

culprit arteries is complete revascularization in the acute phase. The most frequently described multiple coronary artery occlusions are found in the left anterior descending artery and right coronary artery ($\leq 50\%$ of cases),³ as in the patient described here. The peculiarity of our patient lies in the fact that the acute occlusion extended beyond coronary territory to the superficial femoral artery. Furthermore, we have shown that the patient presented drug resistance to conventional antiaggregation therapy. This resistance may share pathologic mechanisms with greater vulnerability to atherosclerotic plaques. New antiplatelet agents, like prasugrel, may well be more appropriate in these patients, in whom the choice of treatment benefits from a platelet aggregation study.⁴

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Sudden Death Due to Histiocytoid Cardiomyopathy

Muerte súbita debida a miocardiopatía histiocitoide

To the Editor,

Cardiac disease must be systematically evoked in the event of sudden death in the child. Histiocytoid cardiomyopathy (HC) is a rare disease responsible for severe ventricular arrhythmia very early in life. We report 2 cases of HC diagnosed after sudden death in young girls.

CASE 1

M. was the first child of healthy, non-blood relative, Caucasian parents. Hypertrophic cardiomyopathy was diagnosed at third trimester by antenatal ultrasound. At birth, clinical examination revealed a systolic cardiac murmur, an axial hypotonia, linear cutaneous erythematous lesions of the face and the neck typical of the microphthalmia with linear skin defects (MLS) syndrome (Fig. 1). The EKG registered Wolff-Parkinson-White syndrome. Transthoracic echocardiography demonstrated a biventricular hypertrophic cardiomyopathy, a small perimembranous ventricular septal defect. At 7 days of life, she presented an orthodromic tachycardia, successfully treated by amiodarone. Etiologic study of hypertrophic cardiomyopathy was noncontributive. At the age of 3 months, she presented

ventricular fibrillation with no resumption of electric activity despite defibrillations. The autopsy confirmed cardiac hypertrophy with micronodules on the mitral and tricuspid valves. Histological analysis revealed areas formed from bundles of myocardial fibers of normal appearance, contrasting with cells of histiocytoid appearance located in the myocardium, the pericardium, and the valves, leading to the diagnosis of HC.

CASE 2

M. was the first healthy child of non-blood relative, Caucasian parents. At the age of 20 months, she presented a cardiorespiratory arrest secondary to a polymorphic ventricular tachycardia refractory to cardiac defibrillations (Fig. 2A). At the autopsy, small yellow nodules were observed on the leaflets of the tricuspid valve (Fig. 2B). These nodules were composed of large, foamy, granular cells in the subendocardium. Immunostaining showed perimembranous reactivity for muscle-specific actin, but not for the histiocytic markers (PS100, CD68). Histological analysis revealed the presence of cellular clusters of histiocytoid cells in the subendocardial region, from the apex to the atrium, in the thickness of atrial septum and atrioventricular valve leaflets, resulting in the diagnosis of HC (Fig. 2C).

The diagnosis of HC should be systematically evoked in case of sudden death or severe ventricular arrhythmia in children, mainly in girls less than 2 years old.¹ Of the reported cases of HC, 19%

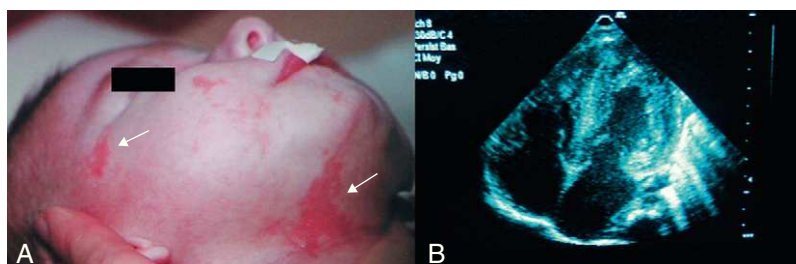


Figure 1. A: linear cutaneous erythematous lesions of the face and the neck (white arrows) typical of the microphthalmia with linear skin defects syndrome. B: trans-thoracic echocardiography showing hypertrophic cardiomyopathy.

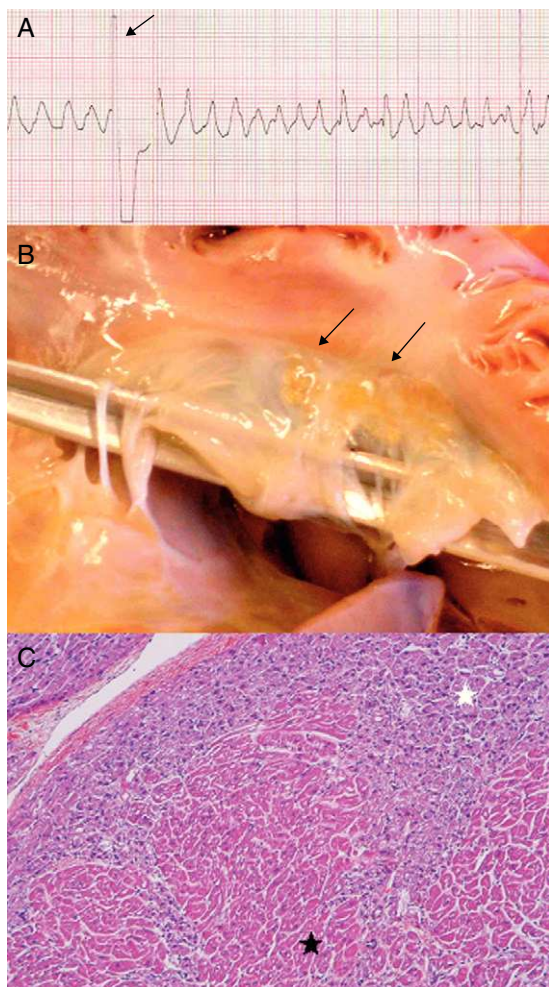


Figure 2. A: polymorphic ventricular tachycardia refractory to cardiac defibrillation. B: yellow arrhythmogenic histiocytoid nodules on leaflets of tricuspid valve (black arrows). C: histological section of the left ventricle displaying large foamy granular cells (white star) and normal myocardial cells (black star).

appeared as sudden deaths in children with no particular history. These patients frequently presented a supraventricular or ventricular arrhythmia uncontrolled by conventional antiarrhythmic treatments. The Wolff-Parkinson-White syndrome, found in almost 10% of the cases, may suggest a diffuse heart disease and may be a marker of gravity. The echocardiographic phenotype is variable. Symmetric or asymmetric hypertrophic cardiomyopathy

with a trabeculated endocardium is very suggestive. Cardiac malformations (ventricular septal defect, atrial septal defect, Shone syndrome) are reported. A normal echocardiography may be found in a focal form of the disease. Histologically, HC is characterized by the presence of nodules with more or less scattering in the myocardium or the endocardium.² The excision of arrhythmogenic nodules is the only treatment that may produce a complete cure and disappearance of arrhythmia.³ Histiocytoid cells have an arrhythmogenic potential and conventional treatment of arrhythmia is not effective. In every case, medical treatment resulted in the death of the patient. For the rare patients who benefit from surgical excision, the long-term prognosis is unknown. Heart transplantation was reported once, in a 30-month-old child.⁴ The etiopathogenesis of the HC is not yet clearly established; HC should be considered as an X-linked mitochondrial disease affecting cardiac muscle. This hypothesis seems confirmed following an association with MLS syndrome, a rare neurodevelopmental disorder that associates linear cutaneous erythematous lesions of the face and the neck, eye abnormalities, and neurological lesions in females monosomic for Xp22.3.⁵

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Pulmonary Embolism Caused by Cysto-Atrial Shunt Fragment

Embolia pulmonar causada por un fragmento de una derivación cistoauricular

To the Editor,

A 40-year-old woman presented to the accident and emergency department complaining of acute onset of headache and dizziness, along with right-sided pleuritic chest pain and shortness of breath. She had a history of a symptomatic right temporal lobe arachnoid cyst treated with a cysto-atrial shunt at the age of 25. Her vital signs were stable and ECG was unremarkable.

Chest radiography depicted a sizable, serpentine-shaped, foreign body within the left hilum (Fig. 1A) and a lateral skull X-ray showed fragmentation of the cysto-atrial catheter at the level of the skull (Fig. 1B). A noncontrast chest computed tomography demonstrated a large catheter fragment embolizing the pulmonary trunk and the left main pulmonary artery (Fig. 2). Patient underwent urgent percutaneous fragment retrieval with a snare apparatus catheter followed by cystoperitoneal shunt implantation, and made an uneventful recovery.

Placement of the distal catheter of a cerebrospinal fluid diversion apparatus in the right atrium is a well-established alternative when peritoneal insertion is contraindicated. Cardiac thrombus formation and chronic thromboembolic pulmonary hypertension are the most common cardiovascular complica-