Liver Hydatidosis Extended to Cava and Lungs

A 21-year-old man was referred to our clinic for evaluation of a two-day evolution of hemoptysis and cough with expulsion of whitish membranous material. He had thoracic pain and shortness of breath for three months. His physical examination was unremarkable. Chest radiographs performed in the Emergency room (ER) showed no abnormalities. Thoracic computed tomography (CT) performed 72 hours later revealed a heterogeneous liver cyst, 7 cm in diameter, multiloculated, with floating membranes [Figure 1, long black arrow] that expanded into the inferior cava vein [Figure 1, short black arrow] and into the right atrium of the heart [Figure 2 white arrow] with multiple pulmonary embolisms [Figures 1 and 2, white arrowhead]. Cytology of the sputum revealed membranes of *Echinococcus granulosus*. The serology (ELISA) for hydatidosis was strongly positive. Diagnosis of liver hydatidosis extending to the cava, heart, and lungs was made.

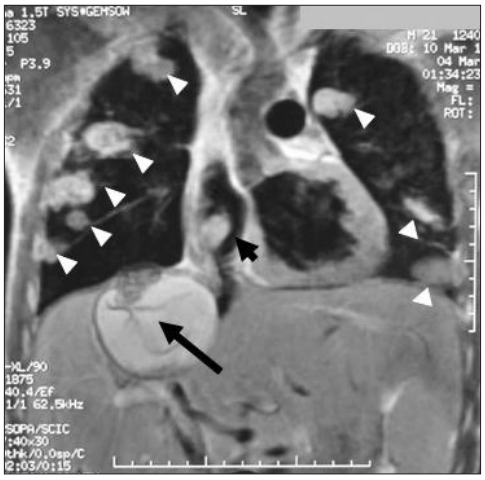

Figure 1: Thoracic computed tomography showed a cyst, 7 cm in diameter, with floating membranes (long black arrow), expanding into the inferior vena cava (short black arrowhead), as well as multiple pulmonary embolisms (white arrowhead)

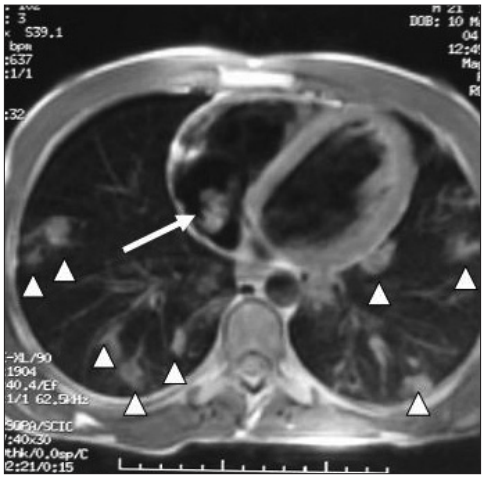

Figure 2: Thoracic computed tomography showed a cyst in the right atrium of the heart that came from the inferior vena cava (long white arrow), as well as multiple pulmonary embolisms (white arrowhead)

Hydatid disease is a zoonotic infection caused by the larval stage of the tapeworm *Echinococcus granulosus,* and is endemic in Europe, including the Mediterranean area and the Balcanic nations, North and East Africa, India, China, Indonesia, and the Southern Cone of the Americas.[1]

Hydatid cysts may be found in almost any site of the body, but the liver (60–70%) and lungs (10–15%) are the most frequently affected.[2] The heart is reported to be involved in less than 2% of the cases of cystic echinococcosis (CE), and only a few cases have been reported. Endovascular extracardiac localization of hydatidosis is even more rare and these are thought to be secondary to the rupture of the primary germinative membrane located at other sites, mostly in the heart or vena cava, with consequent embolization to the pulmonary or systemic circulation.[3]

Cardiovascular hydatid disease is associated with a high risk of potentially lethal complications. The clinical picture and complications vary according to location, size, and disturbance of the normal cardiac function. Unless a cyst is located in a critical anatomic site, the patients can remain asymptomatic for years, although they are under a continuous threat of rupture. Signs and symptoms are extremely variable, although chest pain is the most frequent. Nevertheless, patients may present with life-threatening conditions such as hemoptysis or pulmonary hypertension.

Conventional chest radiography and serological testing for antibodies has been the mainstay of the diagnosis. However, the two, even in combination, may not yield a definitive diagnosis, as radiological cysts can mimic other pathologies. Serological testing yields false-positive or false-negative results, adding to the confusion. Nowadays, CT and magnetic resonance imaging (MRI) are useful additional imaging techniques to confirm the diagnosis and detect complications (especially in the case of hydatid pulmonary embolism).[4]

A therapeutic approach may be complicated due to the heterogeneity of cardiovascular CE, and may require a combination of medical and surgical treatment.[5] Benzimidazoles (albendazole and mebendazole), with or without praziquantel, are the basis of medical treatment, although the dose and duration of treatment have not been clearly established.[6] There is no evidence of what to do with pulmonary embolisms in CE: The surgical approach

is very complex and medical treatment is not effective (as antiparasitic drugs do not reach these locations as well as they are supposed to), hence, an early surgical approach, before the pulmonary cyst embolism appears, is mandatory. However, when a cyst embolism is established, indefinite antihelminthic treatment is necessary.

In the patient reported, a complete surgical resection of the cava and atrium cysts, using cardiopulmonary bypass, was conducted. A midline sternotomy was performed, and cardiopulmonary bypass was instituted with bicaval canulation. Standard myocardial protection with cold-cardioplegic cardiac arrest was achieved. In order to avert embolisms and possible introduction of free scolices to other cardiac structures the cava was cross-clamped. The cyst cavity was carefully opened, the cyst contents were removed, and the remaining germinative membrane was deleted. After the complete removal of the cyst, the remaining pericyst cavity was washed with hypertonic saline solution. Minimal manipulation of the heart occurred during the surgical procedure. The postoperative course of cardiac surgery was uneventful, and five months later, he underwent a pericystectomy in the liver with capitonnage (obliteration of the cyst). No thoracic surgery was performed, as the hydatid cysts were endovascular. After surgery, long-term chemotherapy with albendazole plus praziquantel for 45 months was prescribed, but multiple embolizations were needed due to recurrent hemoptysis. Due to the poor outcome, nitazoxanide was combined with albendazole for 18 months. Clinical and radiological improvement was achieved, with improvement of cough, hemoptysis, and shortness of breath. A new CT scan performed at that time showed disappearance of the liver, caval, and atrium cysts, and size reduction of the endovascular pulmonary cysts.

Marta Díaz-Menéndez, Rogelio López-Vélez

Tropical Medicine and Clinical Parasitology, Infectious Diseases Department, Ramón y Cajal Hospital, Carretera de Colmenar km. 9,1, Madrid 28034, Spain

Address for correspondence:
Dr. Marta Díaz-Menéndez,
E-mail: marta.diazmenendez@gmail.com

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