Cystic Hydatidosis of the Rib—Case Report and Review of the Literature

Andrei Roman, MD, 1 Carmen Georgiu, MD, PhD, 2 Dan Nicolau, MD, 3 Wissam Sabha, MD, 3 Miruna Surariu, MD, 4 and Dorel Precup, MD, PhD 3, 5

The hydatid disease is a zoonosis endemic to rural countries, such as those in the Mediterranean region, South America, North Africa, Central Asia and China. Hydatid cysts commonly affect liver and lungs, but less than 100 cases of costal hydatidosis have been reported in the literature. While diagnosis of the disease in commonly affected organs is relatively easy, uncommon locations can prove to be challenging as is the case with costal hydatidosis. Imaging techniques can suggest the diagnosis, but sometimes it remains uncertain until surgery. The treatment is surgical, assisted by long-time Albendazole chemotherapy. We present a rare case of costal hydatidosis, the first one to be reported in Romania according to our review of the literature.

Keywords: hydatidosis, rib, Echinococcus granulosus, surgery, chest wall

Introduction

Echinococcosis is an anthropozoonosis caused most frequently by Echinococcus granulosus, humans being accidental intermediate hosts. The disease is endemic to countries with significant rural populations. Liver and lungs are the most commonly affected organs, bone hydatid cysts are rare, but less than 100 cases of costal hydatidosis have been reported in the literature. 1, 2 We describe a recently diagnosed case of costal hydatidosis, the first one to be reported in Romania according to our review of the literature.

Case Report

A 25-year old male shepherd was admitted to the Emergency Unit with left-sided chest pain following a moderate trauma. A chest radiograph revealed a 9,5/7 cm mass belonging to the left chest wall. The computed tomography (CT) scan (Fig. 1) showed a multilocular mass with cysts sized up to 3 cm located on the posterior arch of the eighth rib, associated with significant deformation and complete destruction of the internal compact layer of the rib (Fig. 2). No pleural effusions, pulmonary parenchymal involvement, or other thoraco-abdominal masses were visible. The radiological appearance raised the suspicion of costal hydatidosis, requiring surgical confirmation and treatment.

The patient was admitted to the thoracic surgery ward. After preliminary preparation of the patient, a left latero-posterior thoracotomy along the long axis of the mass was performed, with diagnostic and eventually curative purpose. The macroscopic intraoperative aspect was of a multilocular osseous hydatid cyst measuring 13 cm on the long axis, located along the internal face of the eighth left rib without exceeding the extrapleural space. The proliger membrane and numerous daughter cysts were removed, a partial resection of the 8th left rib and debridement of the pericystic tissue being necessary (Fig. 3). Approximately 500 ml 1% formalin were applied for 8 min on the wound with scolicidal purpose.
The histopathological examination revealed fragments of the proliger membrane, daughter vesicles, protoscolices, multinucleated giant cells, inflammatory infiltrate and trabeculae of spongy bone tissue. The histological appearance confirmed the diagnosis of hydatid disease. Postoperative treatment included antiparasitic chemotherapy with Albendazole tablets 400 mg × 2/day. The drug was administered in three one-month cycles with 14 days pause between cycles. Postoperatively, the patient had normal respiratory and cardiovascular functions, any signs of late-onset allergic reactions were absent. Moderate pain in the area of the wound was present, but subsided after 2 days and the wound healed without complications. The chest X-ray upon discharge showed expanded lungs, no pleural effusions, partial absence of the eighth rib and a complete absence of the hydatid cyst. One month after surgery the patient was asymptomatic, with no signs of recurrence on the X-ray. The patient is currently being monitored by the family doctor with radiological surveillance every 6 months.

Discussion

The hydatid disease is one of the most important zoonoses found in Romania. It is a parasitic disease caused most frequently by Echinococcus granulosus, a flatworm. Because of the high number of reported cases in humans and animals alike, Romania is considered to be an endemic country. In 2011, 53 cases have been reported (0.25/100,000), placing Romania on the 5th place in the European Union with an average of 0.18/100,000 reported cases. 3)

The parasite requires an intermediary host, most commonly sheep, cattle, swine, as well as a primary carnivorous host like dogs, foxes, wolves. Primary hosts spread the parasite’s eggs through their feces. Humans become accidental intermediary hosts by ingesting food or water containing the eggs. After being ingested, the eggs hatch in the intestine, releasing oncospheres which pass through the intestinal wall and are transported to the liver, which is the most common affected organ. 5) By passing the liver (52%–77% of cases of hydatid cysts) and entering into the systemic blood flow, the oncospheres can stop in almost any organ, particularly in the lungs (10%–40% of cases of hydatid cysts). 5–8) Bone hydatid cysts are very rare, between 0.5 and 2.5%, involving frequently the spine (44.2%), long bones (30%) and pelvis (16%). 9, 10) Involvement of the ribs is extremely rare, less than 100 cases having been reported in the literature,1) none of which from Romania according to our review of the databases available online.

Costal hydatid cysts can be primary or secondary. Secondary hydatid cysts occur because of a spontaneous or intraoperative rupture of a pulmonary or mediastinal hydatid cyst. 1) Due to the fact that other sites of involvement have not been discovered, we assume that our patient has a primary hydatid cyst of the rib.
The diagnosis of costal hydatid cysts is made by clinical, serological, imaging and surgical means.

Costal hydatid cysts can be asymptomatic and their discovery incidental. Sometimes mild, intermittent chronic chest pain, tumefaction of the area involved, a cutaneous fistula, or an intercostal neuralgia can be signs of the disease.\(^1\)\(^3\)\(^9\) In our case, the pain occurred relatively recently and was attributed to a chest trauma.

Following laboratory tests are used in the diagnosis of hydatid disease: eosinophil count, total and specific immunoglobulin E (IgE), enzyme-linked immunosorbent assay (ELISA), Western Blot, immunoprecipitation, indirect hemaglutination test (IHT). A negative result cannot exclude the diagnosis.\(^1\)\(^11\) The highest sensitivity and specificity obtained for rib hydatidosis were 82.7% (ELISA), respectively 94.7% (immunoprecipitation).\(^1\)\(^2\)

The laboratory tests are often negative for osseous uncomplicated hydatidosis, while rupture of the cysts brings higher serological detection rates.\(^1\)\(^13\)\(^15\) Taking these facts and the additional costs into consideration, we decided not to perform preoperative serological examinations.

Due to the slow progress of the disease and the lack of symptoms, imaging detection of bone echinococcosis is usually achieved by a fortuitous radiograph, usually in advanced stages. Radiographic appearance is nonspecific, consisting of a cystic or irregular bone lysis. In some cases, arcuate calcifications of the cyst wall or multicystic osseous lysis can be present.\(^1\)\(^4\)\(^14\) Although a Computer Tomography examination brings important additional information regarding the location, severity of bone destruction and expansion in the surrounding soft tissues, it remains a nonspecific technique.\(^1\)\(^5\)

The most common findings are multiple cystic images of various sizes with fluid density (10–20 UH) associated with different degrees of osteolysis that can lead to the thinning and eventually rupture of the compact bone.\(^9\) Specific, but rare findings are those of round or ovalary lesions “with a double layered arcuate calcification” and/or containing detached and folded endocysts.\(^1\)\(^4\) The history and CT aspect of our patient strongly suggested the diagnosis, so no other imaging techniques were required.

Magnetic resonance imaging (MRI) is the most accurate imaging technique for osseous hydatidosis. Generally, T1 and T2 weighted images are sufficient for diagnosis, without the need for gadolinium administration.\(^9\) The cysts usually appear as hyposignal areas on T1-weighted images and as hypersignal areas on T2-weighted images.\(^9\)\(^13\) On T1-weighted images, the intensity of the signal belonging to the parent cysts is similar to that of muscle, while the signal belonging to the daughter cysts is similar to that of water. In case of rupture or superinfection of the cyst, the signal is amplified due to increased protein content.\(^1\)\(^4\)

If the presence of a costal hydatid cyst is suspected, ultrasound or CT screening of the liver and spleen is recommended. In our case, the initial CT scan allowed the visualization of lung, liver and spleen, so involvement of these organs could be excluded from the beginning. Imaging investigations can be extended to other organs if necessary.\(^1\)\(^3\) As no other symptoms were present, we decided not to pursue further screening investigations.

The imaging differential diagnosis should include: (1) malignant bone tumors, especially plasmacytoma and metastases (2) benign tumors such as aneurysmal cysts, neurofibromas or giant cell tumors (3) osteomyelitis caused by Mycobacterium tuberculosis or common germs.\(^1\)\(^5\)\(^13\)\(^15\)\(^16\)

Despite advanced laboratory and imaging techniques, in some cases a precise diagnosis cannot be established until surgery.\(^1\)\(^3\)\(^17\)

Costal hydatid cysts can generate multiple complications. Local expansion may cause pathologic fractures, spinal cord compression, thoracic aperture syndrome, pulmonary atelectasis. Cyst rupture can lead to parasitic dissemination into neighboring organs and to anaphylactic shock.\(^2\)\(^1\)\(^4\)\(^17\)\(^18\)

The purpose of treatment in costal hydatidosis is to eliminate all parasitic and pericystic tissues, a similar approach to that of malignant tumors being necessary.\(^2\)

Compared to the surgical treatment of osseous hydatid cysts located elsewhere than the ribs, the treatment of costal hydatidosis has the advantage that all of the affected bone tissue can easily be removed with little functional consequences for the patient. Wide excision of the affected tissues, resection and debridement of pericystic tissues are required.\(^1\)\(^1\)\(^7\)

We performed a partial costectomy with a margin of approximately 2 cm from the healthy rib, but in some cases a total resection of the rib may be necessary. In order to avoid parasitic dissemination in case of an intraoperative rupture of the cysts, scolicidal agents like hypertonic sodium chloride or formalinized solutions (as used in our case) should be applied on the wound.\(^1\)\(^1\)\(^8\)

Despite the known side effects of formalin, we decided to use it because of the surgical team’s experience. After a complete resection of the affected tissues and intraoperative usage of scolicidal agents, complete elimination of microscopic daughter cysts is not always possible.\(^1\)\(^1\)\(^9\)

There is an increased risk of relapse requiring antiparasitic chemotherapy.

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Treatment of choice is Albendazole, a benzimidazole indicated pre- and postoperatively in high doses (10–15 mg/kg/day) for up to 6–9 months after surgery. The postoperative clinical examination has to exclude any signs of late-onset allergic anaphylaxis. A postoperative X-ray is also recommended in order to detect a pneumothorax or a pleural effusion which could raise suspicion of dissemination. Regular checkups are required to detect any relapses.1,13,14

Conclusion

Although costal hydatidosis is very rare, it must be part of the differential diagnosis of costal osteolytic tumors, especially in endemic countries. Imaging techniques can usually suggest the diagnosis, but sometimes it remains uncertain until surgery. The treatment is surgical, assisted by long-time Albendazole chemotherapy.

Disclosure statement

The authors have no conflicts of interest.

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