Surgical treatment of pulmonary hypertension caused by echinococcosis disease

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Echinococcosis is a parasitic zoonosis caused by the dog tapeworm *Echinococcus* and its larval stage, the hydatid cyst. The vast majority of cysts occur in the liver, lungs, or both.¹ Arterial involvement is usually seen after cardiac hydatid cyst rupture. Hydatid cyst within the pulmonary arteries is an exceptional localization, usually a consequence of embolism from primary cardiac locations or intraoperative rupture of a primitive liver or lung hydatid lesion.² We present the case of a patient with bilateral intra-arterial pulmonary involvement leading to pulmonary hypertension.

CLINICAL SUMMARY

A 33-year-old woman had a 3-month history of dyspnea. The patient had a history of disseminated cardiothoracic echinococcosis and had undergone bilateral lung cyst resection twice, 7 and 6 years before the index visit. A right atrial cyst had also been removed 3 years previously. Throughout the years, the patient had received long-term antiparasitic therapy with albendazole.

One month after the cardiac surgery, the patient had symptomatic severe pulmonary hypertension develop. Pulmonary angiography confirmed stenosis of the right interlobar artery, which was successfully treated with a stent implantation. One year before her admission, there was recurrence of severe pulmonary arterial hypertension. Contrast-enhanced computed tomographic scan displayed complete occlusion of the right upper posterior segmental artery and bilateral interlobar arteries, without signs of extrinsic compression. Ventilation-perfusion lung scan showed severe mismatch. Pulmonary angiography revealed right interlobar pulmonary artery stent occlusion, posterior right upper segmental artery occlusion, and absence of pulmonary vessels on the left lower lung (Figure 1). On right heart catheterization, mean pulmonary arterial pressure was 55 mm Hg, and pulmonary vascular resistance was 545 dyne/(s · cm⁻⁵).

FIGURE 1. Pulmonary angiogram (see text for description).

The patient underwent pulmonary endarterectomy with standard cardiopulmonary bypass, deep hypothermia (20°C), and intermittent circulatory arrest. Surrounding tissues were initially protected from spreading of the hydatid cysts with wet sponges soaked with hypertonic saline solution, and the main right pulmonary artery was incised. Loose thrombus and hydatid vesicles were removed from endothelium. Two intervals of circulatory arrest (20 minutes each) were needed to remove the right interlobar pulmonary stent and perform a complete endarterectomy of the right lower lobe arteries. The entire pulmonary tree was irrigated with normal saline solution in an effort to kill any potentially unresected distal vessel located viable *Echinococcus*. Right arteriotomy was closed with a 6-0 polypropylene running suture. A left-side incision was then performed with a single arrest (16 minutes) and the same approach. Inotropic support and nitric oxide was required during cardiopulmonary bypass weaning and the early postoperative period. Immediately after the surgery, mean pulmonary arterial pressure was reduced to 37 mm Hg and pulmonary vascular resistance to 256 dyne/(s · cm⁻⁵). Postoperative stay was uneventful, and the patient was discharged on the 12th postoperative day. Histologic examination confirmed the diagnosis of hydatid cyst caused by *Echinococcus granulosus* (Figure 2). Lifelong antiparasitic therapy with

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FIGURE 2. A, Specimen show extensive disease with the stent in place on the right side (arrow). B, Hematoxylin and eosin–stained specimen section showing the pulmonary arterial wall with thrombotic material and wall of the hydatid cyst (100× original magnification). Note the protoscoleces hook (inset, arrow; 400× original magnification).

albendazole plus praziquantel was administered to diminish recurrence risk.

DISCUSSION

Echinococcosis is a parasitic zoonosis caused by the dog tapeworm *Echinococcus* and its larval stage, the hydatid cyst. This disease has a worldwide geographic distribution. Although approximately 60% of all cysts remain asymptomatic, they can become symptomatic as the result of mass effect or rupture, the latter with life-threatening consequences. The vast majority of cysts occur in the liver, lungs, or both.¹ Arterial involvement of echinococcosis usually results after cardiac hydatid cyst rupture, and embolism of the germinative membrane causes acute symptoms. Hydatid cyst within pulmonary arteries is an exceptional localization and most frequently a consequence of embolism from primary cardiac locations or a result of intraoperative rupture of a primitive liver or lung hydatid lesion (secondary echinococcosis).² This patient had bilateral intra-arterial pulmonary involvement, so most probably she had germinative membrane embolization, intravascular growth of hydatid cyst, and subsequent chronic thrombosis.

The presence of a cystlike mass in a person with a history of exposure to an endemic area suggests hydatid disease even in the absence of previous history of parasitic infection. Imaging techniques, such as computed tomographic scans, ultrasonography, and magnetic resonance imaging are used to detect cysts.¹ Specific confirmation can be obtained either from demonstrating echinococcal antigens by immunodiffusion procedures or from immunoblot assays.¹

Pulmonary arterial involvement with hydatid cyst requiring surgical treatment is uncommon. Previous reports in patients with acute pulmonary embolism treated with embolectomy and suction suggest that they usually do poorly.³ On the other hand, chronic unilateral pulmonary artery obstruction can be successfully treated with arteriotomy and cyst removal with or without pneumonectomy or with pulmonary artery endarterectomy.^{4,5} Bilateral pulmonary arterial disease requires a different approach, because resection is not feasible. Surgical steps to treat bilateral lung hydatidosis mirror pulmonary artery endarterectomy for chronic thromboembolic pulmonary hypertension, but special care is first needed to remove hydatid vesicles attached to the vascular endothelium before starting an endarterectomy. The endarterectomy material is smooth but fragile, and special care is needed to avoid fracture and its consequent spreading containing viable *Echinococcus*. Surgery remains the most effective treatment to remove pulmonary artery hydatid cysts, but aggressive medical treatment is needed to avoid recurrence.

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