Incomplete Cantrell's Pentalogy—A Case Report

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Abstract We present a case of an 11-year-old boy, who presented to us with a ventral hernia since birth. He was found to have the following: A large ventral midline defect starting from umbilicus to xiphisternum. Sternum was bifid at the lower end. Liver had three separate lobes. Extrahepatic biliary apparatus was normal and liver was ptotic. Diaphragm had an anterior midline defect. Diaphragmatic defect was in continuity with pericardial defect and heart could be seen directly pulsating through the skin. Despite modern surgical standards, Cantrell's syndrome represents a challenge to the surgeon because of the wide spectrum of anomalies, the severity of the abdominal and cardiac malformations, and the high mortality.

Keywords Congenital cardiac defect · Pentalogy of Cantrell · Situs inversus of the liver

Introduction

Cantrell's pentalogy (CP) a rare syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. The characteristics of this syndrome are as follows [1]:

- A midline supraumbilical abdominal wall defect
- A defect of the lower part of the sternum
- A deficiency of the anterior diaphragm
- A defect in the diaphragmatic pericardium
- A congenital heart malformation

The pathogenesis of CP has not been understood since its first description. Cantrell et al. postulated these malformations to result from developmental failure in differentiation of a segment of intraembryonic mesoderm between splanchnic and parietal planes around 14–18 days of embryonic life [1]

In 1972, Toyama proposed the following classification of CP, based on 60 cases described in the literature [2]:

1. Confirmed diagnosis—all elements of pentalogy present
2. Probable CP—four defects (intracardiac, involving the abdominal wall and the sternum)
3. Incomplete CP—various combinations (always including sternal anomalies)

Echocardiography usually gives the diagnosis of intracardiac defects and the treatment of defects is difficult, and therefore it requires multistage corrective procedures. The main aim is to repair the abdominal, thoracic, and cardiac malformations.

A Case Report

An 11-year-old boy presented to us with a ventral hernia since birth (Fig. 1). The defect did not close or decrease in size with age but increased in size. He never complained of dyspnea or respiratory infections. He presented to us very late because of illiteracy and poor socioeconomic conditions.

On examination, patient was averagely built and nourished, and all vital parameters were within normal limits. His respiratory and cardiac examinations were also within normal limits. Umbilical scar was absent. His CXR and echo was normal. USG was also normal.

On exploration for hernia, he was found to have the following:

- A large ventral midline defect starting from umbilicus to xiphisternum.
- Sternum was bifid at the lower end.

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Fig. 1 Preoperative ventral hernia since birth

- Liver had three separate lobes. Extrahepatic biliary apparatus was normal and liver was ptotic (Fig. 2).
- Diaphragm had an anterior midline defect (Fig. 3).
- Diaphragmatic defect was in continuity with pericardial defect and heart could be seen directly pulsating through the skin (Fig. 3).

We modified ligamentum teres which was redundant and sutured to excess of diaphragm and pericardium. Some loose sutures are taken to avoid tamponade. A Prolene mesh was sutured underneath to repair diaphragmatic defect (Fig. 4). Liver anomaly was not disturbed. Anterior abdominal wall defect was sutured in anatomical manner with Prolene. The patient tolerated the procedure well. His parameters and CXR were normal after 1 month.

Discussion

Pentalogy of Cantrell is a rare syndrome characterized by a partial sternal cleft, anterior abdominal wall defects, and anterior diaphragmatic defect and intracardiac defects of which ventricular septal defect is the commonest and is invariably present. Left ventricular diverticulum is present in 20–50 % of cases [3, 4]. Other intracardiac defects, which

Fig. 2 Liver with three separate lobes and ptotic

Fig. 3 Diaphragm with an anterior midline defect in continuity with pericardial defect and heart could be seen directly pulsating through the skin

can be seen, are tetralogy of Fallot (17–20 %), atrial septal defect (34.6 %), pulmonary stenosis or atresia (31.5 %), transposition of great arteries, tricuspid atresia, truncus arteriosus, atrioventricular septal defects, anomalous pulmonary venous return, and right ventricular diverticulum [3].

These deformities are postulated to be due to developmental failure of appropriate segments of the mesoderm. The time of origin of Cantrell's pentalogy must be before or immediately after the differentiation of the primitive mesoderm into the splanchnic and somatic layers (between days 14 and 18 of embryonic life) [1]. The etiology is still unknown [5].

Echocardiography usually leads to the diagnosis of intracardiac defects, but in a few cases the diagnosis is made only during operation. The management of the syndrome is the most controversial issue. Because of the complexity of the syndrome, the treatment of defects is difficult and often requires multistage corrective procedures. According to many authors, the effectiveness and success of treatment as well as long-term survival depend primarily on the type of diagnosed heart defect [6].

Fig. 4 Modification of ligamentum teres sutured to excess of diaphragm and pericardium—a Prolene mesh sutured underneath to repair diaphragmatic defect

The closure of the abdominal wall defects only may compromise the cardiac or respiratory function. With the exception of an omphalocele, which should be corrected as soon as possible, the main indications for surgery are those of intracardiac defects and the aim is to repair the abdominal, thoracic, and cardiac malformations [7].

Sternal clefts are best corrected in neonatal period by approximation of sternal bands. For abdominal defects mesh reinforcement with or without relaxing incisions in the rectus sheath may be needed. Severe abdominal wall anomaly with a relatively stable cardiac defect may need correction of the former first. Minor abdominal wall defects can be repaired after correction of intracardiac defects [8].

In our case we modified ligamentum teres, which was redundant and sutured to excess of diaphragm and pericardium. Some loose sutures are taken to avoid tamponade. A Prolene mesh was sutured underneath to repair diaphragmatic defect. Liver anomaly was not disturbed. Anterior abdominal wall defect was repaired in the anatomical manner with Prolene.

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