A complete case of Cantrell's Pentalogy with isolated left ventricular diverticulum

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Abstract

The congenital left ventricular diverticulum is a rare cardiac malformation, and it may associate with Cantrell's Pentalogy with other cardiac defects. However, isolated ventricular diverticulum without any other cardiac defect in complete Cantrell's syndrome is very rare. We describe a 6-year-old male patient with a complete Cantrell's syndrome with isolated left ventricular diverticulum.

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Abstract:

The congenital left ventricular diverticulum is a rare cardiac malformation, and it may associate with Cantrell's Pentalogy with other cardiac defects. However, isolated ventricular diverticulum without any other cardiac defect in complete Cantrell's syndrome is very rare. We describe a 6-year-old male patient with a complete Cantrell's syndrome with isolated left ventricular diverticulum.

Keywords: Cardiac diverticulum, Cantrell's syndrome, congenital heart defect

Introduction:

The incidence of the left ventricular diverticulum is reported to be 0.05% of all congenital heart malformations. It is currently believed that it occurs as a result of impaired development of the endocardial tube during the 4th week of embryologic development. Although it may present as an isolated disorder, this entity is often associated with other cardiac abnormalities or with Cantrell pentalogy in around 70% of cases (1,2). Cantrell's Pentalogy is a rare syndrome with an estimated incidence of 5.5 per 1 million live births. It was defined by Cantrell et al. in 1958 as a condition with different degrees of five defects: supraumbilical abdominal wall defect, lower sternal cleft, defect of the diaphragm in the central tendon, pericardium

defect, and various intracardiac anomalies. They considered the intracardiac defects as an integral of this association and further described the frequency as 100% for ventricular septal defect, 53% for atrial septal defect, 33% for pulmonary stenosis (either valvular or infundibular), 20% for left ventricular diverticulum, and described Tetralogy of Fallot as the only established cardiac syndrome to be accompanying this syndrome with a frequency of 20% (3). In 1972, Toyama et al. proposed a classification of this syndrome into three subtypes. The first subtype is the complete syndrome with all five defects. The second subtype is the probable syndrome with four defects, including intracardiac and ventral wall anomalies. The third subtype is the incomplete syndrome with various combinations of defects sternal abnormality (4).

We describe a 6-year-old male patient with a pulsatile mass extending from the lower chest to the upper abdomen. The IRB approval, consent statement and clinical trial registration were waived.

Case report:

After an unsuccessful percutaneous intervention to close the ventricular diverticulum, the patient was admitted to our clinic. The history obtained from his family revealed that he was referred to a pediatric cardiologist in his hometown with the complaint of a pulsating mass in his abdomen before his admission to our department. ECG, 24h Holter monitor, and abdominal USG were performed for further investigation. ECG and 24-Holter monitoring were found to be normal. Abdominal USG revealed a 6mm wide and 20 mm long pulsatile lesion and a 12mm wide umbilical herniation. He presented to our clinic after an unsuccessful attempt of diverticulum closure with an endovascular plug. The combination of umbilical herniation and left ventricular diverticulum led to a probable diagnosis of Cantrell's Pentalogy. Figure 1 shows the preoperative angiographic view of the left ventricular diverticulum.

The patient underwent corrective surgery at six years of age. A midsternal incision and median sternotomy were made. The lower third of the sternum was absent, and the anterior pericardium was observed as a thin membrane. The pericardium was opened, and a piece was reserved to be used as a pledget. The apical left ventricle diverticulum with a length of 8 cm was observed, and the incision was extended to the umbilicus. The diverticulum was separated from the surrounding fascia. Aortic and two-stage venous cannulations were performed to initiate the cardiopulmonary bypass. The diastolic arrest was provided with a cross-clamp to ascending aorta using antegrade tepid blood cardioplegia. The diverticulum was excised, and the three layers (endocardium, myocardium, and epicardium) were observed. Figure 2 shows the operative view of the diverticulum before excision and three layers of the diverticulum on both sides. The excised diverticulum was sent to pathology for examination, and the ventricular side of the defect was closed with 4-0 prolene sutures and pericardial pledgetes. After gradual reduction, the cardiopulmonary bypass was ended, and the heart was decannulated. Wires for the temporary ventricular pacemaker and the drainage tubes were placed. After primary closure of the umbilical defect and the anterior abdominal wall, sternotomy and the abdominal incision were closed.

Discussion:

Cantrell's Pentalogy is a rare syndrome with a high mortality rate and poor prognosis. The long-term prognosis mainly depends on the severity of the cardiac and accompanying defects and requires multidisciplinary management due to the involvement of multisystem anomalies (5). The syndrome is characterized by a partial sternal cleft, anterior abdominal wall defects, anterior diaphragmatic defect, and intracardiac defects (6). The steps for closure are usually chest wall closure, closure of the sternal defect, closure of the omphalocele, placement of the heart in the thorax, and repair of the intracardiac defect (7). Nearly one-third of the cases with this syndrome have left ventricular diverticulum, which is a life-threatening abnormality that may cause systemic embolization, fatal ventricular arrhythmias, and sudden death due to ventricle rupture. Early detection and surgical intervention for this pathology might be lifesaving (8).

Jaime F. Vazquez-Jimenez et al. published a literature review of 153 Cantrell's Pentalogy cases in 1998. According to the analysis, ventricular septal defect was the most common cardiac malformation (72%). Atrial septal defect was present in 44 (34.6%) of patients, left ventricular diverticulum in 41 (32.3%), pulmonary stenosis or atresia in 40 (31.5%), Tetralogy of Fallot was present in 22 patients (17.3%), dextrocardia in 19

(15.0%) and it was accompanied by various malformations in lesser numbers (6). The analysis showed 3 cases of Cantrell's Pentalogy, of which the intracardiac defect is limited with Left Ventricle Diverticulum (6%), without any accompanying intracardiac defects. First of these three cases was a 2-year-old female patient with a lower sternal defect, an umbilical hernia, and a ventral diaphragmatic defect; the second case was a 2-year-old male patient with a short sternum, an omphalocele, and a diaphragmatic defect; and the third case was a 7-month-old male patient with a huge ventral hernia, a diaphragmatic defect and a pericardial defect coherent with the incomplete syndrome. In addition, a case of complete syndrome of which the intracardiac defect is limited to the left and right ventricle along with a bifid sternum, an omphalocele, a ventral hernia, and absent pericardium has been reported (9).

Turbendian et al. reported another incomplete case of Cantrell's Pentalogy with an intracardiac defect limited to the left ventricular diverticulum along with a PDA, an omphalocele, a pericardial, and a diaphragmatic defect (10).

According to the Toyama classification, our case fulfills all requirements of the complete syndrome with supraumbilical abdominal wall defect, lower sternal cleft, defect of the diaphragm in the central tendon, pericardium defect, and left ventricular diverticulum. According to the previous research, the left ventricular diverticulum is a defect related to Cantrell's Syndrome almost always accompanies other intracardiac defects (11). However, the intracardiac defect is limited to the left ventricle diverticulum in our patient.

In summary, our case is interesting because of the presence of left ventricular diverticulum alone. Also, isolated ventricular diverticulum often accompanies incomplete Cantrell's cases, and our case is one of the rarest complete Cantrell's syndromes with no other intracardiac defects.

AUTHOR CONTRIBUTIONS

All authors have contributed to the concept, critical revision, and approval of the final article.

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