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Sudden death due to congenital left ventricular aneurysm in a 21-year-old woman

Muerte súbita por aneurisma ventricular izquierdo congénito en una mujer de 21 años de edad

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To the Editor:

Sudden cardiac death remains a great challenge for cardiologists and health systems in the world[1]. It is defined as a sudden unexpected death in an otherwise healthy person or patient who is not expected to die within 6 hours after the onset of symptoms or, when unwitnessed, within 24 hours after the person was last seen in good health[1,2].

There are several causes that may produce it[3,4], including left ventricle aneurysm (LV); but its most frequent presentation is when it appears secondary to ischemic heart disease, due to the widely affected myocardial zone[5].

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Congenital LV aneurysm is infrequent, as it is often acquired, but may be fatal[6]. Diagnosis of left ventricular aneurysms can be made after exclusion of the various diseases that may cause them[6,7]; therefore, although found in life, their congenital origin can be difficult to discern.

We present the case of a 21-year-old white woman who although was born with a heart murmur considered unimportant, had a normal quality of life, with no worrisome cardiovascular symptoms; as those she presented were related to palpitations to physical effort which relieved after a brief period of rest. She had an active normal life and, according to her relatives, reported frequent palpitations, accompanied by dyspnea, which she attributed to her daily physical activity. A year before her death, she had a eutocic birth, a newborn with good weight and Apgar 9/9, without any warning signs or symptoms related to her underlying illness, evidently unknown. No family history of congenital heart disease or other type of heart disease was recorded.

After travelling a long distance on bicycle, she

had a dramatic episode of palpitations, sweating, coldness and cyanosis, which led to her death. The diagnosis was sudden arrhythmic cardiac death due to congenital LV aneurysm. The **figure** shows the anatomical piece, without signs of rupture or thrombus inside.

According to Pérez-Fernández *et al*[6], the clinical presentation of this type of aneurysm is very variable and the treatment is not standardized due to its low prevalence. It is important to distinguish it from the congenital left ventricular diverticulum, as it may cause confusion[8,9]. Congenital LV aneurysm has been associated with developmental anomalies, endocardium and myocardium dysplasia, with no evidences of viral infection[9]; unlike the diverticulum, they have a wider connection with the LV, histologically they lack myocardial muscular layer presenting a single one of fibroelastic tissue, that is often calcified; Moreover, it is rarely associated with other congenital malformations[6,9]. Aneurysms and diverticulum can be differentiated by several criteria. Contractility is an important parameter, as aneurysms expand, whereas diverticulum contract during ventricular systole[6,8].

Congenital LV aneurysms may initially go unnoticed, or be a diagnostic finding; but their clinical manifestations, when they appear, are highly variable and include arrhythmias, mainly ventricular (as, apparently, occurred in this case), heart failure, peripheral embolisms and sudden death[6-9]. However, the feared rupture with cardiac tamponade is less frequent[9].

Basso *et al*[4] studied 650 cases with sudden cardiac death post-mortem (201 women, 31%) and found a mechanical cause in 7% and an arrhythmic in 93% of the cases, which coincides with what, apparently, happened in this case. Both age and anatomopathological confirmations of absence of coronary disease, previous infarction or hypertrophic cardiomyopathy, as well as the histological study (presence of fibroelastic tissue), contributed to establish the congenital origin of the aneurysm.

Non-invasive imaging techniques such as: radiography, echocardiography, computed tomography and magnetic resonance imaging are very useful for diagnosis[6-10], but its treatment is controversial and must be individualized: from conservative, in the absence of symptoms, to surgical[6,7,9]. According to Savío Benavides *et al*[9] and Martín-Trenor[11], surgery is reserved for patients with: thin aneurysm wall and greater probability of rupture, presence of arrhythmias or thromboembolic phenomena, increase in size, or presence of symptoms and signs of heart failure.

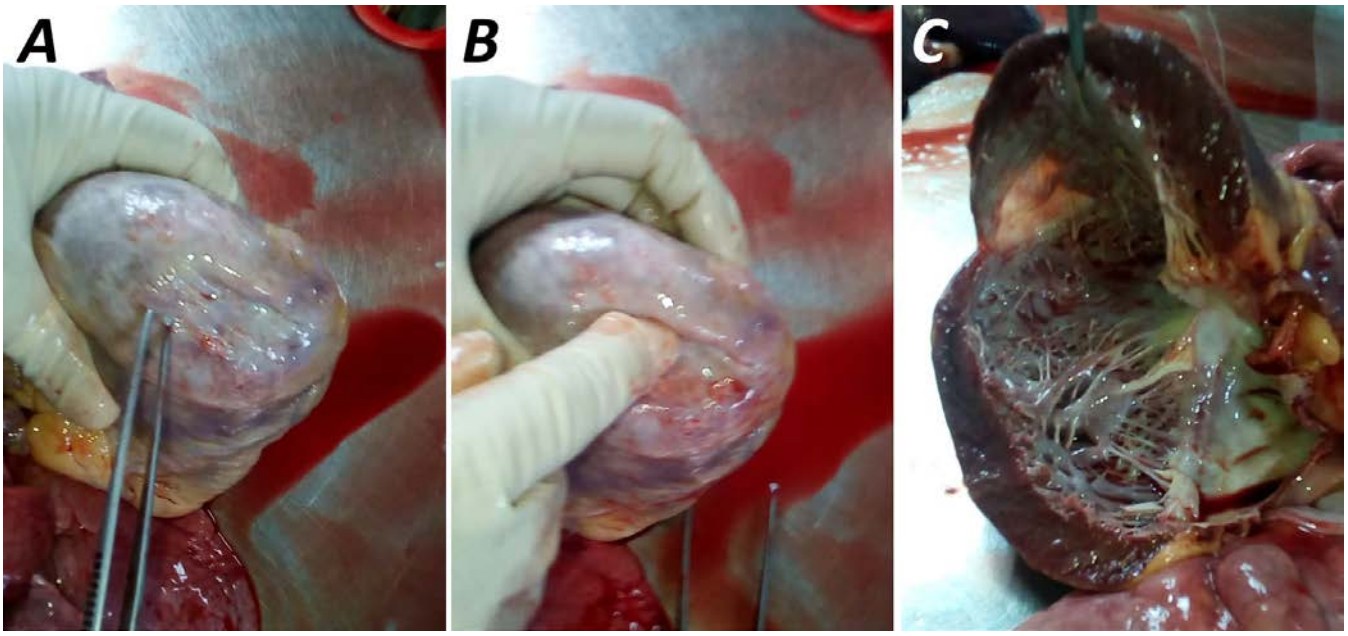

Figure. Anatomical piece where the congenital left ventricle aneurysm is observed. **A**. External view. **B**. Digital compression shows wall weakness at the aneurysm level. **C**. Section of the left ventricle showing apical involvement extending to the posterobasal wall region where fibroelastic tissue is evident.

CONFLICTS OF INTERESTS

None.

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