

RESEARCH ARTICLE

Non-spinal Hydatid Disease of Bone: A Series of Nine Cases

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Abstract

Background: Extra-spinal osseous hydatid disease is reported in a small number of case series. In the present study, we report our experience with extra-spinal hydatid disease of the bone in a series of nine patients.

Methods: In this retrospective study, the patients who were diagnosed with an extra-spinal osseous hydatid disease were included. All patients were treated surgically. Preoperative anthelmintic drugs were employed for the cysts that were diagnosed before the operation. Postoperative chemotherapy was performed for all patients.

Results: The study population included nine patients, including seven males and two females, with a mean age of 45.2±7.9 years and an average follow-up of 4.1±2.7 years. Non-specific pain was the most common symptom at presentation. Pelvic bones were the most frequent site of involvement. Serologic tests were false negative in seven patients. The disease was diagnosed preoperatively in five patients, and all of them were located in flat bones. The cysts were treated by radical excision in four patients, extended curettage in four patients, and amputation in one patient. The recurrence of the lesion was recorded in two patients who were treated by intralesional curettage. One case of suppuration was the only postoperative complication of this series.

Conclusion: Osseous hydatid disease is a serious disease with challenging diagnosis, difficult treatment, and significant morbidity. Preoperative diagnosis is generally easier in flat bones. Radical resection is the optimal treatment of this disease, while non-radical resection is associated with a higher risk of recurrence.

Level of evidence: II

Keywords: Bone, Hydatid disease, Treatment

Introduction

Cystic echinococcosis, also known as hydatid disease, is a potentially serious condition caused by the *Echinococcus granulosus* or hydatid worm. The larval stage of this parasite forms hydatid cysts in intermediate hosts, with the liver and lung being the most involved site of hydatid cysts (70% and 12%, respectively) (1, 2). The involvement of bone is rare, even in the endemic regions, accounting for 0.5%-2.5% of all hydatid cysts, half of which occur in the spine (3, 4).

The cysts generally remain asymptomatic for a long time; as a result, the diagnosis is usually made at the late stage, when the extension of cysts makes the surgical resection difficult. Radiological findings are non-specific, and many cases are diagnosed accidentally (5). Serological assays also lack enough sensitivity and

specificity, making the diagnosis of these cysts more challenging (5). The prognosis is poor, particularly where the radical excision of the lesion is anatomically difficult, leading to an increased risk of cyst recurrence (5, 6). The disease acts like a malignant tumor, and chemotherapy is used as an adjunct treatment or when the surgery is not possible due to an anatomic barrier (5).

The high morbidity and mortality rate of hydatid disease highlights the need for improved diagnostic and therapeutic procedures. A more clear insight into the characteristics of the disease is a critical step toward the development of new diagnostic and therapeutic modalities, and case series studies are valuable tools to achieve this goal. Nonetheless, due to the rare incidence, a small number of case series have reported osseous

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hydatid disease in extra-spinal regions, and therefore, several aspects of these lesions are not well-understood. For instance, the differences in the diagnosis, prognosis, and treatment of extra-spinal hydatid disease of the long and flat bone have not been discussed in earlier studies.

In light of the aforementioned issues, the present study aimed to report the characteristic features, as well as the treatment approach and outcome of the treatment, in a series of nine patients with the osseous hydatid disease of extra-spinal regions.

Materials and Methods

This retrospective study was approved by the review board of our institute, and patients provided written consent to the publication of their medical data. Between 2002 and 2018, the patients who were diagnosed with an osseous hydatid disease at our referral orthopedic hospital were evaluated. Patients were included if histological documentation of the diagnosis was available. On the other hand, patients with spinal involvement (n=5) and concomitant involvement of other organs were excluded from the study (n=3). Patients with a follow-up of fewer than two years (n=2) were excluded from the study as well. Finally, nine patients were included in the study.

The demographic characteristics of the patients, including age and gender; clinical characteristics, such as the presenting symptom and location of the lesion, and treatment characteristics, including the method and outcome of treatment, were extracted from the patients' profiles. The serologic test results for hydatidosis (either indirect hemagglutination or enzyme-linked immunosorbent assay) were studied before the surgery. The peripheral blood eosinophil count was also checked as a complementary test, and an eosinophil count of $> 500/\text{mCL}$ ($> 0.5 \times 10^9/\text{L}$) was defined as Eosinophilia.

Radiologic evaluations were performed using plain radiographs, computed tomography (CT) scans, magnetic resonance imaging (MRI), and sonography. Complete radiologic records were available for five patients. For the remaining four patients, sonography was not performed. For one patient who was presented with a pathologic

fracture (case 5), a CT scan and MRI were not performed as well. A plain radiograph was used for initial screening. The CT scan was used for the evaluation of cortex destruction, while MRI was utilized for the identification of extraosseous lesions. A sonographic evaluation was used for the detection of cystic floating membranes and hydatid sand in soft tissue.

The surgical procedure included the excision of the soft-tissue cyst in the first step. Regarding the bone cyst, the lesion was radically excised if it was possible without significant morbidity. For cysts involving the ilium (n=3), the involved bone was excised with about a 5-mm margin using an iliofemoral approach. For the cyst of the scapula (n=1), partial scapulectomy was performed through a posterior longitudinal incision (7). When the radical excision of bone was associated with a serious disability, the lesion was treated with extended curettage (8, 9) and irrigation with hypertonic saline (30%) for 3 min [Figure 1]. To facilitate early mobilization, the defect was augmented with bone cement whenever the lesion was located near the diarthrodial joint; otherwise, the defect was filled with bone graft.

For patients in whom the disease was diagnosed preoperatively, both neoadjuvant and adjuvant chemotherapy were implemented. The chemotherapy regimen was albendazole (10 mg/kg/day) with cycles of 28 days and two weeks of rest. Patients in whom the disease was diagnosed preoperatively (n=5) received two cycles of chemotherapy before the operation. All patients were administered five to seven courses of chemotherapy (average six cycles) after the operation.

The follow-up visits were every three months for one year. In each follow-up session, the sonographic and radiographic evaluation was performed to rule out the recurrence. Serologic tests and peripheral blood eosinophil were also checked in each follow-up session to help the diagnosis of recurrence. Recurrence was treated with the same protocol used for primary hydatid disease.

Results

The study population included nine patients, including seven males and two females. The mean age of the



Figure 1. (a & b) anteroposterior and lateral radiograph of the hydatid disease of the distal tibia in a 46-year-old male (case 6), (C & D) anteroposterior and lateral radiograph of the same patient two years after treatment with extended curettage and cementation.

Table 1. Characteristic features of patients with osseous hydatid cyst

ID	Age (year)	Sex	Symptom	Symptom duration (month)	Location	Eosinophilia	Serologic test	Preoperative diagnosis	Treatment	Follow-up (year)	Recurrence
1	39	Female	Pain	84	Ilium	Positive	Positive	Yes	Radical resection	6	No
2	59	Female	Pain & Mass	120	Tibia	Negative	Negative	Negative	Extended curettage & bone graft	5	times 3
3	51	Male	Mass	60	Ilium	Positive	Positive	Yes	Radical resection	5	No
4	42	Male	Pain & Mass	30	Ilium	Negative	Negative	Yes	Radical resection	4	No
5	41	Male	Pain & fracture	96	Femur & Pelvis	Positive	Negative	Negative	Hind quarter amputation	3	No
6	46	Male	Pain	8	Distal Tibia	Positive	Negative	Negative	Extended curettage & cementation	2	No
7	51	Male	Pain & Mass	3	Sacroiliac joint	Negative	Negative	Yes	Extended curettage & bone graft	5	No
8	46	Male	Pain & Mass	10	Scapula	Negative	Negative	Yes	Radical resection	4	No
9	32	Male	Pain & Mass	6	Radius	Negative	Negative	Negative	Extended curettage & bone graft	2	times 2

patients was 45.2 ± 7.9 years (range of 32 to 59 years). The mean follow-up of the patients was 4.1 ± 2.5 years, ranging from to 6 years. The main symptom at presentation was non-specific pain that was observed in eight patients. The swelling (mass) was the second most frequent symptom at the presentation that was detected in six cases. One patient presented with a pathologic fracture. Two osseous hydatid cysts were observed in this patient (one in the femur and one in the pelvis). The pelvis was the most common location of the cyst ($n=5$). The mean symptom duration was 46.3 ± 44.8 months, ranging from 3 to 120 months. The characteristic features of patients are summarized in Table 1.

Out of nine patients, preoperative diagnosis was made in five cases in flat bones (three iliums, one sacroiliac

joint, and one scapula). In these patients, the CT scan revealed a well-defined partial disappearance of one or both cortex without sclerosis and periosteal reaction. The MRI showed a multilocular extra-osseous cyst with locules of varying sizes. Sonographic characteristics were only positive in two of these five patients. Plain radiographs revealed an osteolytic lesion in all patients. The eosinophil count was above the normal range in four patients. The serologic test for hydatid disease was only positive in two patients. Laboratory tests, such as erythrocyte sedimentation rate, C-reactive protein, complete blood count, biochemical tests, and other serologic tests, were in the normal range [Table 1].

The lesion was treated with wide excision [Figure 2] in four patients, extended curettage [Figure 3] in four

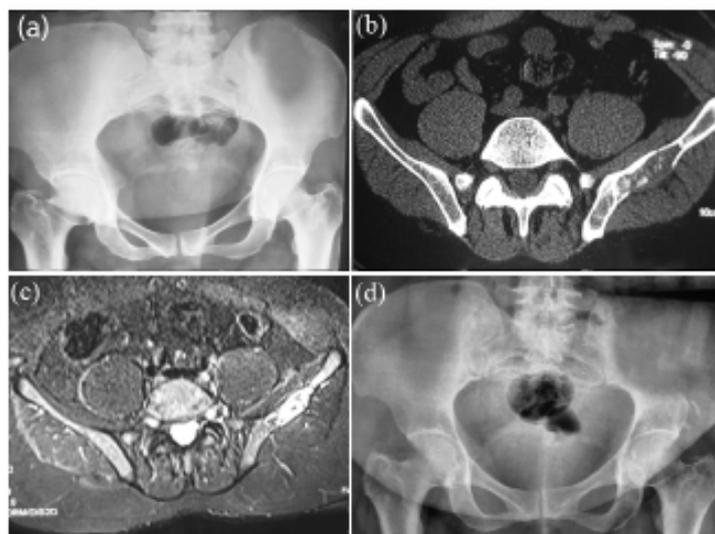


Figure 2. Hydatid disease of the ilium in a 39-year-old female with a preoperative diagnosis (case 1), (a) anteroposterior radiograph showing the cystic lesion in the ilium; (b) CT scan of the lesion showing partial disappearance of the cortex without sclerosis and periosteal reaction, (c) Axial fat-suppressed T2 MRI showing a lesion with the hyperintense signal, (d): Five-year postoperative radiograph of the patient showing the healing of the lesion after wide excision.

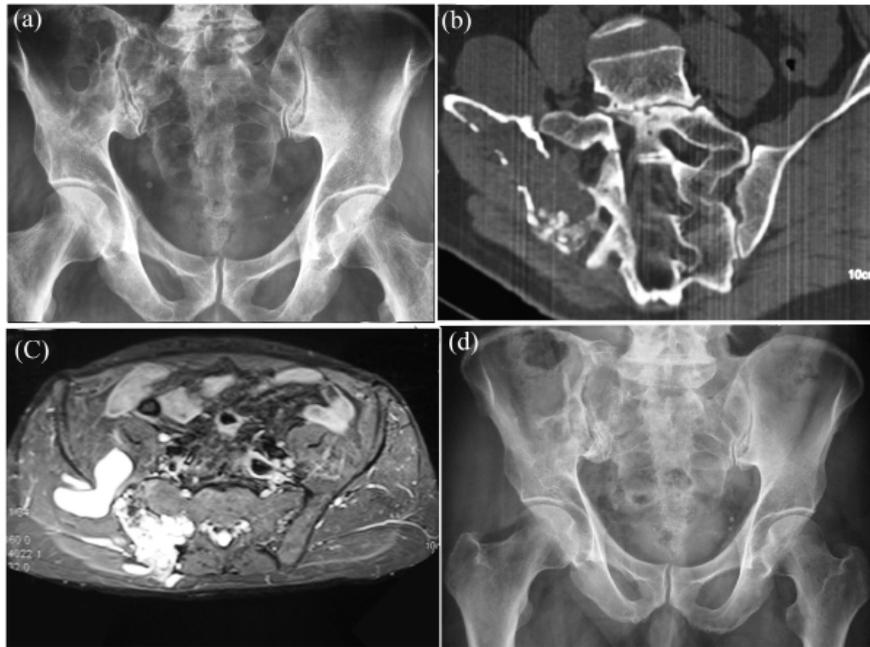


Figure 3. Hydatid disease of the sacroiliac joint in a 51-year-old male with a preoperative diagnosis (case 7), (a) anteroposterior radiograph showing the cystic lesion in the sacrum and ilium; (b) CT scan of the lesion showing partial disappearance of the cortex without sclerosis and periosteal reaction, (c) MRI of the lesion showing multilocular extra-osseous cyst with locules of varying size; (d): seven-year postoperative radiograph of the patient showing the healing of the lesion after extended curettage & bone graft.

patients, and amputation in one patient. In patients who underwent extended curettage, the defect was augmented with bone graft in two patients and bone cement in one patient. The lesion recurred in two patients, and both of them were managed with extended curettage. In one of them (case 2), the lesion recurred three times during a six-year follow-up. In the other one (case 9), the lesion recurred two times during a three-year follow-up. The only observed postoperative complication was a case of suppuration that occurred in a patient with the involvement of the tibia (case 2). This complication was managed with drainage and antibiotic therapy.

Discussion

As evidenced by the results of this study, patients with osseous hydatid disease of extra-spinal regions generally present with long-term non-specific symptoms, such as pain and mass. Pelvic bones are the most frequent site of hydatid disease of bone in extra-spinal regions. Laboratory tests lack enough sensitivity to detect these lesions. The cysts might recur in a considerable number of patients, particularly if the lesion is not radically extracted.

In the present study, hydatid disease of flat bones revealed characteristic features in the CT scan, which included the disappearance of cortex with no sclerosis and periosteal reaction. These features, combined with a multilocular extra-osseous cyst in MRI, could be regarded as unique characteristics of hydatid disease of flat bones and help preoperative diagnosis, thereby providing a minimized risk of intraoperative disease spread through

implementing preoperative chemotherapy. Chen et al. assessed the imaging feature of the hydatid disease of the pelvis in 15 patients. They concluded that owing to the thin pelvis cortex, the lesion easily penetrates, destroys the cortex, and invades the surrounding soft tissue. In their series, out of 15 cases of pelvic hydatid disease, soft-tissue invasion was observed in 13 subjects. Thinning of bone cortex and cystic expansive bone destruction were discerned in nine patients in their study. Erosive bone destruction was detected in three patients (10). The results of the present study are in accordance with the finding of the stated research regarding the imaging characteristics of hydatid lesions in flat bones.

Loudiye et al. reviewed 11 cases of extra-spinal hydatid disease of bone. The mean time interval from symptom onset to diagnosis was 22.7 months. The pelvis was the most frequent site of involvement (n=6). The eosinophil count was above normal in four patients. Serological tests were positive in five patients. Surgical treatment was performed in 10 patients and it was radical in only three patients. The lesion recurred in five patients at a mean follow-up of six months. They attributed their high rate of recurrence to difficult access to the lesion (11). The mean time interval to diagnosis was 46.3 months in our patients, and the pelvis was the most frequent site of involvement in our series as well. The lesion recurrence was more prevalent in patients who were treated with intralesional curettage. Nonetheless, the reason to select intralesional curettage was not the difficult access but to prevent the significant morbidity that radical excision might cause.

Herrera et al. reported extraspinal bone hydatidosis in 26 patients with a mean age of 51.5 years and a mean follow-up was 12.8 years. The ilium was the most frequent site of involvement in this series (n=14). Up to the last follow-up session, the lesion recurred in seven (27%) patients. They reported that cases with the involvement of both the ilium and the hip are the most challenging to treat (12). In agreement with the study by Herrera et al., the recurrence rate was 33.3% in the present series. The simultaneous involvement of the ilium and hip was detected in one of the patients in the current study.

Arazi et al., in a case series study, reported the hydatid diseases of the bone (n=8) and muscles (n=7). The lesions were treated with both surgery and antihelminthic chemotherapy. The average follow-up of the patients was 30 months. Recurrence was recorded in four patients with bone involvement, while it was not observed in patients with muscle involvement (13). These results further support the role of radical resection in the local control of hydatidosis since the cysts could be more radically resected in muscles than bones.

Xie et al. reviewed the clinical data of 40 patients with osseous hydatid disease, out of whom 24 cases were treated surgically, and the remaining 16 patients were managed by radiotherapy. Lesions recurred in 14 (58%) and 3 (21%) patients in the surgery and radiotherapy groups, respectively. Patients in the radiotherapy group were significantly more satisfied, and radiotherapy was not used for any patient in the present series (14).

Technically, filling the defect with cement could have a more preventing effect on the recurrence of the lesion since the monomer toxicity heat generated by cement polymerization can kill the remaining larvae (15-17). In this respect, both recurrences in the present series were observed in the lesions with bone graft augmentation. Neoadjuvant chemotherapy is also attributed to the reduced rate of recurrence; however, the recurrent cases of the present series did not receive neoadjuvant chemotherapy since the lesion was not diagnosed before the surgery (18).

In line with the results of the present study, a review of the literature revealed that extra-spinal osseous hydatid disease could be a serious disease with a high rate of mortality and morbidity. Serological tests are not reliable for diagnosis, and other diagnostic modalities, such as CT and MRI, could be helpful. The resection of the lesion in regions with an anatomic barrier is challenging and prone to recurrence. Among the notable limitations of this study, we can refer to the small sample size and the retrospective design of the research. However, these limitations are inevitable when reporting rare disorders, such as extra-spinal hydatid disease of the bone.

Extra-spinal osseous hydatid disease is more frequent in the pelvic area. Serological tests are false negative in the majority of cases. In flat bones, CT findings (cortex disappearance), along with MRI findings (multilocular extra-osseous cysts), could help preoperative diagnosis and provide a more efficacious therapeutic plan. Radical resection is the optimal treatment of choice, and non-radical resection is associated with a high risk of disease recurrence.

Informed consent was obtained from the patients to use their medical data for publication.

Disclosure:The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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