT may be helpful for assessing residual hydatid cysts and/or for detecting recurrence at an earlier stage. MRI provides further information on the involvement of the spinal cord in spinal echinococcosis. Myelography yields no specific findings (Rong and Nie 1985).

The preoperative diagnosis is difficult and usually is determined only after operation or open biopsy, because there are no specific disease characteristics to distinguish it from the other commoner causes of bone lesions (Markakis et al. 1990, Torricelli et al. 1990). Surgical exploration with histologic examination can provide a definitive diagnosis (Torricelli et al. 1990, Rao et al. 1991). Fine-needle aspiration must be avoided, because of the danger of spread. Preoperative diagnosis is reached in half of the patients (Booz 1993). This was true in our cases. Although in 2 cases (cases 1 and 5) operations took place in other institutions, the diagnosis was missed in both. In 5 of our cases, the primary lesion was not found. The differential diagnosis includes tuberculosis, large foci of fibrous dysplasia, enchondroma, metastatic carcinoma, multiple myeloma and giant cell tumor (Bouras et al. 1984, Markakis et al. 1990).

Appropriate immunodiagnostic tests, such as the Casoni intradermal test and the Weinberg complement fixation test, are helpful, but they are of no value if only bone is involved in the absence of lung or liver involvement. However, eosinophilia may be found (Markakis et al. 1990).

The only definitive treatment when bone is involved is a complete resection of the involved area, with a wide healthy margin (Voutsinas et al. 1987). This is difficult, but incomplete removal results in recurrences (Ocete et al. 1986, Charles et al. 1988). Operative treatment of spinal echinococcosis involves the selection of an approach that allows adequate removal of the cyst and cyst contents without spillage, management of the remaining cavity and avoidance of reactions hypersensitivity to the hydatid antigen; leakage of the cyst fluid at surgery may lead to anaphylactic shock (Rao et al. 1991). Osteosynthesis can be improved by the use of acrylic cement, considering that osseous grafts can be invaded by hydatidosis extension or recurrence. Most authors recommend a combination of surgical resection (Ocete et al. 1986), bone grafting (Pintilie et al. 1966) and postoperative treatment with mebendazole or albendazole (Porat et al. 1984, Ocete et al. 1986, Charles et al. 1988, Rao et al. 1991). Missed diagnosis and therefore delayed treatment, as seen in 2 of our cases, may compromise the prognosis.

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