

Total femoral replacement in patient with complicated bone hydatid cyst.

Case report

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ABSTRACT

We report the case of a 31-year-old male patient presenting a femoral fracture whose complementary studies revealed an image consistent with a cystic lesion. The first biopsy study of this cyst erroneously reported a simple cyst. Osteosynthesis with intramedullary nailing was performed. The patient had a poor postoperative course associated with diaphyseal osteolytic images and loss of fracture reduction. The material was removed and intramedullary canal specimens were taken, which were positive for *Pseudomonas aeruginosa* and revealed remains of hydatid cysts through pathologic examination. The patient was referred to our center for the treatment of an infected pseudarthrosis secondary to a complicated hydatid bone cyst. A two-stage procedure was performed. The first stage involved the placement of an antibiotic-impregnated cement mega-spacer and the second stage involved the implantation of a total femur prosthesis as definitive treatment. The bone hydatidosis involves a challenging diagnosis, is very rare, and can cause severe dissemination. While there is no consensus or gold standard treatment for severe cases, two-stage total femoral replacement represents an encouraging option for limb preservation, showing satisfactory short- and medium-term outcomes.

Key words: Complicated hydatid cyst; hip mega-prosthesis; infected pseudarthrosis.

Level of Evidence: IV

Prótesis total de fémur en un paciente con quiste óseo hidatídico complicado. Reporte de un caso

RESUMEN

Se presenta un caso de un hombre de 31 años con fractura de fémur. Los estudios complementarios mostraron una imagen compatible con lesión quística. El primer estudio por biopsia de dicho quiste informó erróneamente un quiste simple. Se realizó una osteosíntesis con enclavado endomedular. El paciente tuvo una evolución tórpida asociada a imágenes líticas diafisarias y pérdida de la reducción de la fractura. Se retiró el material y se tomaron muestras del canal endomedular, cuyo cultivo reveló *Pseudomonas aeruginosa* y restos de quistes hidatídicos en la anatomía patológica. En nuestro centro, fue tratado por una pseudoartrosis infectada secundaria a un quiste óseo hidatídico complicado. Se le colocó un megaespaciador de cemento con antibiótico y, luego, una prótesis total de fémur como tratamiento definitivo. El compromiso óseo es muy raro y de difícil diagnóstico, y puede llevar a una diseminación severa. Si bien no hay consenso ni tratamiento de elección para los casos graves, el reemplazo femoral total en dos tiempos representa una opción alentadora para conservar el miembro, con resultados satisfactorios a corto y mediano plazo.

Palabras clave: Quiste hidatídico complicado; megaprótesis de cadera; pseudoartrosis infectada.

Nivel de Evidencia: IV

INTRODUCTION

Hydatidosis or cystic echinococcosis is a zoonotic infection caused by the larval stage of the tapeworm *Echinococcus granulosus*, and is responsible for significant morbidity and mortality in endemic areas from countries in the Mediterranean, Asia, Africa and Latin America.¹ The definitive hosts are domestic dogs and wild canidae, such as wolves and jackals, where the adult stages of the larva take place. Sheep are the most common intermediate hosts, but these tapeworms can also occur in goats, pigs, cattle, guanacos and other domestic livestock, where the

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larval stage (metacestode) takes place. Humans are accidental hosts in the normal life cycle of this parasite. Human infestation usually occurs in rural areas where livestock is raised, especially sheep and goats, with precarious hygienic and work conditions (no slaughter facilities, no water supply systems, no offal pit disposal, etc.), an inadequate understanding of the disease and a canine population lacking proper veterinary care.²

Bone hydatid cysts are rare, and the diagnosis is usually based on their complications, according to their localization and progression, such as neurological deficit due to spinal cord compression in cases of spine localization, pathologic fracture in long bones or fistulization and secondary infection in advanced stages with soft tissues involvement.^{3,4}

The purpose of our paper is to report a clinical case of complicated hydatid bone cyst of the femur, associated with osteomyelitis and skin and soft tissue infection, which was treated using a two-stage procedure for the implantation of a total femur mega-prosthesis and comprised a 2-year follow-up.

CLINICAL CASE REPORT

A 31-year-old male patient presenting a femoral fracture from a high fall sustained while engaging in agricultural activities in a rural environment, who was treated with intramedullary nailing. A previous examination of the needle biopsy of a cystic lesion found at fracture site through X-ray imaging, revealed a simple bone cyst.

Short-term postoperative course was not favorable: osteosynthesis failure associated with diffuse osteolytic and heterogeneous images at diaphyseal level (Figure 1).



Figure 1. Radiographic evolution of the first surgical intervention.

Fixation material was removed and an external fixation was performed. New bone and soft tissue specimens were culture-positive for *Pseudomonas aeruginosa*. The histopathological study revealed several fibro-hyaline membrane fragments consistent with the adventitial layer of a hydatid cyst, and no alive parasites were observed possibly due to the intramedullary nailing.

Soft tissue evolution remained poor, with active fistulas, and the external fixation was removed. The patient was then referred to our Center with a diagnosis of infected pseudarthrosis of the right femur associated with a complicated hydatid cyst.

New X-rays were taken along with a CT scan that revealed heterogeneous images consistent with femoral pandiaphysitis and pseudarthrosis in its proximal third. MRI revealed hypointense signal on T1-weighted images throughout the diaphysis, with a punctate pattern, wide cortical bone involvement, with preservation of condyles and femoral head, and hyperintense on T2-weighted of both bone and soft tissue images, consistent with the superinfection process (Figure 2).



Figure 2. Right thigh imaging studies.

The patient underwent surgical debridement and specimens were collected, of which from bone and soft tissue tested positive for *Pseudomonas aeruginosa*. Antibiotic chemotherapy was therefore instituted with piperacillin-tazobactam. In addition, a new histopathological study confirmed the presence of remains of hydatid cysts. Once the improvement of both the soft tissue function and the biochemical parameters was achieved, the patient underwent total femoral resection and placement of a modular, articulating, gentamicin-impregnated cement mega-spacer (Figure 3).

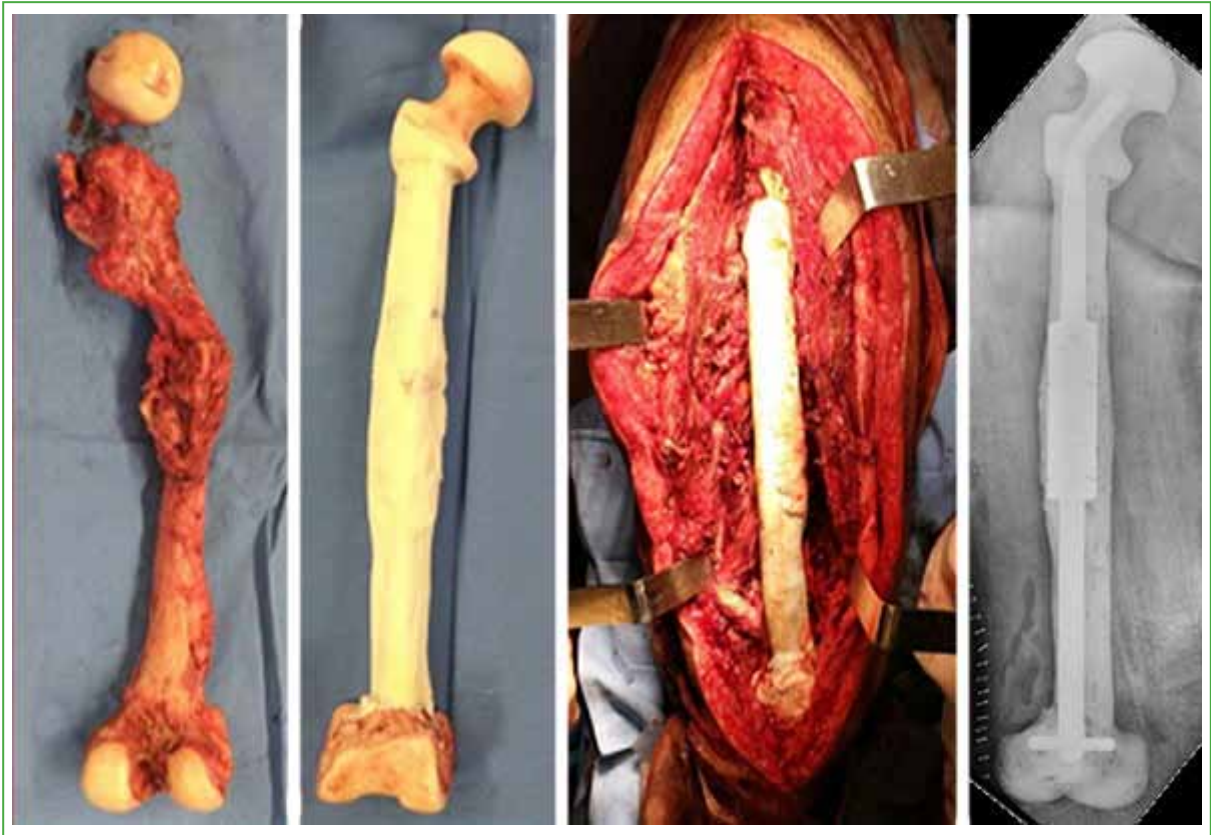


Figure 3. Femoral resection and placement of an antibiotic-impregnated cement mega-spacer.

Three months after this first procedure, the patient presented an adequate infection course which allowed him to undergo the definitive treatment: cement spacer removal and mega-prosthesis implantation within the total femur replacement, from an extended posterolateral approach up to the anterior tibial tuberosity and preserving the gluteal muscles from the metaphyseal component of the prosthesis (Figure 4). The patient received postoperative therapy with albendazole for 28 days. Within the following 7 months, the patient had normal inflammation parameters, no eosinophilia, and an adequate soft tissue evolution.

The patient was discharged with a healed surgical wound and a progressive postoperative physical therapy program.

Following the definitive surgery, both clinical and functional evolution was favorable. At one year after the procedure, the patient presented the following passive ranges of motion: right hip, 90° of flexion, 0° of extension, 25° of abduction, 15° of adduction, 10° of medial rotation, and 10° of lateral rotation; right knee, 90° of flexion and complete extension. The patient sustained no subsequent vascular or neurological impairment. Functional evolution was assessed using the Harris Score: 72.35% at 1-year follow-up, 80.50% at 2-year follow-up, and a 19% preoperative score. A marked functional recovery was observed, and the patient's satisfaction with treatment was high.

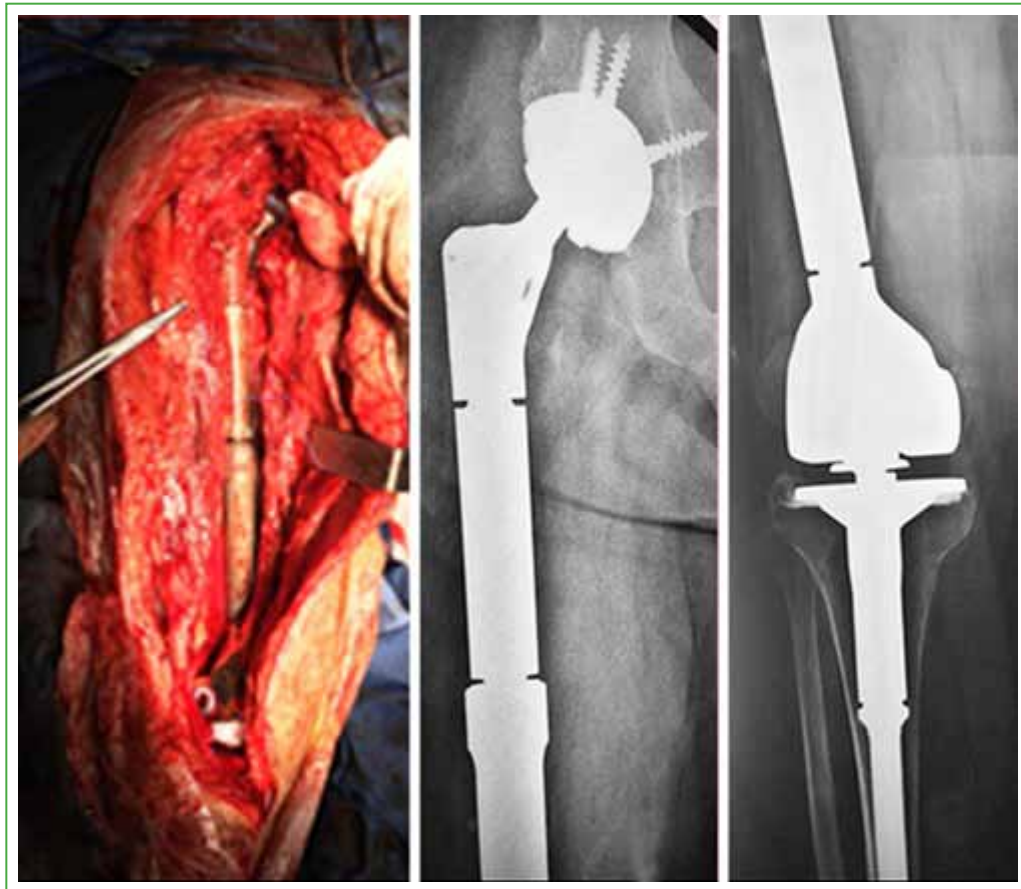


Figure 4. Extended posterolateral approach and postoperative radiographic control.

DISCUSSION

Hydatidosis is a major public health issue in Argentina due to these patients' medical care costs borne by the health system, involving patients that may require prolonged hospitalizations and may have their quality of life disrupted.²

Several strains of *Echinococcus granulosus* have been identified using criteria based on morphological, biological, biochemical, and epidemiological parameters. In South America, hydatidosis reaches endemic levels in Argentina, Chile, Uruguay, southern Brazil and the mountainous regions of Peru and Bolivia. *E. vogeli* and *E. oligarthrus* cases have been reported in Central America and western South America.⁵

Most cases of human hydatidosis are caused by the sheep strain and, to a lesser extent, by cervid and cattle strains. In 2010, the Argentinian National Health Monitoring System received reports of 385 hydatidosis cases.

Echinococcal transmission does not occur directly from one person to another nor from one intermediate host to another.² People become infected through ingestion of parasite eggs (excreted in dog feces), which is associated with contaminated hands and vegetables, pica and geophagy disorders, and close contact with host animals. Therefore, echinococcal infection is more common during childhood.

Once inside the human organism, the ovum releases the hexacanth embryos, which pass through the intestinal mucosa, enter the portal bloodstream and are carried to the liver, which acts as a first filter. Those who manage to pass the liver 'filter' are transported to the lungs (second most common location) and then through the systemic circulation to the rest of the organism, and may develop hydatid cysts.⁵

The most common locations of hydatid cysts are liver (67-89%) and lungs (10-15%); less frequent locations, spleen (1-3%), kidneys (1.5-2%), peritoneum (3-5%) and brain (2%); rare locations, pancreas, bone, heart, and orbits.⁵

Bone involvement is very rare and has an incidence of 1.4-3% of all cases.⁶⁻⁸ Cystic bone lesions may involve the spine (40-50%), long bones (25-30%) and the pelvis (15-20%), and only rarely the skull, sternum, scapulae, and phalanges.^{9,10} Bone dissemination occurs through three mechanisms:

- A) The cyst gradually compresses the adjacent tissues, eventually causing atrophy and necrosis.
- B) The cyst may obstruct nutrient vessels, resulting in bone ischemia and necrosis.
- C) Osteoclasts proliferate around the infectious focus of hydatidosis.

Any of these mechanisms or their combination may result in complications.²

In such a context, most patients with bone involvement present with non-specific symptoms or with bone hydatid cyst complications, including pathological fracture, neurological deficit (spine involvement) or superinfection with or without active fistulas.¹

Bone hydatidosis usually remains asymptomatic for long periods of time only to be diagnosed when patients are adults even if they had been infected while they were teenagers or children. For that reason, its diagnosis poses a challenge or may only be made on severe and disseminated presentations, and a not insignificant number is only diagnosed postoperatively.^{3,10}

Early diagnosis is key, as it allows for the institution of adequate patient management and treatment. Therefore, cystic tumors should raise the clinical suspicion of this disease.

Although there are multiple complementary studies, we relied on X-ray, CT and MRI imaging to assess the bone and soft tissue involvement since our patient presented with a concurrent severe infection affecting the entire diaphysis. We ordered a magnetic resonance angiography, although not a routine test, to assess the great vessels of the affected leg.

Laboratory studies included:

- Direct testing method: direct observation of fresh or formalin-fixed surgical specimens (sputum, urine, cerebrospinal fluid, daughter vesicles, scolices).
- Indirect or serological testing method:
 - Indirect hemagglutination
 - ELISA (IgG) and Western Blot: They have a higher diagnostic sensitivity than indirect hemagglutination and are the method of choice for the serological confirmation in suspicious and complicated cases.
 - Arc-5 test: this detection method is no longer used for the diagnosis of hydatidosis.⁵

Despite all these methods, a definitive diagnosis is established on the basis of histopathological examination of the needle biopsy and/or resected specimen, such as our case.¹¹

Differential diagnoses vary according to lesions characteristics and location and include: abscesses, simple cysts, hemangioma, tuberculosis among others.⁵

Albendazole is currently the benzimidazole of choice. Treatment response is affected by several factors, including age, cyst size and anatomical site. Albendazole is given at 10-15 mg/kg /day, twice daily, every 12 hours, following a fatty meal as it improves the drug absorption. Albendazole treatment should be given for at least 3 months and may be extended up to 6 months unless the patient experiences medication intolerance or has altered laboratory findings.² Praziquantel may be useful in case of spillage of cyst contents during surgery. Albendazole (10 mg/kg/day) and praziquantel (25 mg/kg/day) combination therapy given for 30 days before surgery, has been found to result in an increased number of nonviable protoscoleces as compared to albendazole or praziquantel treatment alone; however, more studies are warranted to assess the actual effectiveness of this combined treatment.²

Although there are currently no direct comparative studies between albendazole and surgery, chemotherapy stands as a second-choice treatment since its clinical outcomes include cyst disappearance in less than 50 % of patients, no cyst size reduction in at least 20-25 %, and an approximately 15-30 % recurrence rate during follow-up.¹²

Although there is no consensus on the type of resection and the need to adapt it to the different presentations, the surgical procedure of choice in cases of bone hydatidosis is complete resection of the lesion. However, most cases do not allow for this procedure as they involve diffuse and extensive lesions and such treatment would result in a mutilating surgery and severe functional sequelae. Despite the parasitic nature of the disease, some reports

suggest adhering to surgical procedures with an oncological criterium on account of their locally destructive behavior.¹⁰

The available literature is consistent on bone involvement being rare, and there is little material on severe femur involvement being treated with total femur arthroplasty.

Wirbel *et al.* suggest treating hydatid disease of the pelvis and the femur by means of radical resection and hip replacement with mega-prosthesis. Such treatment strategy may result in extended surgical procedures that are likely to involve infections at the surgical site.¹³

Natarajan *et al.* published three cases of hydatidosis of the femur treated by mega prosthesis replacement,¹⁴ as did Moore *et al.*, who, in 2015, published the first reported case of severe femoral involvement treated with a total femoral replacement.¹⁵

CONCLUSIONS

Reaching an accurate diagnosis is critical to avoid making poor decisions, such as the intramedullary reaming and nailing of our case, which may result in significant hydatid dissemination and a subsequent superinfection.

While there is no consensus or gold standard treatment for severe cases, total femoral replacement or mega-prosthesis continue to be an encouraging option for limb preservation, showing satisfactory short- and medium-term outcomes.

Conflict of interests: Authors claim they do not have any conflict of interests.

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