Cardiac echinococcosis causing unexpected death. A case report

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Abstract: Echinococcus is endemic in some parts of the world but cardiac involvement with hydatid cyst is rare. Here, we report a 50 year-old man having at presentation a history of atypical chest pain. During routine clinical evaluation abnormal findings in ECG were found. In echocardiography, a large mass was detected in the anterior part of the interventricular septum. The patient died shortly after admission by anaphylactic shock, secondary to hydatid cyst endocavitary rupture.

Key words: Cardiac echinococcosis, Hydatid cyst, Anaphylaxis

Echinococcus is endemic in sheep-raising areas of the world, particularly in Argentina, New Zealand, Greece and Australia [1, 2]. The usual host of Echinococcus granulosus is dog. Sheep is the usual intermediate host, but occasionally, human may serve as an intermediate host if they accidentally ingest the tapeworm ova [3]. Cardiac involvement with hydatid cyst is rare. The myocardium of the interventricular septum or left ventricular free wall is the usual site for cardiac cyst involvement, but occasionally right atrium or ventricle may be involved, as well [4]. Cardiac cysts are usually associated with fatal complications [5].

A myocardial cyst may degenerate and calcify or rupture into pericardium or heart chambers. Rupture of the cyst is the most serious complication, inducing either acute or advanced chronic constrictive pericarditis, or systemic or pulmonary emboli [6, 7]. The passing of the hydatid fluid into the circulation may also produce fatal circulatory collapse in response to anaphylactic reactions to protein constituents of the fluid [1].

Few patients infected with cardiac hydatid cyst may not have any obvious clinical symptoms [1,2]. However, in patients with left ventricular involvement, ECG may show ischemic changes in T wave, while the involvement of the interventricular septum may be associated with atrioventricular conduction defects, or right bundle branch blocks [8,9].

The ischemic changes are due to compression or complete obstruction of the coronary arteries present in the area of the cyst [10]. Therefore, a complete cardiac evaluation should routinely be performed in all patients with cardiac echinococcosis, including coronarography.

Case report

A 50 year-old man, from a rural zone, sheep-raising area, was admitted in hospital for evaluation of an atypical anterior chest pain by only 4 weeks. The lab tests showed a moderate...
inflammatory syndrome and lack of eosinophilia. Ischemic changes in T wave were found on ECG recording. The echocardiography has evidentiated a moderate cardiomegaly.

CT scan showed the presence of a septal cystic mass. The patient died at 6 days from hospital admission through a progressive cardiovascular collapse.

Fig. 1. The internal germinative layer with daughter cysts (HE X20)

Fig. 2. Pericystic chronic inflammatory infiltrate (HE X20)

Fig. 3. Pericystic chronic inflammatory infiltrate and fibrosis (VG X20)

Fig. 4. Pulmonary emboli with anhisto membranes (VG X20)

In the macroscopic examination of the heart we detected a cystic lesion of 6 cm in diameter located at the level of antero-inferior part of the interventricular septum. The cyst was typically composed of a stratified wall: fibrous adventicea, an anhisto and proligere membranes.

Grossly, we also identified two openings: (a) one more evident, oval in shape, of about 0.5 cm in diameter, connecting the cyst with left ventricular cavity; we found the presence of small daughter cysts into ascending aorta, as well, confirming the systemic embolism; (b) the other opening, was an
irregular right ventricular endocardial fissure, of about 0.3 cm length, permitting the right ventricular entrance of the hydatid fluid, and in this way in the pulmonary circulation; this appearance was confirmed histologically, by the presence of anhiste membrane emboli into peripheral pulmonary vessels.

In the microscopic examination of the heart, the internal germinative layer with daughter cysts (fig. 1), the external laminated layer, and the cystic fibrotic wall was detected. We also revealed an adjacent myocardial area presenting a moderate inflammatory cellular infiltration, mainly composed from eosinophils, lymphocytes and plasma cells (fig. 2), myocyte atrophy and diffuse interstitial fibrosis, as signs of chronic myocardial ischemia (fig. 3). On the microscopic analysis of the lung sections, the presence of multiple emboli into small peripheral pulmonary vessels was detected, represented by fragments of anhiste membranes filling the peripheral pulmonary arterioles (fig. 4).

No other gross or microscopical pathological lesions were detected in the chest or abdominal organs in the systematic examination.

The cause of death in this case was referred as an anaphylactic shock due to intravascular spread of the hydatid cyst content.

Discussion

Hydatid disease is a parasitic infestation caused by a tapeworm. Echinococcus is endemic in many parts of the world, but cardiac involvement with hydatid cyst is uncommon, occurring in less than 2% of cases [1]. Cardiac echinococcosis is mostly symptomatic. These symptoms include angina because of the pressure effects of the cyst on the coronary arteries, dyspnea, and palpitation [8, 10].

Morbidity is usually secondary to free rupture of the echinococcal cyst associated with anaphylaxis or dysfunction of the affected organs. So, after a progressive enlargement, hydatid cyst may rupture, usually happening after trauma or during interventions, but spontaneous form is much less frequent. Mortality rates of asymptomatic cases of cardiac cyst involvement, following perforation of the cyst, are relatively high, because of acute anaphylactic reaction and cardiogenic shock, which may occur in these patients [1, 2].

In the past decades, the treatment of hydatid disease has been limited to surgical excision [11]. Nonetheless, new experiences suggest that benzimidazole derivatives are useful drugs for the treatment of hydatid cyst [12].

Despite the availability of valuable medical treatments, the surgical excision is generally recommended for both symptomatic and asymptomatic patients. This is because of the high risk of rupture of the cyst and its serious consequences. So, the surgically cyst removal can prevent the systemic emboli and fatal circulatory collapse, which may occur in such cases [11].

Hydatid disease is a dangerous condition because of risk of complications. We commented the death as the result of anaphylactic phenomena following the rupture of echinococcal cyst, by intravascular spread of cyst content.
References