



## Long term follow-up of a patient with disseminated spinal hydatidosis

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A rare case of a 27-year-old male patient with disseminated sacral hydatidosis is presented. Because the diagnosis was missed initially, the patient underwent only partial resection of the tumour to obtain tissue for histology. The resection was followed by deep wound infection, and re-exploration had to be performed, thereby resecting the remaining cyst tissue and the S1-S3 vertebral bodies. Adjuvant anti-helminthic therapy was started postoperatively. Unfortunately, the hydatid cyst further progressed and could only be controlled with multiple decompression procedures and continuance of anti-helminthic therapy. We review the diagnosis, treatment and prognosis of this uncommon condition, which is a serious challenge for the spinal surgeon.

**Keywords** : hydatid cyst ; spinal hydatidosis ; sacrum ; echinococcosis.

### INTRODUCTION

Echinococcosis is a term used to describe infection with the adult or larval stages of tapeworms (cestodes) belonging to the genus *Echinococcus* (3, 8,12). With one of the echinococcus species, *Echinococcus Granulosus*, hydatid cysts may develop in internal organs (3,8,12). The lifecycle of this parasite is mainly domestic, involving dogs as definitive and sheep as intermediate hosts (3,8). The highest endemicity of the parasite therefore is seen in regions with extensive sheep farming, like the Mediterranean basin and parts of Great Britain (12).

However, the parasite has a worldwide distribution and seems currently to emerge in certain parts of China, Asia, Eastern Europe, and Israel (8). In Western Europe countries, such as Belgium and the Netherlands, the incidence has remained low and clinicians are not always aware of the disease. This may lead to a delay in diagnosis and suboptimal treatment, especially when rare manifestations of the disease are encountered (12). In the current article, we present the long-term follow-up of a patient with sacral echinococcosis.

### CASE REPORT

A 27 year-old-male, born in Turkey, was referred to our hospital in the spring of 1990 with progressive low back pain radiating to the lateral part of

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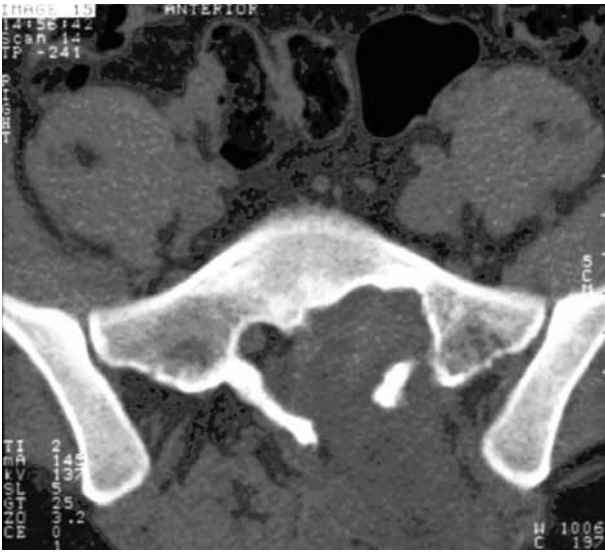
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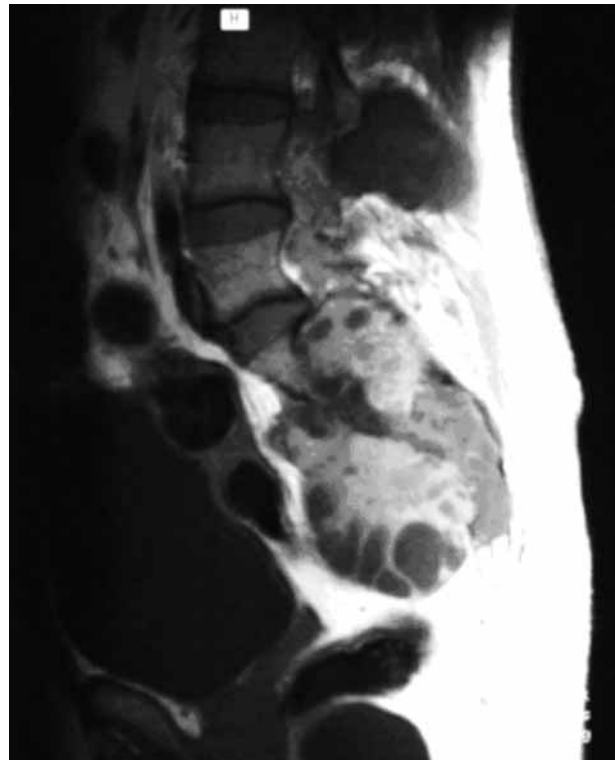
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*Fig. 1.* — Axial CT-image showing destruction of the second sacral vertebra.

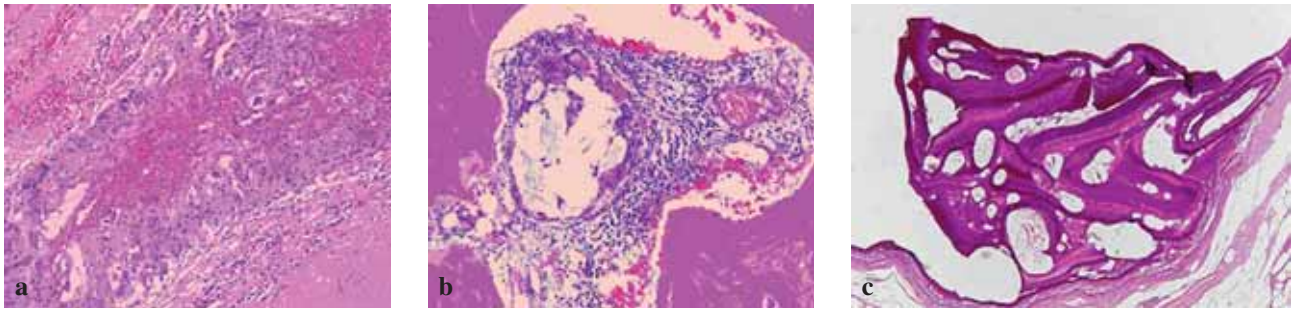
the left leg and toes. The pain had started 6 months previously. Walking distance had diminished to less than 100 meters. Coughing, laughing and sneezing precipitated the pain, and defecation had become problematic. There were no complaints regarding micturition or sexual functioning. Further history taking revealed no relevant information. On physical examination a healthy young man was seen with a painless swelling on the back, arising besides the midline of L4 on the left. The motion range of the lumbar spine was within normal limits. No abnormalities were found on digital rectal examination. Neurological examination revealed diminished sensibility on the left leg and left sacral-area (S3-S5) compared to the right. Reflexes and forces were normal and symmetrical on both legs. Straight leg raising tests were negative on both sides.

Blood samples were taken and revealed a slightly increased erythrocyte sedimentation rate (ESR) [14 mm/hr] and leucocyte count [10.0 million/ml]. Plain radiographs of the lumbosacral spine showed a large soft density shadow as well as a lytic type of sacral destruction. On Computed Tomography (CT) images multiple hypo densities in and around the sacrum and extensive intra-osseous destruction were seen (fig 1). There were no signs of calcific



*Fig. 2.* — Sagittal T1-weighted MRI image of the lumbar spine showing a large cystic lesion.

deposits in the mass. MRI investigation showed a mass composed of numerous round and oval shaped lesions highly variable in size (fig 2). Their low to intermediate T1 and high T2 signal intensities as well as faint rim enhancement after intravenous gadolinium were indicative of a cystic nature of the mass. The mass covered a cranio-caudal distance of 26 cm in the sacrum and left dorsal soft tissues of the back, reaching up from the level of the anus to the third vertebral body. Vertebral bodies as well as the intervertebral disc spaces were spared. Invasion of the mass into the sacral canal caused extradural compression and encroachment of both the dural sac and nerve roots. Ventrally, the tumour was seen to extend into the presacral space and left pelvis floor causing compression and displacement of the adjoining visceral structures. In the differential diagnosis degenerative cystic schwannoma, cystic extramedullary ependymoma, chordoma, were all



**Fig. 3.** — Histological images showing numerous clusters of giant cells and bone destruction (a, b). Other fragments contain only hyaline and possibly lectin-like, PAS-positive fragments probably derived from the echinococcus eggs (c).

preferred above hydatidosis. A partial resection of the tumour was performed and histologic evaluation strongly suggested an inflammatory lesion, most likely an echinococcal invasion (fig 3 a-c). ELISA IgG for *Echinococcus granulosus* turned out to be positive [1:80]. Additional immuno-electrophoresis against echinococcus and CBR were both positive. ELISA directed against *Echinococcus multilocularis* was negative. Meanwhile, the patient had developed progressive fever, malaise and local signs of a bacterial infection. Two surgical re-interventions were performed : the remaining part of the lesion, the sacral vertebral bodies and laminae of S1-S3 were resected. No reconstruction was performed. In addition, high dosed intravenous antibiotic therapy against *Staphylococcus Aureus* was initiated. After local infection control, the patient could be discharged continuing intake of high dose oral albendazole. CT scanning on discharge unfortunately showed a lesion suspicious for a remaining hydatid localisation in the intervertebral foramina of L4-L5. Treatment with albendazole was continued and the patient was seen on regular intervals at the outpatient clinic. The clinical picture, the infectious laboratory measures as well as the imaging studies remained steady. After ten years, however, he returned with a drop foot and progressive weakness and pain in the left leg. MRI studies showed an increase of the soft tissue lesion around the left SI-joint with extension into the left iliac bone. In addition, the lesion in the spinal canal at the level of L4 showed an increase in size as well. Another surgical decompression was performed, including

spondylolysis of L3-L5 in combination with resection of the laminae and left pedicle/corpus of L4. The echinococcus lesion was macroscopically resected and the bone defect filled with polymethylmethacrylate (PMMA) bone cement (fig 4). After the surgery the pain disappeared and the patient was able to walk again, although an incomplete drop foot persisted. Unfortunately, after another steady period, further progression occurred recently and a fifth decompression surgery had to be performed. With a follow-up of over 17 years a point has arrived at which further surgery has become a serious dilemma.

## DISCUSSION

Bone is affected in 0.5 to 4% of the cases of hydatid disease (9,17,18). In half of these cases, vertebral bones are involved, reflecting the preference of the parasite for highly vascularised bone (7,9,18). In order of frequency, spinal lesions are located at the thoracic, lumbar, and least often, cervical region (18). Involvement of the sacral spine, as in the current case, is very rare, with less than 10 cases reported in literature (4,11,4,15,20). After ingestion, the parasite reaches the vertebral bone through porto-vertebral shunts by paradoxical flow during moments of increased intra abdominal pressure (2,13,20). Five different patterns of spinal involvement have been described : primary intramedullary, intradural intraspinal, extradural intraspinal, vertebral and paravertebral (7,20). In the current case, a combination the last three types was seen.



**Fig. 4.** — Post-operative radiograph showing the bone defect of L4 being filled with PMMA.

The parasite cyst generally consists of an inner layer (endocyst) and outer layer (ectocyst). The formation of new scolices and daughter cysts takes place inside the cyst, at the internal (germinal) layer, leading to the typical multilocular aspect of the cysts. The host defence reaction leads to a fibrous capsule (pericyst), which is vascularised to provide nutrients for the parasite (15,18). Because the host defence is only marginally in bone tissue, the outer fibrous capsule is usually thin or even missing (2,9,15). On plain radiographs, the cyst generally presents as a small osteolytic lesion, with ill-defined margins with absence of any periosteal reaction (2,15,18). More advanced disease is recognised at multiple sharply demarcated, expanding, osteolytic lesions, with thin septa inside, giving the typical honey-comb appearance (2,15,18). Rarely, a cyst can be seen on conventional radiographs as a round calcified lesion. Calcification usually means death of the parasite, but this is certainly not a rule, since cases of calcified lesions that restart to grow after more than 40 years of dormancy have been reported (18). Moreover, most cysts do not calcify at

all, and typically enlarge gradually over time (18). CT studies typically reveal one or several closely related, well defined, osteolytic lesions (fig 1). Other features that are associated with bone hydatidosis are bone expansion, cortical thinning, cortical destruction, sclerosis and extension into adjacent soft tissue (2,15,18). On T1-weighted MRI images the smaller cysts present as low signal intensities against a relatively light background of the larger cyst (fig 2) (15). Contrast administration may only result in mild enhancement that remains restricted to the fibrous capsule (15). Additional features revealed by MRI studies are the relationship of the lesion with neural structures, spinal cord, and extension in soft tissue (2,15,18). In cases presenting with rather non specific cystic lesions that are few in number, lacking daughter cysts and involve only one tissue compartment, diagnosis may be difficult. Aneurismal bone cysts, cystic components of giant cell tumours, cystic metastasis or epidermoid cysts should be considered in the differential diagnosis (14,15,18). In case of reactive enhancing tissue or calcification, neurogenic tumours related to the spine like cystic schwannoma or cystic extramedullary ependymoma should be considered as well (14,15,18).

Histologic evaluation usually shows numerous clusters of giant cells, bone destruction and bony fragments (fig 3 a, b). Characteristic scolices may not be identified even after extensive additional sections. This can be explained by the osseous resistance, in which larvae develop by exogenous budding, and scolices are rare (2,9).

There have been no well-designed clinical trials performed for any treatment modality and it is generally agreed that treatment of spinal hydatid disease should be reserved for symptomatic lesions (8). Ideally, complete resection of the cyst is performed followed by albendazole therapy for at least one year (5). Unfortunately, radical resection is generally impossible in spinal hydatidosis and decompression is the highest achievable goal (1). Spillage, which can induce severe anaphylactic reactions and implantation of vesicles during excision, is very common (1,5). These risks however, can be minimised by prevention of rupturing of cysts and injecting formalin, alcohol, 0.5% silver

nitrate or hypertonic saline into the cyst and surroundings (5). Chemical sterilisation however, does not kill all microscopic daughter cysts (16). Initially, we only performed a subtotal resection of the cyst to obtain tissue for histologic evaluation, which retrospectively was certainly not the treatment of choice. At that time however, hydatidosis occurred incidentally in our area, and susceptibility was not raised. During the second surgery we performed extensive debridement of bone and soft tissue, and all surrounding tissue was thoroughly lavaged with hypertonic saline followed by antihelminthic therapy. According to Karray *et al* (6), this is the maximum effort to increase the remission period. Some authors recommend PMMA reconstruction of the vertebral body defect for its antihelminthic effect (fig 4) (19). Little is known about the exact prognosis of spinal hydatidosis. After initial presentation of symptoms the average survival is less than 5 years (5,16). Even after the combined strategy of surgery and antihelminthic therapy, the reported mortality exceeds 50% within 5 years follow-up (5,10,16).

In conclusion, we present a case of disseminated lumbosacral hydatidosis, which was treated with incomplete decompression surgery followed by chemotherapy. With multiple decompression procedures and continuance of antihelminthic therapy, further progression could be controlled for almost two decades. However, further progression has now become inevitable.

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#### Note from the editors

The editors are sorry to announce the sudden death of Dr Paul Wuisman on July 25, 2007. They condole with the family and the Nederlandse Orthopaedische Vereniging.

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