

A Rare Case Report of Congenital Left Ventricular Diverticulum Complicated with Mesenteric Ischemia

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ABSTRACT

Pentalogy of Cantrell includes defects involving the heart, lower sternum, anterior diaphragm, diaphragmatic pericardium and midline abdominal region. Incomplete expression of Pentalogy of Cantrell is reported in the literature; therefore, the documentation of any one or more of these defects warrants further investigation. The major complications include valvular regurgitation, thrombosis, embolism, rupture, congestive heart failure, and ventricular arrhythmias. Our patient had incomplete expression of the Pentalogy with left ventricular diverticulum, lower sternal defect and supraumbilical abdominal wall defect. And the situation was complicated with superior mesenteric artery thrombosis and small bowel gangrene for which he had undergone small bowel resection and anastomoses. After 5 months of the surgery, patient developed multilobar consolidations in chest, myocarditis and had sudden deterioration with septic shock, acute kidney injury and despite of all resuscitative measures, the patient could not be saved.

INTRODUCTION

45 year old male patient presented to the casualty with acute onset of diffuse abdominal pain. Abdominal pain was associated with non-bilious vomiting. He had no history of systemic hypertension or diabetes mellitus in the past. The patient had a pedunculated pulsatile oval swelling in the epigastrium since birth, for which no evaluation was done previously. There was no significant change in the size of swelling over the past years. On examination, vitals were stable, diffuse tenderness was present in the abdomen. The epigastric swelling measured 6x5cm, non-tender, non-reducible. Thus the clinical suspicion was paraumbilical hernia. Routine blood investigation showed hemoglobin of 12 mg/dl, total count – 7800, ESR- 25 mm/1st hour. All the solid intra-abdominal organs were normal in the ultrasonogram of abdomen. USG Doppler examination of the swelling revealed a vascular mass with echogenic thrombus filling the lumen which showed synchronous pulsations with the heart. He was further investigated with CECT abdomen with lower chest to evaluate the cause of abdominal pain. CECT showed sternal defect of size 2.2 cm in the lower aspect, through which a 3.8x3.7 x 13 cm (APxTRxCC) sized focal outpouching of the left ventricle of the heart was projecting outside the thoracic cavity. The sac was directed antero inferiorly. The wall of the diverticulum showed focal thinning with non-enhancing areas in the distal part. However, there was no associated pericardial / diaphragmatic

Key Words: Left Ventricular Diverticulum, Pentalogy of Cantrell, Mesenteric Ischemia.

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defect or cardiac septal defects. Thus, a diagnosis of thrombosis in the congenital left ventricular diverticulum was considered. There were no bowel pathologies detected in the first CECT study. After 3 days, the patient had worsening symptoms of abdominal pain associated with malena and loose stools. CECT abdomen was repeated which revealed non enhancing hyperdense thrombus in the superior mesenteric artery 2.5 cm distal to the origin. The wall of the small bowel loops appeared thinned out with non-enhancing with multiple intra mural nondependent air foci.

Thus a diagnosis of mesenteric ischemia, with gangrenous small bowel loops, possibly secondary to the thrombo embolism from the left ventricular diverticulum was given. Patient underwent laparotomy. Intra-operatively, the small bowel loops appeared gangrenous and non-viable, which was resected and anastomosed. Patient was discharged on oral anticoagulants and other supportive measures.

Again, after 5 months, the patient presented with fever, cough, and breathlessness. Blood investigations showed anemia with elevated total count. Trop I was found to be positive which was suggestive of myocarditis. Chest x ray showed features of multilobar consolidation. Subsequently, patient went for septic shock, acute kidney injury. Despite all resuscitative measures and prompt management, the patient could not be saved.



Fig 1: Showing the pulsatile swelling in the epigastrium, projecting through the anterior abdominal wall defect

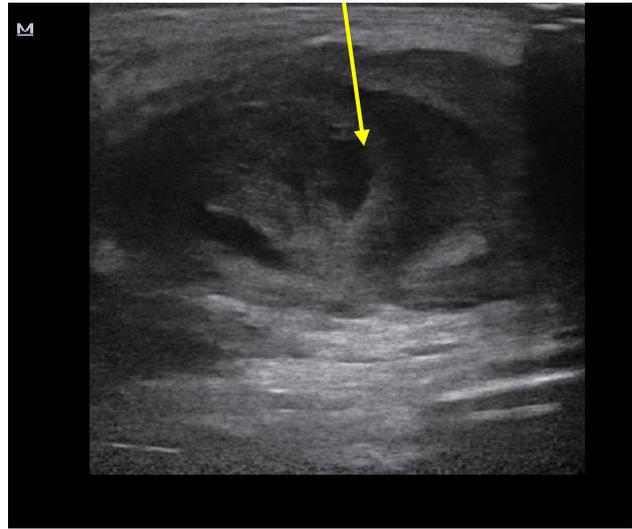


Fig 2: USG of the swelling showed multiple concentric echogenic areas in the diverticulum representing thrombosis.

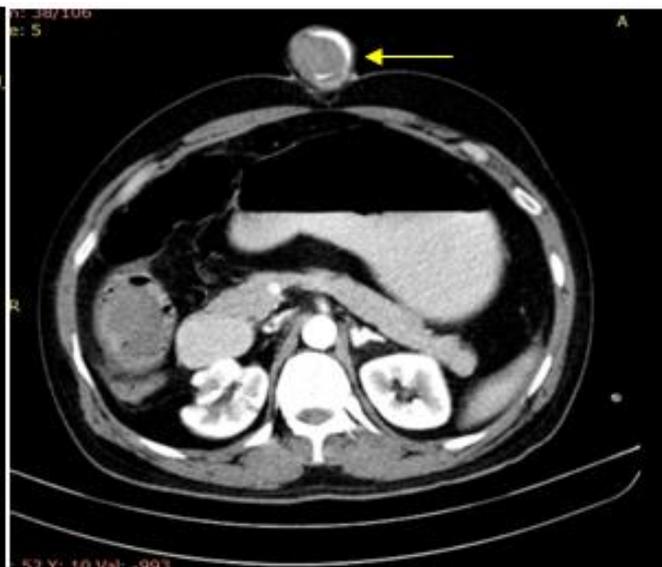
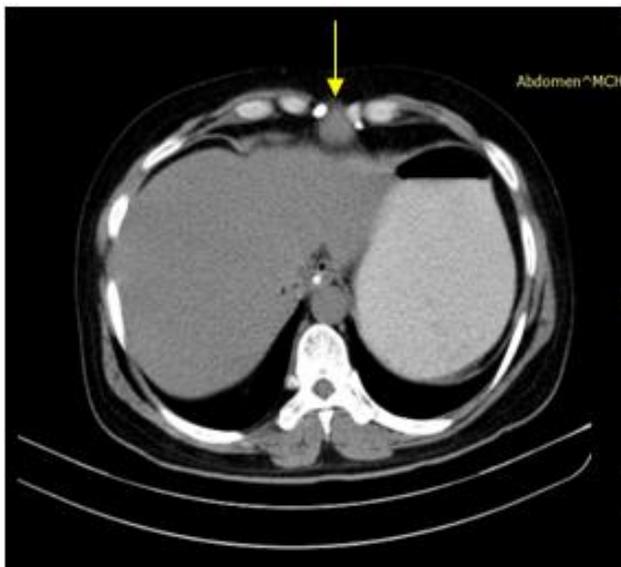


Fig 3: Axial CT sections of abdomen showing the defect in the lower part of the sternum and the left ventricular diverticulum with the thrombus.

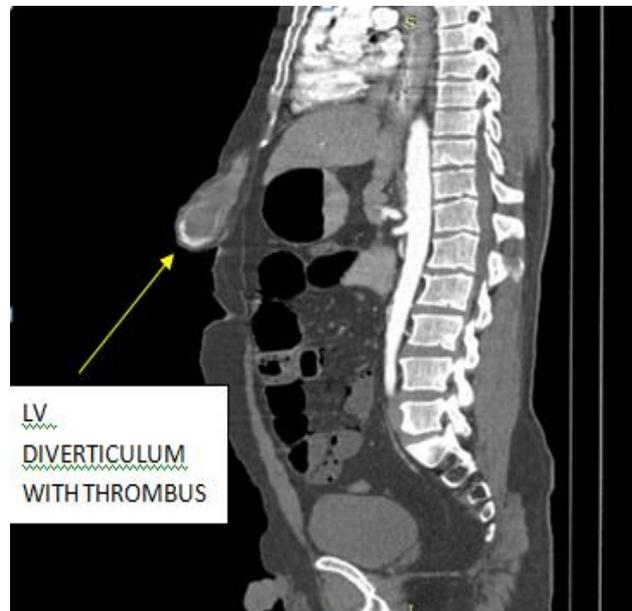
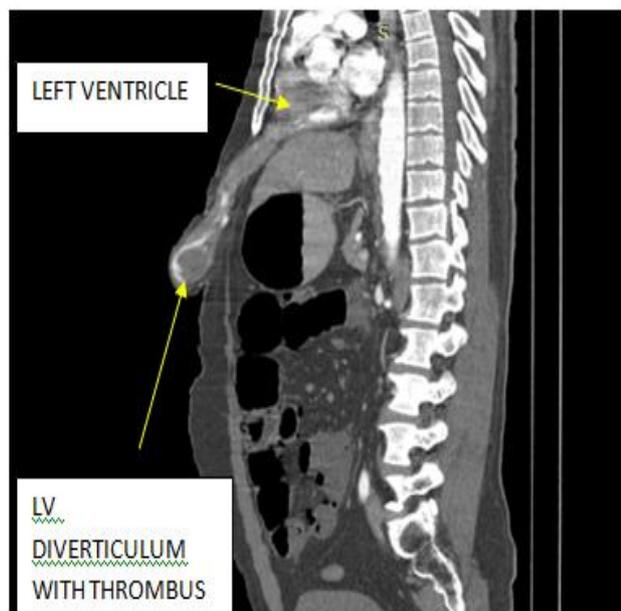


Fig 4: Sagittal post contrast images showing the left ventricular diverticulum, with thinned out walls and non-enhancing thrombus.



Fig 5: Axial sections of CT abdomen showing filling defect in the superior mesenteric artery.



Fig 6: Axial CT sections of abdomen showing the dilated, thinned out, non-enhancing, small bowel loops with non-dependent air foci in the wall.

DISCUSSION

Left ventricular diverticulum can be defined as an out pouching that contains all three layers of the heart and displays normal contraction.¹ The prevalence is reported as 0.4% in necropsy studies. There has been approximately 100 cases reported in the English literature with a prevalence estimated at 0.013%; however, the true prevalence is not clearly known.² Congenital left ventricular diverticulum starts in the 4th embryonic week. Possible etiologies are intrinsic abnormalities of embryogenesis or in utero acquired malformations (like viral infections, arrhythmia-related vascular accidents, or cardiomyopathies).³

In 1958, Cantrell described an association of cardiac defects with anterior abdominal defects, sternal defects, anterior diaphragmatic defects, and pericardial defect.⁴ Toyama subsequently published a modified classification for Pentalogy of Cantrell, enabling inclusion of patients who present with variable quantities of the classic spectrum of defects.⁵ Type 1 is the classic Pentalogy of Cantrell. Type II is partial expression with 4 defects present from birth which always include intracardiac abnormalities and type III incomplete expression which include less than 4 defects of variable combination which always include sternal defect. Our patient had incomplete expression with anterior abdominal wall defect, left ventricular diverticulum with sternal defect.

LV diverticulum is characterized by a finger or hook-like appendix of the entire left ventricular wall, beyond the myocardial margin, frequently localized in the left ventricle. It can be differentiated into two types: muscular or fibrous. The muscular type is more frequent and not prone to rupture. The fibrous diverticulum is usually located either in the base of the heart or in the subvalvular area, leading to possible aortic or mitral regurgitation. It is characterized by a fibrous layer, a narrow neck, no contractile function, a tendency to rupture and is not associated with other malformations.

Differential diagnosis may include true LV aneurysm and LV pseudoaneurysm. LV aneurysm has been strictly defined as a distinct area of abnormal left ventricular diastolic contour with systolic dyskinesia or paradoxical bulging. This pathologic

condition involves bulging of the full thickness of the left ventricular wall with thinned fibrous tissue (scar) as remnant of the left ventricular muscle. False aneurysms of the left ventricle or left ventricle pseudoaneurysms may be caused by acute contained rupture of the ventricle wall, often after myocardial infarction, or after circumflex coronary arterial occlusion.^{1,4} True LV aneurysm and LV pseudoaneurysm are characterized by a common contractile function, exhibiting either akinesia or dyskinesia.

ECG alterations remain the first clinical sign.⁶ Coronary angiography is the gold standard in the evaluation of coronary artery disease (CAD). Left ventricular catheterization, as part of the examination, is very useful in assessing the morphology and dynamics of the left ventricular chamber. The cardiac MRI is a fundamental tool for the performance of a differential diagnosis with true aneurysms and left ventricle pseudoaneurysm. In the case of a true aneurysm, the wall of the aneurysm shows delayed enhancement, indicating scar tissue as a result of infarcted myocardial muscle. A pseudoaneurysm is composed only of pericardium and does not show delayed enhancement within the sac, while the border of the aneurysm shows enhancement that indicates a perianeurysmal infarcted area.⁷

The major complications include valvular regurgitation, thrombosis, embolism, rupture, congestive heart failure, and ventricular arrhythmias.¹ The natural history of the disease depends on the associated anomalies. Long-term follow-up of uncomplicated diverticulum have been shown to have a benign course.⁸ The treatment for complicated or symptomatic cases is surgical resection. However, some suggest surgical resection for all patients.

CONCLUSION

Cantrell's pentad includes defects involving the lower sternum, anterior diaphragm, diaphragmatic pericardium, midline/ventral abdominal region, and heart. A patient presenting with any lesions associated with this pentad warrants a thorough evaluation to comprehensively assess mortality risks, the feasibility and eligibility for surgical repair. Incomplete expression of the Pentalogy is reported in the literature; therefore, the documentation of any one or more of these defects warrants

investigation with 2D/3D ultrasound or MRI. The major complications include valvular regurgitation, thrombosis, embolism, rupture, congestive heart failure, and ventricular arrhythmias. Management of the diverticulum depends on the clinical situation and the associated abnormalities. Most asymptomatic diverticula can be managed with a conservative approach. Treatment options for high risk cases include surgery, anticoagulants and management of the complications.

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