Pulsating abdominal mass in a newborn – Pentalogy of Cantrell with left ventricular diverticulum

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ABSTRACT

Pentalogy of Cantrell is a rare congenital anomaly involving the anterior diaphragm, pericardium, sternum, peritoneum, and associated intracardiac defects. In this report, we describe a neonate with pentalogy of Cantrell evaluated with multimodality imaging and successfully managed by a multidisciplinary team.

Keywords: Left ventricular diverticulum, neonate, pentalogy of Cantrell

INTRODUCTION

Pentalogy of Cantrell is a rare congenital anomaly consisting of defects of supraumbilical peritoneum, anterior part of the diaphragm, apical pericardium, and lower part of the sternum and associated congenital intracardiac defects like tetralogy of Fallot, ventricular septal defect (VSD), and left ventricular (LV) diverticulum. We describe a neonate who presented with a pulsating abdominal mass and was successfully operated.

CASE REPORT

A neonate at birth was noted to have a pulsating mass in the epigastrium and was referred to our hospital. The baby was otherwise asymptomatic, and the vital parameters were stable. On examination, the neonate had a pulsating mass in the epigastrium extending to the umbilicus and herniating to the umbilical cord [Figure 1 and Video 1]. The skin over the swelling was intact. There was a bruit on the swelling. Chest X-ray showed mesocardia with no cardiomegaly. Echocardiography showed a large perimembranous VSD and an apical LV diverticulum contracting simultaneously with the LV [Figure 2]. An ultrasound abdomen showed LV diverticulum extending to the umbilical cord with thinned out wall at the tip [Figure 3]. Multisliced computed tomography (CT) showed LV apical diverticulum traversing caudally in the anterior abdominal wall to herniate through an omphalocele [Figure 4]. There was a small anterior diaphragmatic defect, absence of apical pericardium, and herniation of bowel loops through the anterior abdominal wall defect. Sternal abnormality could not be made out as the epiphyses were not well developed. A diagnosis of partial pentalogy of Cantrell was made.

Surgical resection of the diverticulum was indicated as the tip of the diverticulum was thinned out and there

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was a risk for rupture. Surgery was done via an upper midline abdominal incision. The diverticulum was ligated at the apex through the diaphragmatic defect and excised [Figure 5]. Two small hernial sacs in the abdomen were separated, reduced, and transfixed. The diaphragmatic and midline abdominal wall defects were repaired by direct closure using 6-0 PDS sutures. The umbilicus was reconstructed excising the gangrenous skin. The patient had a smooth postoperative recovery and was discharged after 1 week. At 4-months of follow up, the child is doing well and is planned for surgical closure perimembranous VSD.

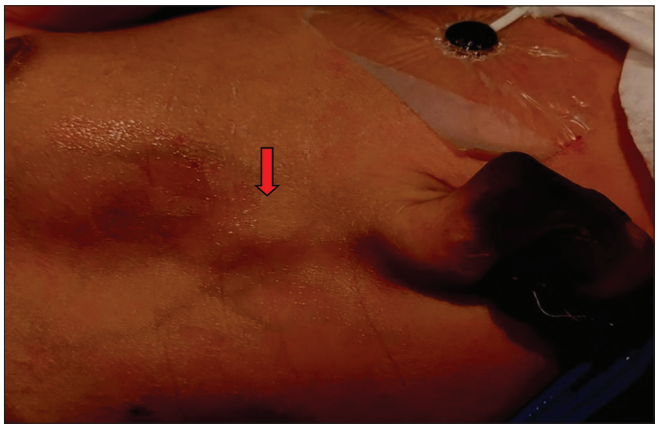

Figure 1: Pulsating abdominal mass extending into umbilical cord with herniation (Red arrow)

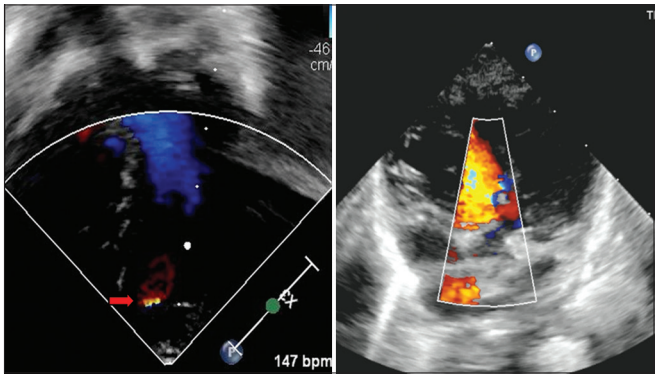

Figure 2: Echocardiography showing left ventricular apical diverticulum (arrow) and large perimembranous ventricular septal defect

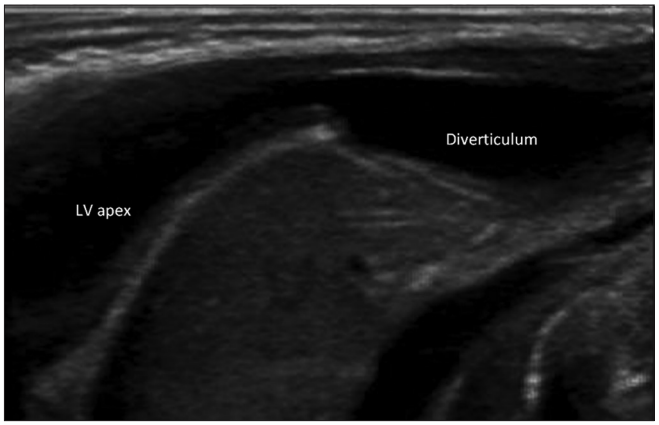

Figure 3: Ultrasonogram in sagittal plane showing diverticulum arising from left ventricular apex traversing caudally along the anterior abdominal wall

DISCUSSION

In 1958, Cantrell, Haller, and Ravitch described a constellation of multiple congenital anomalies including a midline anterior abdominal wall defect, a distal sternal cleft, a defect in the anterior diaphragm, and a defect of the apical pericardium with pericardioperitoneal communication, as well as intracardiac anomalies. The cardiac defects reported in pentalogy of Cantrell include tetralogy of Fallot, VSD, double outlet right ventricle, hypoplastic left heart syndrome, and LV diverticulum.[1] The prevalence of this syndrome varies between 1:65,000 and 1:200,000 cases.[2] The embryological basis of these anomalies could be the failure of development of mesoderm between 14 and 19 days of the early embryonic period. Complete or partial Cantrell syndrome develops due to the differences in the timing of involvement of mesoderm. If an insult occurs after the differentiation of mesoderm into splanchnic and somatic layers, it does not involve the heart.[3] Variable combinations of components of the pentalogy of Cantrell have been reported. The abdominal wall defects include omphalocele, diastasis recti, umbilical hernia, or a combination of these. Sternal defects include bifid sternum, absent xiphoid process, or short sternum. Anterior diaphragmatic defect is the common diaphragm defect and an absent apical pericardium with pericardioperitoneal connection is the common pericardial defect. Cantrell syndrome has been classified into three classes by Toyama *et al.*: in Class I, there is the occurrence of all 5 defects; in Class II, there are 4 defects with intracardiac and ventral abdominal wall abnormalities present; and in Class III, there is incomplete expression of the disorder showing various combinations of defects, although sternal anomalies are definitely present.[4]

In the above case, the patient had a large VSD requiring surgical management. However, as the neonate was only 7 days old and the LV diverticulum needed immediate surgical management, the surgery was done through an abdominal incision by a team of cardiothoracic and pediatric surgeons. LV diverticulum was excised, the diaphragmatic defect was repaired, the rectus sheath was closed, and the umbilical hernia was repaired with direct sutures.

In conclusion, Cantrell syndrome with LV diverticulum can be diagnosed by echocardiography and abdominal ultrasound, which can be confirmed with a CT scan. These patients may need early surgical repair in view of the possible risk of rupture and thrombosis.

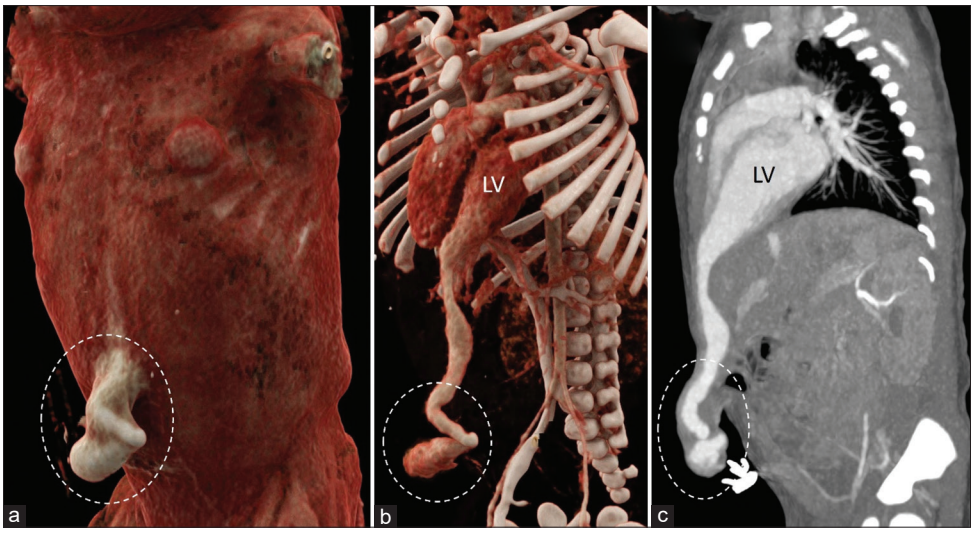

Figure 4: Volume rendered images (a and b) and oblique sagittal reconstruction (c) of ct angiography image showing the left ventricular (LV) diverticulum arising from the LV apex, traversing caudally and herniating through the anterior abdominal wall defect (dotted circle)

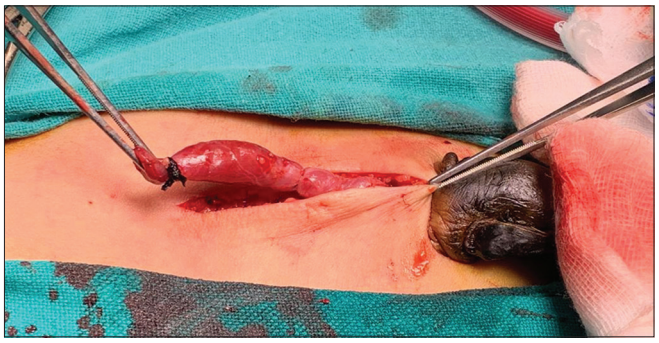

Figure 5: Operative picture showing the upper abdominal incision and LV diverticulum being excised

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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