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Case Report

Giant primary muscular hydatid cyst with a secondary bone localization

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ABSTRACT

Primary musculoskeletal hydatidosis is less frequent than hydatidosis of the parenchymal organs. This localization has been little studied and so there is little information in the literature on the subsequent disease evolution. We present a case of primary hydatidosis of the abductor muscle that came to medical attention very late. After complete surgical removal of the huge mass, a secondary bone localization developed, causing a femoral pertrochanteric pathological fracture. The case described is exceptional in view of both the localization and the great size of the primary multi-lobed muscle hydatid cyst. We underline the difficulties of diagnosis and treatment of both the primary muscle localization and the secondary bone recurrence.

1. Introduction

Human hydatidosis is an endemic infection in the Mediterranean Basin, Central Asia, East Africa, the southern part of South America, and in Oceania.[1–3] The disease is caused by *Echinococcus granulosus*, whose larvae develop in cystic form, most often in the liver and lung.[1,4] Involvement of other organs, and particularly of the muscles, is rare, even in endemic areas.[1,5] The case we describe demonstrates that in cases of onset of a cystic mass in endemic areas for this zoonosis, it is essential to take into account the possibility that it might be echinococcosis, even when the lesion presents in a rare localization. In addition, the peculiar rarity of the size of the hydatid cyst we report, as well as the subsequent bone involvement, make it necessary to consider whether treatment associating surgical excision with anthelmintic treatment is indicated in such cases.

2. Case report

A 79-year-old housewife from southern Italy was admitted to the General Surgery Unit of Bari Polyclinic for a voluminous painful mass in the medial region of the left thigh. The mass had started developing three years before. The patient's clinical history was unremarkable except for intermittent episodes of mild fever and pruritus, not present at the time of admission. Clinical examination showed that the mass had a solid consistency, was localized in the abductor region and measured approximately 40 × 10 cm. Blood tests, X-ray of the thigh, cranial and chest–abdomen–pelvic computed tomography (CT) scans were all within the normal range. Magnetic resonance imagining (MRI) demonstrated a multi-lobed mass extending from the flexor to the abductor muscles. The mother cyst showed a low intensity signal in weighted T1 and T2 and the numerous daughter cysts were surrounded by intracystic fluid (Figure 1).

On the suspicion of a hydatid cyst, more detailed questions were put to the patient, who revealed that she used to keep a dog, which had died 5 years before.

A diagnosis of primary muscular hydatidosis was made and surgical excision was performed. Passing between the abductor muscles (long and short) and the sartorius and gracilis muscles, a voluminous cyst, partially infiltrating the great abductor muscle, was individuated. The cyst was completely removed with particular care to prevent accidental dissemination of the daughter cysts (Figures 2 and 3).

Histopathological examination of the mass, measuring 37 × 12 × 10 cm, revealed that the cyst wall consisted of largely lamellar, acellular, eosinophilic matter. The internal surface was covered by a squamous cuboid epithelium, connective tissue, and a mononuclear inflammatory infiltrate. Indirect hemagglutinin tests for Echinococcus were positive at 1:3200, confirming the diagnosis of hydatidosis, classified as primary in view of the absence of other localizations.

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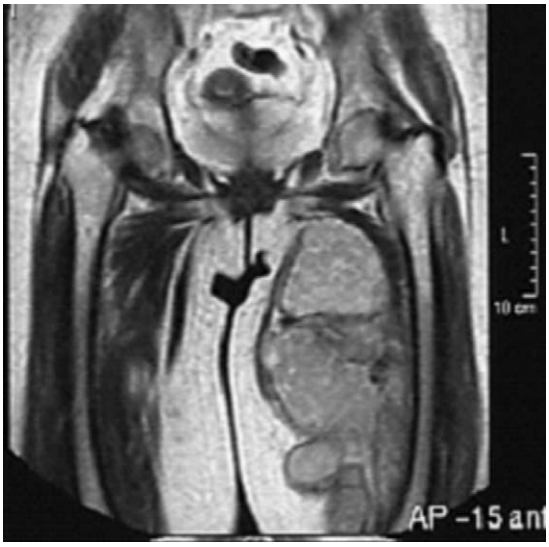

Figure 1. MRI showing the primary muscle hydatidosis localized in the abductor region of the thigh.

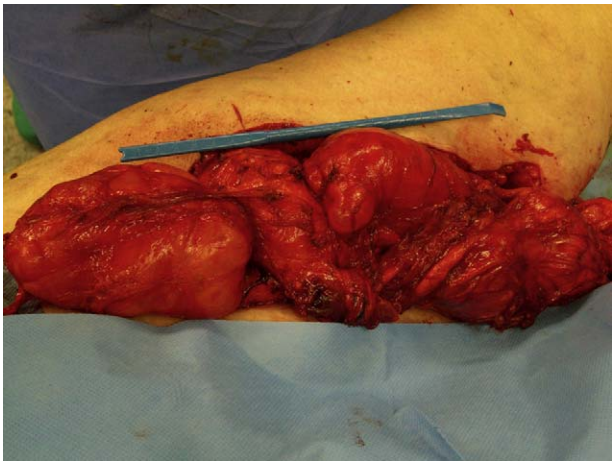

Figure 2. Surgical resection of the giant muscle cyst.

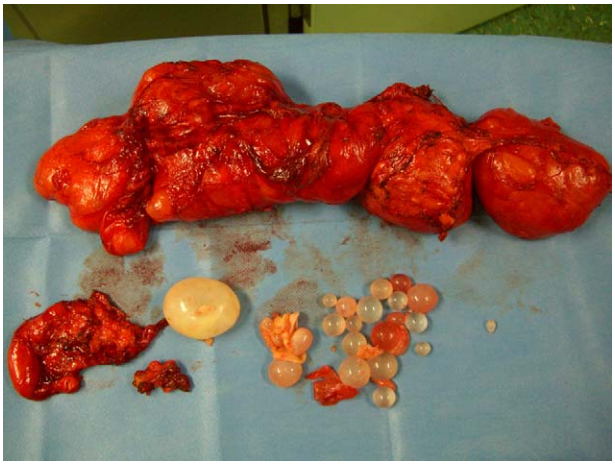

Figure 3. Macroscopic aspect of the hydatid cyst after the surgical excision.

At the 12-month follow-up, the patient showed no signs of reactivation of the disease. However, after 14 months she suffered a trauma of the left thigh that resulted in a pertrochanteric fracture of the femur. Radiography demonstrated the presence of a cystic lesion, revealing a pathological origin of the fracture (Figure 4).

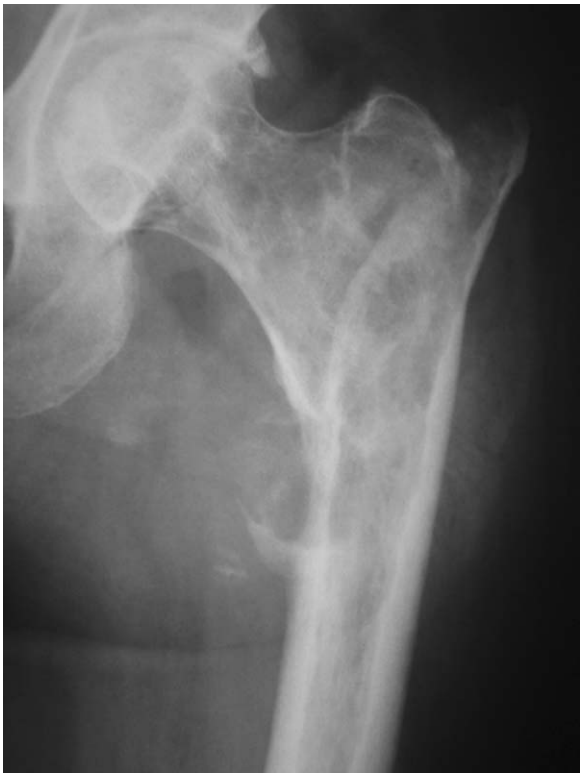

Figure 4. X-ray of the left femur showing the pertrochanteric fracture and the secondary hydatid bone cyst.

Although blood tests were still normal, a secondary localization of the hydatidosis was suspected and so ample local curettage was performed intraoperatively, before placing a femoral prosthesis. Microbiological, serological and microscopic tests of the cyst fluid revealed the presence of bone tissue with a hyaline, germinative membrane, lymphocytes and monocytes, confirming the diagnosis of secondary bone hydatidosis. The patient was prescribed albendazole therapy at a dosage of 10 mg/kg/day, to be taken for 6 months. After surgery and up until the last follow-up at 20 months from the first diagnosis of hydatidosis, no infectious spread or septic complications have been observed, or any other secondary localizations. The patient gave her consent to the reporting of her case.

3. Discussion

We describe a unique case of a giant multi-lobed hydatid cyst localized in the striped muscle of the thigh, complicated after surgical excision by disease recurrence in the adjacent femoral bone.

Cystic echinococcosis is a cosmopolitan parasitosis that is endemic in zones where there is widespread sheep farming.[6] Regions with the highest prevalence are the Mediterranean Basin, the Balkans, the Middle-East and northern Africa, as well as the southern part of South America, Central Asia, Mongolia, Xinjiang and Tibet in China, and East Africa. In Italy, the disease is present in the south and the islands.[7] This parasitic anthropozoonosis is caused by invasion of the tissue by the larvae of *Echinococcus granulosus*, whose intermediate host is the sheep and final host the dog.[8] The larvae develop in cystic form, usually in the liver (60–70%) and lungs, and ultimately cause symptoms due to local compression, as well as systemic symptoms of allergic type. Some less frequent localizations have been reported in the literature, such as the spleen, soft tissues, breast, heart, and extradural

space.[8–10] The striped muscle is a particularly rare localization, especially as the primary site.[9]

In fact, although the muscles account for 25% of the body mass and receive a large quantity of arterial blood, skeletal muscle infection by *E. granulosus* is estimated to account for less than 1–4% of cases of hydatidosis.[11,12] This low prevalence of muscular localization could be because the hepatic sinusoids and lung capillaries pose a physical barrier to dissemination of the cysts through the blood. Moreover, some authors suggest that the high concentration of lactic acid and the continual contraction–relaxation of the skeletal muscle masses could hinder implantation of the cysts.[11]

In any case, in the few reported cases of primary muscular hydatidosis, the most frequent localizations are the trunk[13] and upper limbs,[3,14,15] while the lower limbs are very rarely affected.[16–24] It has been hypothesized that implantation could occur by direct contact, following a dog bite, for instance. Another proposed hypothesis is that the liver or lung circulation might be bypassed by the creation of pre-capillary anastomoses at the pre- and post-parenchymal level.[25]

Few cases of giant hydatid cysts, i.e. cysts measuring more than 10 cm in diameter, have been described in the literature.[26] They have been reported in the lung, liver, abdominal cavity and spleen,[27–29] while at the muscular level they have been described in the diaphragm and heart.[30–32]

The clinical importance of large hydatid cysts is correlated to the risk of performing surgery before the infection has been correctly diagnosed. In fact, when there is rapid growth of a mass, compression of the adjacent organs may dictate emergency excision. In such cases there is a greater risk of dissemination of the disease, as well as of anaphylactic shock.[14] Thus, a careful clinical history needs to be taken to exclude any exposure to this parasite, particularly in endemic areas, as well as MRI, and it must be borne in mind that blood tests can yield false-negatives.[16]

In our case the diagnosis was made late since the patient had not consulted a doctor for some years, during which time the mass had grown to a gigantic size. Indeed, she must be considered lucky that no untoward event occurred, since she could very easily have accidentally hit the thigh, causing rupture of the cysts and subsequent dissemination and septic shock. When the patient finally came to our observation, the diagnosis was fairly simple, both because our area is known to be endemic and in view of the MRI features, showing the presence of many daughter cysts inside the mother cyst, with a low intensity signal on weighted T1 and T2.[1]

Our decision not to proceed with chemotherapy after surgical excision did not prove entirely justified. This subject remains controversial in the literature.[33] Many authors believe that complete removal is sufficient for a primary muscle localization, provided a correct preoperative diagnosis of hydatidosis has been made and the mass is completely eliminated, as in our case. However, other authors advise chemotherapy with mebendazole or albendazole for at least 3 months, in view of the high risk of development of a secondary lesion.[34] The great size of the mass we observed likely precluded total eradication, without an associated anthelmintic treatment, resulting in the secondary femoral bone involvement.

Due to the rarity of the disease and the difficulties in making an early diagnosis, bone hydatidosis often goes unnoticed. Failure to institute proper treatment, consisting of amputation and chemotherapy, can have severe consequences leading to sepsis and death.[35] In the literature, bone localization is reported to have a very low frequency (0.5–2%) and is generally secondary to contamination by contiguity or very rarely to spread through the bloodstream.[36,37] The sites most commonly affected are the vertebrae (50%), followed by the pelvis (25%) and long bones (15–25%).[38] Due to the slow growth of the cyst, there are often no local symptoms prior to the onset of a complication, such as a pathological fracture or paraplegia.[39] Even blood serum tests may remain negative for a long time and so the definitive diagnosis is often made on postoperative histological findings.[40] Radiography is useful in the advanced stages of the disease, when osteolytic lesions are evident, sometimes with the formation of internal lacunae or external fistulae.[41]

In the presented case, the secondary localization was diagnosed after the patient suffered a pathological fracture. At this stage, placement of a femoral prosthesis was necessary after resection of the infected bone tissue, and then prophylactic anthelmintic treatment was administered to prevent subsequent spread. Twenty months after surgery, the patient shows no recurrence and is still entirely functionally autonomous.

4. Conclusions

We have described a case of giant muscular hydatidosis localized in the medial region of the thigh. Considering the uncommon primary muscle localization and the huge size of the cyst, we made a search of the literature. We found that hydatidosis was only rarely reported with a primary muscular localization, while giant size was observed only at the level of the diaphragm or myocardium. The likelihood of onset of this infection is much higher in endemic areas like the Mediterranean Basin, and there is a risk that the cyst, if not promptly diagnosed, may grow to a very large size.

The aim of the present work was to underline the importance of including hydatidosis in the differential diagnosis of muscle masses, especially in endemic areas. On the basis of the clinical experience we describe, characterized by a recurrence in a bone segment near the primary site, we stress the need to associate exeresis with chemotherapy. In particular, this decision should be taken in cases of very large lesions for which the risk of spread to neighboring structures by proximity may be greater. In the case we describe, this choice might have obviated the need for the second surgical treatment, which was more demolitive than the first.

Conflict of interest: All authors declare no financial and personal relationships with other people or organizations that could inappropriately influence (bias) their work.

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