

Acute arterial embolism as the clinical presentation of a disseminated hydatidosis: case report

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ABSTRACT

Hydatidosis is a parasitic infection caused by the *Echinococcus granulosus* larvae, transmitted by the ingestion of infected food, characterized by the formation of cysts in vital organs. In Argentina, the disease is spread throughout the territory, constituting an important public health issue.

The disease usually affects the liver and lungs. The affection of other organs is rare, and even more uncommon the affection of the heart.

We present the case of a disseminated hydatidosis in a pediatric patient, whose initial clinical manifestation was an acute arterial embolism of the right limb caused by the rupture of a hydatidic cardiac cyst.

Key words: Thrombosis and embolism, echinococcosis.

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INTRODUCTION

Hydatidosis is a parasitic infection caused by the *Echinococcus granulosus* larvae, transmitted by the ingestion of infected food, and it is characterized by the formation of cysts in different organs, such as liver, lungs, etc.

Echinococcus granulosus infection shows a universal geographic distribution, with

description of cases in every continent. In South America, it exists in most countries, but in Argentina, Bolivia, Brazil, Peru and Uruguay, it is prevalent and constitutes an important public health issue.¹

In Argentina, cases are spread all along the national territory, with its highest prevalence in rural areas, especially in areas of sheep and goat breeding. This parasite can be found in approximately 30% of the national territory. According to the 2017 integrated epidemiologic bulletin of surveillance, in Argentina, during the 2016-2017 period there were 234 notified cases and 116 confirmed cases. Provinces with the highest number of confirmed cases were Buenos Aires, Córdoba, Entre Ríos and Santa Fe.²

In humans, hepatic involvement happens in 67-89% of cases, and pulmonary involvement in 10-15% of cases. Localization in other organs accounts for less than 10% of reported cases. Cardiac compromise is infrequent (0.05-2%) and the rupture of a cardiac hydatid cyst, with subsequent dissemination in different organs, has been rarely reported. In the present report, a pediatric patient with disseminated hydatidosis with acute arterial embolism is described.¹

CASE REPORT

A 15-year-old male patient, native of the Carmen de Areco locality, Province of Buenos Aires, whose family lived in a rural area, was admitted to the hospital for epigastric pain, followed by paresthesia in the right lower limb with less than 24 hours of evolution. Pain had been increasing and a change in color of the entire right lower limb was added, also associated with severe headache. He consulted at a zonal hospital, where an acute arterial thrombosis was suspected. Single dose of intravenous heparin and hydrocortisone were administered, and referral to a center with greater complexity was made.

The family lived in a field in their locality, where they raised and ate their own animals, giving the remains to the dogs of the field.

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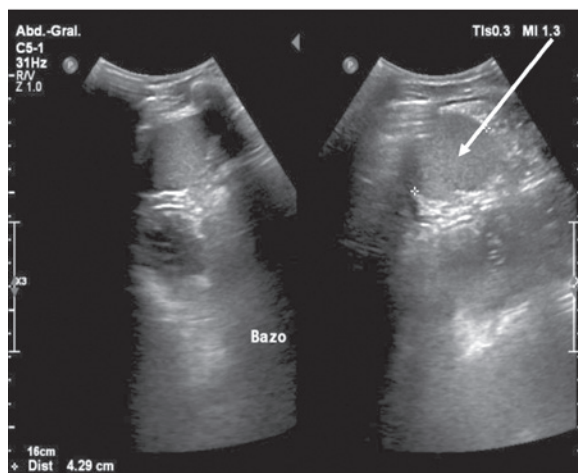
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At his admission in the Emergency Department of "Prof. Dr. Juan P. Garrahan" Hospital, the patient was somnolent, hemodynamically stable and eupneic. At physical examination, he had normal heart sounds, no murmurs or gallops. He had normal respiratory auscultation, soft nonpainful abdomen, and no hepatosplenomegaly. His right lower limb was cold and pale, and presented absent femoral, popliteal, posterior tibial and dorsal pedal pulses.

Diagnosis of acute arterial embolism was suspected. The next complementary exams were performed:

- Vascular doppler ultrasonography: an echogenic endoluminal image was observed in right common femoral artery and deep femoral artery through its proximal end.
- Abdominal ultrasonography: cystic image in the superior spleen pole, with an 11*10*10 cm. diameter. Normal liver. Preserved aortic flow. (Figure 1).
- Brain scan: without abnormalities.
- Blood tests: neutrophilic leukocytosis (WBC 14050/mm³) (neutrophils = 13340/mm³), lymphopenia (lymphocytes= 453/mm³), and normal liver and renal function. Negative HIV serologic testing.
- Echocardiogram: a 3-cm cystic image was observed inside the left ventricle (LV). There seems to be a communication between the interior of the cyst and the LV cavity. Preserved LV function, no structural congenital defects and no pericardial effusion (Figure 2).

FIGURE 1. Abdominal and renal ultrasonography



Cystic image at the level of the upper pole of the spleen (arrow).

With such image and report compatible with cardiac hydatidosis, complementary studying was completed with a chest x-ray, with no abnormalities. ELISA for hydatidosis disease was performed, with negative results.

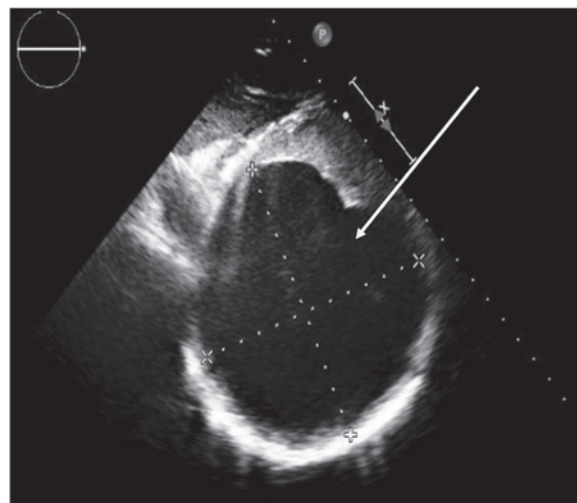
Twenty four hours after admission, longitudinal arteriotomy of the common femoral artery was performed and, according to the operation note, a 10 cm long material compatible with hydatidosis was extracted (Figure 3). At this opportunity, albendazol administration was initiated to prevent further hydatid dissemination.

Since there were no hemodynamic compromise, cardiovascular surgery staff decided not to perform cardiac exeresis at admission.

With the compatible pathology sample, hydatidosis diagnosis was confirmed. Because of the severity of the case, praziquantel was added to the antibiotic regimen (load dose for 5 days, then weekly administration). At 21 days of treatment, with good clinical evolution, surgery to extract cardiac and spleen cysts was performed.

Procedure lasted six and a half hours, requiring an hour of extracorporeal circulation. Left ventriculotomy and cyst extraction, which was attached to the posterior leaflet of the mitral valve, was done. During surgery patient suffered ventricular fibrillation for 5 minutes, which required defibrillation and lidocaine drip. Superior hemisplenectomy was also performed. Samples for pathology and cultures were sent. Samples showed the presence of the cuticular wall of a hydatid cyst (Figure 4).

FIGURE 2. Doppler echocardiogram

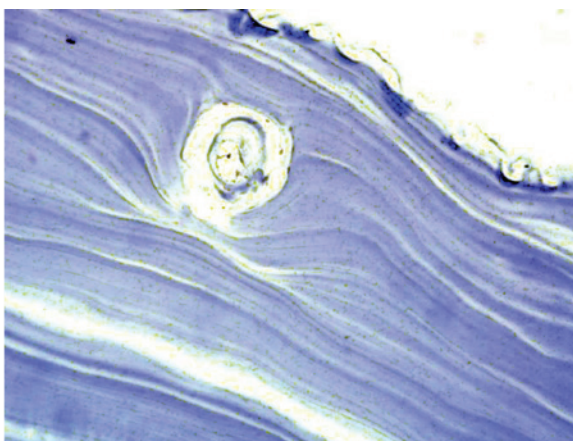


Cystic image of 3 cm in diameter inside de left ventricle (arrow).

Postoperative echocardiogram was later made, showing moderate left ventricular dysfunction, for what he was started on carvedilol, spironolactone and enalapril. Holter monitor was done 13 days after surgery, presenting permanent sinus rhythm alternating with ectopic atrial rhythm during sleep hours with chronotropic range in its superior limit, isolated premature ventricular complexes and no symptoms were reported in diary. Cardiology didn't initiate any antiarrhythmics drugs.

Due to good clinical evolution, he was discharged from the hospital 38 days after admission. He continues follow-up by pediatric clinic service, infectology and cardiology. He maintains cardiological treatment with carvedilol, spironolactone and enalapril. He completed antibiotic treatment with praziquantel weekly for a period of fifteen months, and daily albendazole for a period of eighteen months. After two years of hospital follow-up, at the moment there are no recurrences of the disease, with good response to the treatment established. The last abdominal ultrasound shows residual spleen, and no cystic images are evident.

FIGURE 3. Sample of material removed from inside the right femoral artery in the first surgery (femoral arteriotomy)



In the histological section, a wall corresponding to a hydatid cyst is observed, constituted by the laminated membrane that partially exhibits the germinal layer and in the interior, a scolece is recognized. (H & E 10X).

DISCUSSION

Cardiac hydatidosis with peripheral embolism is an infrequent presentation of the disease but it has high morbimortality. *E. granulosus* accesses the heart mainly through the coronary circulation, but a patent foramen ovale, the pulmonary circulation, intestinal lymphatics, the thoracic duct, rectal and cava veins may be other places of access.³

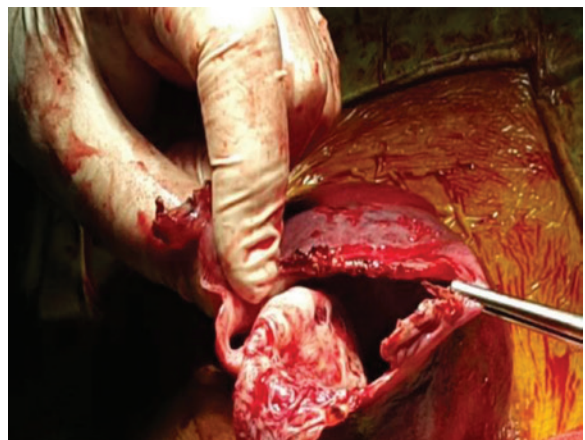
Every part of the heart may be affected, but the most common site is the left ventricle (50-77%), being the right ventricle and the interventricular septum (IVS) places of less frequency, 18% and 7% respectively.³

Generally, it presents as a single intramyocardial left cyst and, in the majority of cases, extracardiac involvement is present. Miralles et al., indicated that in up to 55% of cases, extracardiac involvement is associated, being the liver the most frequent site. Most of them are asymptomatic but carry a high risk of spontaneous rupture and complications, from embolic to severe anaphylaxis.⁴

Left heart cysts tend to grow to the epicardium and break in the epicardiac space. Right heart cysts tend to expand in the cavity beneath the subendocardium, and tend more frequently to break, thus, developing pulmonary embolism, anaphylaxis or sudden death.⁵

When the IVS is involved, arrhythmias are characteristic. Conduction block, as well as obstruction of the right and left ventricle outflow tracts, may be possible.⁶

FIGURE 4. Hydatidic splenic cist



Although cases related to cyst-vascular fistula has been described, arterial involvement generally develops due to cardiac cyst rupture and secondary embolism. Even so, cardiac hydatid cyst and secondary embolism, as this reported case, is extremely rare. In the literature, hydatidosis disease presented with peripheral arterial embolism has been reported in few cases, secondary to intracardiac cyst mainly.⁷⁻¹⁴ The presentation in pediatric patients was scarce.

CONCLUSION

Although acute arterial embolism is a rare form of atypical presentation of disseminated hydatidosis, in our country, an endemic area for hydatidosis, it should be considered as a differential diagnosis for arterial thromboembolism or cardiac mass in patients with compatible imaging and epidemiology. ■

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