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MEDICINE

Inoperable Primary Hydatidosis of Pelvic Bones with Involvement of Gluteal Muscles: Diagnosis of a Rare Presentation and Follow-up

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Abstract

Primary hydatid cysts typically occur in the lungs or the liver, but can be found elsewhere without involving these organs. Primary bone cysts are rare and frequently missed. No standard treatment exists, especially for inoperable patients. A 53-year-old female patient with pain and swelling in the left gluteal region for a month was first admitted 3 years ago, with an initial diagnosis of DVT. Incidentally, magnetic resonance imaging showed cystic involvement of the pelvis, and an iliac bone biopsy suggested a diagnosis of hydatidosis. The cyst was inoperable, and praziquantel and albendazole chemotherapy was initiated. The patient was followed after 3 years, having a palpable 12 × 9 cm mass and showing minor radiographic changes. We report an unusual and inoperable case of primary bone hydatid cyst, with involvement of pelvic bones and surrounding soft tissue. This case suggests that cystic mass, especially where disease is endemic, should raise suspicion of echinococcosis. The chemotherapy regimen appears to have limited the growth of the cysts.

Keywords Hydatid cyst · Musculoskeletal hydatidosis · Bone cyst · *Echinococcus granulosus*

Introduction

Hydatidosis is a common parasitic disease resulting from the infestation of various organs by tapeworms of the genus *Echinococcus* [8]. Most commonly, hydatid cysts remain dormant even in older adults, but parasitic overload, enlarging cysts, or cysts in unusual body parts can cause various symptoms [8]. Hydatidosis can theoretically involve every organ, including the liver (63%), lungs (25%), and muscles (5%) and less commonly the skeletal system, kidneys, brain, spleen, and even the eyes [13]. Most of the symptoms of the disease are caused by local damage, and thus are specific to infected organs.

Bone involvement comprises 0.5–4% of all primary hydatidoses [14], and the most commonly affected bones are the vertebrae, the long bones of the lower limbs, and the pelvis [2]. Pelvic hydatidosis accounts for 28% of all bone hydatidosis cases [20]. Primary peripheral muscle involvement is rarer and happens in less than 1% of cases [10].

Here, we report a case of inoperable primary hydatid cyst with involvement of the pelvic bones and gluteal muscles. Albendazole chemotherapy seems to be the only widely used drug for inoperable osseous hydatidosis, even in cases of extensive disease, but its effectiveness in such cases is questionable [11]. The addition of praziquantel is relatively rare in literature on osseous hydatid cysts [11], but it might increase the effectiveness of albendazole [1].

Case Presentation

A 53-year-old woman was admitted with a slowly growing painful swelling in the left gluteal region for 1 month. The patient's vital signs were stable on admission and the left hip had a limited range of motion, and evidence of local inflammation was observed. There was a notable history of occasional contact with farm animals.

She was initially hospitalized with a diagnosis of deep vein thrombosis, and received warfarin. While being treated, she developed neurological symptoms due to intracranial hemorrhage, and a craniotomy was performed. Her laboratory test results indicated systemic inflammation (ESR > 120, CRP 88).

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During further workup of the gluteal swelling in a three-phasic bone scan, angiographic and blood pool images of the pelvis revealed mild hyperflow hyperemia in the left hip, and static images showed an area of increased tracer uptake in the left acetabular roof and the left femoral head. These findings were consistent with chronic osteomyelitis or osteoarthritis. Magnetic resonance imaging (MRI) showed several large cystic and multiseptated lesions with ring enhancement in the pelvic cavity with extensions to buttock and inguinal area and also left pubis. The left femur, acetabulum, and iliac crest all showed signal changes, and involved soft tissue included the iliopsoas muscle (Fig. 1). The total size of the lesion was about 105 × 69 × 89.

Surgical drainage of the mass was performed (339 cc total volume, and about 200 cc pus was aspirated), and fine-needle aspiration cytology (FNAC) produced a yellowish creamy fluid and microscopic examination revealed some granular debris material in the background, few macrophages, some inflammatory cells, and few degenerated cells with undetermined significance. A second MRI, 7 months after the first one, also revealed a large heterogeneous signal intensity mass involving the left pelvic bones, and a heterogeneous signal involving the left femoral head and neck. Bacterial cultures of the cystic fluid were negative and in serological tests, anti-*Echinococcus* IgG was positive. Finally, a year after the initial presentation and based on a differential diagnosis including hydatid cyst, a left iliac bone biopsy was performed. The biopsy confirmed suspicions of hydatidosis of the pelvic bones with soft tissue and muscle involvement, based on the history, examination, imaging, and laboratory results.

Treatment was initiated with a combination of albendazole (400 mg BD) and praziquantel (500 mg TDS). Surgery was deemed to pose an undesirably high risk considering the sensitive location of the cyst, its large skeletal and extra-skeletal components, and its proximity to spinal nerves, and chemotherapy was continued.

A year after the initiation of treatment, computerized tomography (CT) scan of the hip expectedly showed expansible lytic lesions with soft tissue density every part of the left iliac bone as well as in the left femoral head; but notably, no further involvement of the iliopsoas muscle and the gluteal region was reported, and the cyst had shrunk by 25% to 90 × 26 mm.

The patient was admitted once again in 2018, 2 years after the start of treatment. The patient again had a complaint of reduced range of movement, pain, swelling, and tenderness in the left hip joint which had worsened progressively in the month before admission. The patient's vital signs were stable on admission, with a blood pressure of 160/80 mmHg, pulse rate of 80/min, and temperature of 37 °C. On physical examination, the range of motion of the left hip (passive and active) was limited. A diffuse, tender, and fluctuating cystic swelling (approximately 12 × 9 cm) in the left gluteal area was palpated. Sedimentation rate was 40 mm/h and C-reactive protein was 20 mg/L.

With the signs of a new local inflammation, the cyst was drained, which produced a yellow fluid. Chemotherapy was continued and MRI of the pelvis and thigh was performed. A multiloculated fluid in the left pelvis was observed, with transfacial spread from the iliopsoas region involving the iliac medullary

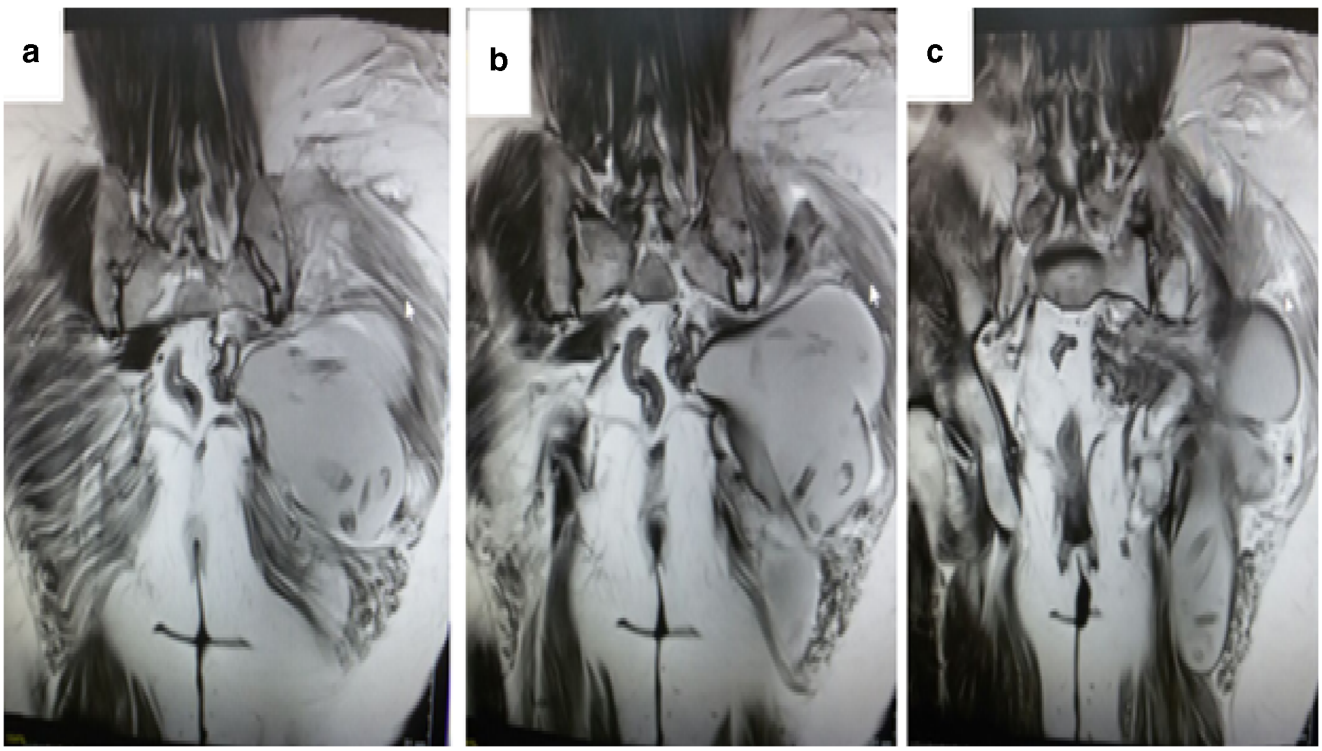

Fig. 1 MRI of the left thigh, sequence T2-weighted: ovular mass in the gluteal muscle, with a cystic aspect containing numerous cystic formations with regular outlines, before treatment

cavity (first locule, 85 × 44 mm) extending inferiorly to the acetabulum, pubis and ischemic, and also the iliopsoas bursa (second locule, 51 × 38 mm). A third locule (56 × 17 mm) in the left gluteus minimus muscle was detected. The pelvic lesion descended into the proximal thigh just beneath the posterior medial fascia, with a maximum diameter of 75 × 32 mm and total length of 124 mm (Fig. 2). Considering the probability of a new superinfection causing cystic swelling, we decided to follow the patient and wait for the acute inflammation to subside before considering further treatment options while continuing chemotherapy. After the resolution of the acute inflammation, symptoms notably improved. In a follow-up interview after 3 months, the Harris Hip Score (HHS) was 44.50. The HHS was used because it is a well-validated scale used to assess hip joint function and has been validated in the context of a variety of disorder [12], and has also been previously used in Iran [4]. While this is considered a poor result, the HHS score would have been about 25–30 in the course of the superinfection episode and 30–35 prior to the initiation of the anti-helminthic treatment. The patient also considered her symptoms to have been significantly alleviated.

Discussion

Musculoskeletal hydatidosis is considered a differential diagnosis for any painful mass in the gluteal area in patients with a previous history of hydatid cyst [10] but primary involvement of the musculoskeletal system is rare and commonly overlooked [14]. Musculoskeletal hydatidosis is frequently overlooked in the differential diagnosis of musculoskeletal masses, and mistaking it for a tumoral mass has been reported in the literature [17]. While the first case of primary bone hydatid cyst was reported two centuries ago [18], case reports in the literature are still rare. A recent review found only 101 cases (both primary and secondary) of pelvic hydatidosis [16].

The definitive diagnosis of hydatidosis is based on discovering the cysts within infected tissue, but it can alternatively be diagnosed with a combination of serological tests, fine-needle aspiration (FNA), and imaging modalities [6]. FNA and cell block have also been used to diagnose isolated musculoskeletal hydatidosis [5].

There are no widely accepted treatment guidelines, and the disease has proven to be difficult to treat and has relatively high chances of recurrence [7, 16]. For medical treatment of hydatid cysts, albendazole is commonly chosen due to its significant scolicidal effect, and it can be used with or without surgery or aspiration. It has been noted that no drug has been specifically developed for the treatment of musculoskeletal hydatidosis [17]. For bone hydatid cysts, surgery and cyst excision are usually necessary, and common options include hemipelvectomy, curettage, and debridement [7, 9, 17]. A case report and review of literature somewhat similar in scope to ours, concerning gluteal musculoskeletal hydatidosis, have also emphasized the necessity of both extensive surgical excision and anti-helminthic pharmacotherapy [15]. Unfortunately, our patient was deemed inoperable due to the location and extent of involvement. We elected to continue albendazole and praziquantel chemotherapy, considering that albendazole is a popular and effective drug (with or without praziquantel) in these cases [1]. One promising modality which might help bone hydatidosis patients such as ours (and which we indeed considered) is radiotherapy, but not much evidence is available [19].

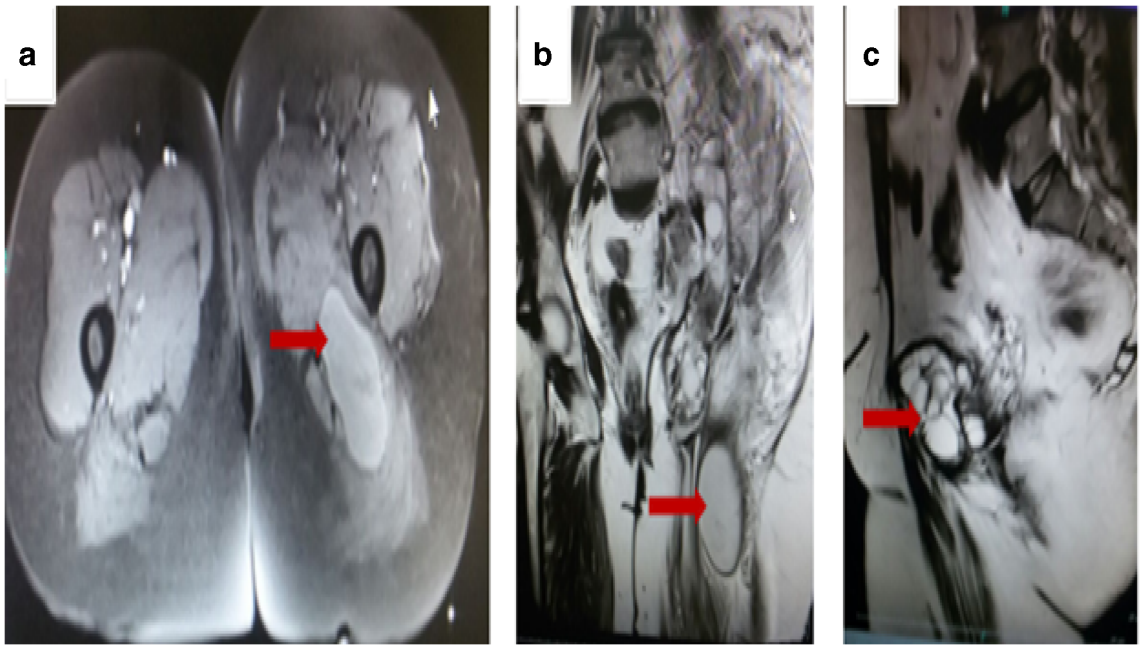

Fig. 2 MRI of the left thigh, sequence T2-weighted. A multiloculated fluid in the left pelvis with involving the muscles and bones, after medical treatment

To conclude, primary musculoskeletal hydatidosis without any evidences of liver or lung involvement is rare and easy to overlook. Delayed diagnosis might lead to extensive and inoperable lesions, bacterial superinfections [3], and permanently worsened quality of life. In our case, the diagnosis was based primarily on sonography, CT scanning, MRI, and biopsy, and treatment with drainage combined with long-term antihelminthic therapy was chosen. The cyst was not excised, and the patient will be followed for further complications.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Statement An independent ethics committee in Tehran University of Medical Sciences reviewed and approved the study protocol and the consent form. Informed consent was obtained from the patient for every diagnostic and therapeutic procedure. Separately, informed consent was obtained to write this report and obtain the relevant information contained therein.

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