CASE REPORT

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Incomplete pentalogy of Cantrell: a case report

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Abstract

Adult congenital heart defect operations still maintain their importance in cardiovascular surgery practice. Atrial septal defect (ASD) is the most commonly seen congenital heart defect in this group, and it may be associated with various anomalies. This report presents the case of a patient with ASD who was also diagnosed with pentalogy of Cantrell, a rare multiple congenital malformation syndrome characterized by a combination of features: a midline supraumbilical abdominal wall defect, a defect of the lower sternum, a defect of the diaphragmatic pericardium, deficiency of the anterior diaphragm, and congenital cardiac anomalies. In the absence of concomitant severe anomalies in patients with pentalogy of Cantrell, preoperative transthoracic echocardiography (TTE) may not be adequate to detect the congenital defects. Therefore, further imaging modalities are vital for the evaluation of these patients.
Key words: ASD, pentalogy of Cantrell, pectus excavatum.

Streszczenie

Operacje wrodzonych wad serca u dorosłych wciąż zajmują istotne miejsce w chirurgii serca i naczyń. Ubytek w przegrodzie międzyprzedsionkowej (*atrial septal defect* – ASD) jest najczęściej obserwowanym ubytkiem serca w tej grupie schorzeń; może być on też powiązany z różnymi anomaliami. W poniższym artykule prezentujemy przypadek pacjenta z ASD, u którego zdiagnozowano pentalogię Cantrella – rzadki zespół wrodzonych malformacji. Do jego składowych należą: ubytek w linii środkowej ściany brzusznej w okolicy nadpępkowej, ubytek w dolnej części mostka, ubytek przeponowej części osierdzia, przednia przepuklina przeponowa oraz wrodzone wady serca. W przypadku braku współwystępowania innych ciężkich anomalii u pacjentów z pentalogią Cantrella, przedoperacyjna echokardiografia przezklatkowa (*transthoracic echocardiography* – TTE) może nie wystarczyć do wykrycia wrodzonych ubytków. Dlatego tak istotne jest zastosowanie innych modalności obrazowania w celu poprawnego zdiagnozowania tych pacjentów.
Słowa kluczowe: ASD, pentalogia Cantrella, *pectus excavatum*.

Introduction

Adult congenital heart defects, including atrial septal defect (ASD), may be associated with various anomalies [1]. This report presents the case of a patient with ASD, who was also diagnosed with pentalogy of Cantrell, a rare multiple congenital malformation syndrome characterized by a combination of features: a midline supraumbilical abdominal wall defect, a defect of the lower sternum, a defect of the diaphragmatic pericardium, deficiency of the anterior diaphragm, and congenital cardiac anomalies [2]. Our patient demonstrated an incomplete form of pentalogy of Cantrell, which is very rare. In our case, the "abdominal wall defect" and "deficiency of the anterior diaphragm" components of pentalogy of Cantrell were absent.

Case report

A 21-year-old man was diagnosed with ASD, which appeared unsuitable for percutaneous closure due to rim deficiency visualized by transthoracic echocardiography (TTE). The TTE examination did not reveal any concomitant pathology. During elective ASD repair, we did not encounter any complications while performing median sternotomy to treat the pectus excavatum of the anterior chest wall. After the sternotomy, we realized that the pericardium covering the heart anteriorly was defective. There was a minimal amount of thymic tissue on the sides of the superior vena cava, and additional tissue was present instead of the pericardium, covering the heart from the left side down to the diaphragm (Fig. 1). The left phrenic nerve was visualized

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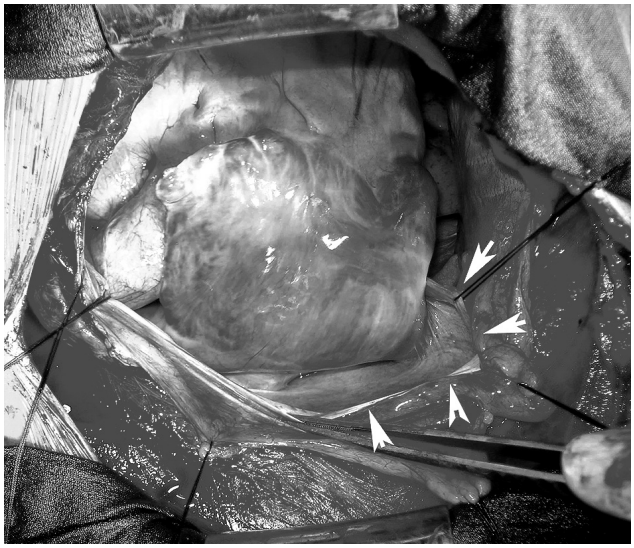

Fig. 1. A view of the defective pericardium absent on the left side of the heart

as defective, and its ventral location in relation to the pericardial tissue was abnormal (Fig. 2). A left-side dissection confirmed the presence of a free pericardial edge and agenesis of the anterior and diaphragmatic pericardium.

The ASD was repaired with a PTFE patch via routine cannulation using a cardiopulmonary bypass. The rest of the procedure was uneventful. The pectus excavatum defect was also repaired. No complications occurred during the postoperative course, and the patient was discharged on postoperative day 7 without any complications.

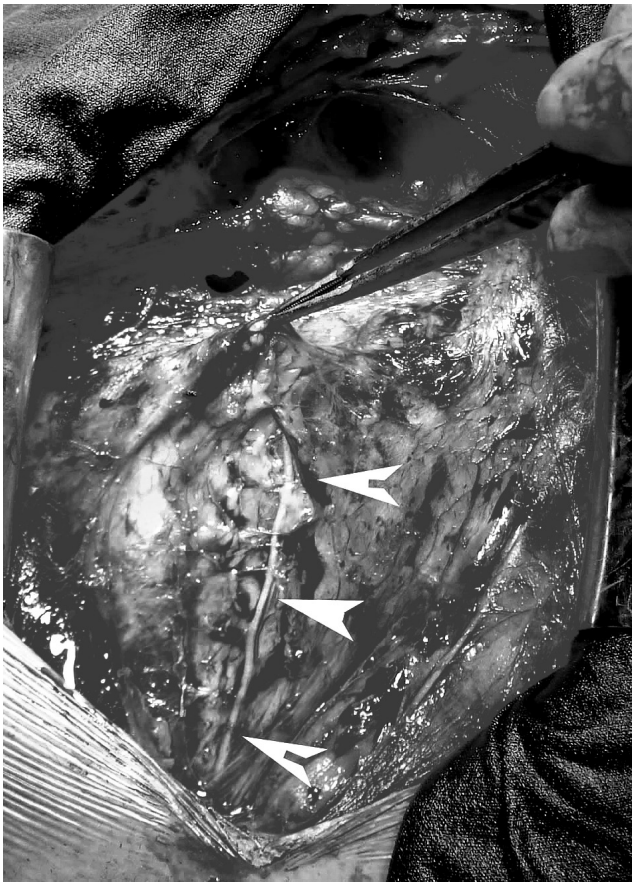

Fig. 2. The left phrenic nerve visualized as ventral to the heart, passing through the defective pericardial tissue

Discussion

In the absence of concomitant severe anomalies in patients with pentalogy of Cantrell, preoperative TTE may not be adequate to detect the congenital defects. Therefore, further imaging modalities might be necessary for the evaluation of these patients [3]. This was the case with our patient, as the absence of pericardium had not been diagnosed before surgery: the absence was only revealed after the median sternotomy. Further imaging modalities, such as computed tomography, might be useful in determining all the associated anomalies preoperatively. At this point, we believe that the pectus excavatum defect should be a warning sign of accompanying congenital defects.

The "midline supraumbilical abdominal wall defect" and "diaphragmatic pericardium" components of pentalogy of Cantrell were absent in our patient. If these two components had been present, a more complex surgical strategy would have been necessary [4]. In conclusion, we were lucky in the regard that the cardiac anomaly was easy to repair.

Another point that should be emphasized is that a defective course of the phrenic nerve, anterior to the heart, makes this nerve susceptible to damage during dissection, when the pericardial tissue is to be exposed for cardiac repair. The surgeons have to take great care not to damage the phrenic nerve while performing this dissection. We believe that this report of a rare clinical syndrome in adult congenital heart surgery will contribute to the literature.

The authors declare no conflict of interest.

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