VENTRICULAR DIVERTICULUM

Image in cardiology

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FIGURE 1: Protuberant umbilicus
A markedly pulsating protuberant umbilicus was noticed at presentation.

PRESENTATION

A six-week old girl presented at Paediatric Cardiology after a pulsatile umbilical mass was noticed during routine post-natal visit. The baby was asymptomatic with no feeding difficulties and no history of cardiac failure.

General examination revealed a healthy infant with a protuberant and markedly pulsating umbilicus which was easily palpable (Figure1). Vitals signs were normal for age except oxygen saturation of 88% in room air. On cardiovascular examination the pulses and blood pressure were normal. Auscultation revealed an ejection systolic murmur over the precordium, radiating into the lung fields. She had no signs of cardiac failure.

Chest X-ray showed a normal cardiothoracic index with oligemic lung fields. Sinus rhythm with a rate of 155bpm and no Q- or T-wave abnormalities was seen on ECG.

Echocardiography demonstrated double outlet right ventricle anatomy with infundibular and valvular pulmonary stenosis. A long pulsating apical left ventricular structure stretching into the umbilicus was observed. Left ventricle function and contractility was preserved.

Angiography confirmed the diagnosis of double outlet right ventricle and an elongated pulsating mass originating from the apex of the left ventricle. The mass had synchronous systolic contraction and could be followed into the umbilicus.

Cardiac MRI demonstrated a structure that was thick walled and contained myocardium. The diagnosis of a diaphragmatic hernia was also confirmed.

Differential diagnosis at this stage included a left ventricular diverticulum or aneurysm.

Congenital diverticulae and aneurysms are rare cardiac malformations and can be differentiated on an etiologic, functional and anatomical basis.[1,2,3] These conditions are two separate well-defined entities and the terminology should not be used interchangeably. Patients may be asymptomatic or present with serious complications including sudden cardiac death due to ventricular arrhythmias, rupture or systemic embolism.[1,2,4,5]

The etiologic basis for ventricular diverticulae is uncertain and has been described as congenital disruption of the left ventricle during embryogenesis, as part of Cantrell syndrome.[2] Pentalogy of Cantrell is associated with midline thoraco-abdominal defects with combinations of abdominal wall, diaphragmatic, sternal, peri-cardial and cardiac defects, most frequently Tetralogy of Fallot or a left ventricular diverticulum. This may be due to failure of normal fusion of the paired primitive mesoderm in combination with abnormal fusion of the cardiac loop to the yolk sac. The cardiac wall may become attached through the septum transversum to structures forming the yolk sac and then become drawn out as the embryo grows thus resulting in the diverticulum.[1,6,7] Diverticulae usually have narrow connections with the left ventricle and contain all three layers of the myocardium, are muscular and show synchronised systolic contraction.[6,8]

Ventricular aneurysms on the other hand are usually the consequence of ischaemia, infection or trauma. They have a wide connection with the LV, do not contain all the muscle layers and are noncontractile or dyskinetic.[7,10,11]

The size of diverticulae usually remains stable whereas aneurysms will increase in size over time which is an important prognostic factor.[12]

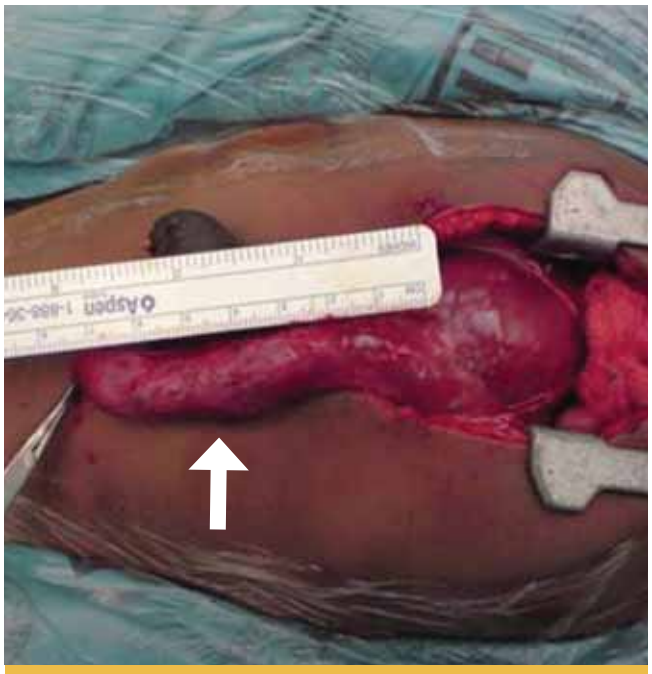

FIGURE 2: Surgery
During surgical repair an elongated left ventricle diverticulum was resected.

The treatment of left ventricular diverticulae is controversial and varies from surgery for those with complications and associated lesions to medical follow-up for the uncomplicated, isolated and "silent" left ventricular diverticulae.[1,7,12]

FINAL DIAGNOSIS

The patient was then referred for surgery and the diagnosis of left ventricular diverticulum was confirmed.

The diverticulum was dissected free from the umbilicus through a median sternotomy and ligated at the base using 3/0 polypropylene suture. It was then resected (Figure 2) and the ventricular aspect over sewn with a polypropylene suture. The associated Morgagni and umbilical hernias were repaired. The post-operative course of the patient was uncomplicated. The patient has been followed up for 16 months and is doing well. The associated cardiac lesions will be repaired at a later stage.

Conflict of interest: none declared.

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