

Congenital pericardial defect with Gerbode type septal defect in rotated heart: report of a case

Ubytek worka osierdziowego oraz ubytek w przegrodzie międzykomorowej typu Gerbode w zrotowanym sercu – opis przypadku



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Abstract

Asymptomatic pericardial defects are rare and found mostly incidentally during cardiac surgery. In one third of all cases absence of pericardium accompanies various congenital heart defects, and is diagnosed intraoperatively. We present a case of an 8-month-old male infant with Gerbode-type septal defect (LV-RA communication) with rightward rotation of the heart and left-sided partial pericardial defect, with pericardial adhesions and symptoms of left-sided pressure pneumothorax while opening the pericardial sac. The unique co-existence of Gerbode septal communication and position anomaly of the heart with congenital defect of the pericardium caused the effect of valvular air-trapping mechanism in the area of pericardial wall discontinuity that needed a change of the operative strategy prior to cardiopulmonary bypass. Taking into account the patient's past medical history the pericardial defect could be responsible for pericardial adhesions as a reaction to recurrent pulmonary infections spreading via persistent pleuro-pericardial communication.

Key words: congenital pericardial defect, Gerbode septal defect, congenital heart defects, paediatric cardiac surgery.

Background

Congenital pericardial defects are rare malformations with variable clinical presentations [1, 2]. Pericardial sac defects can be partial or complete, as it is commonly known in some mammals. The anomalies are still poorly known, the majority of literature reports are based on incidentally diagnosed cases, thus it is impossible to ascertain their total prevalence. Asymptomatic patients remain undiagnosed, otherwise the defects are discovered incidentally during cardiac surgery. The patients with congenital pericardial defects are referred for surgery for unrelated conditions, or the diagnosis is given postmortem [1]. To the best of our

Streszczenie

Bezobjawowe wrodzone ubytki osierdza rozpoznaje się przeważnie przypadkowo podczas zabiegów kardiochirurgicznych. W ok. 30% przypadków ubytkom osierdza towarzyszą inne wrodzone wady serca. W pracy zaprezentowano przypadek 8-miesięcznego chłopca z ubytkiem w przegrodzie międzykomorowej typu Gerbode (komunikacja lewa komora – prawy przedsionek) w zrotowanym sercu z historią przewlekłych infekcji dróg oddechowych. W trakcie zabiegu operacyjnego korekcji wady serca stwierdzono zrosty osierdziowe oraz lewostronną odmę opłucnową. Powstała ona po otwarciu worka osierdziowego w mechanizmie pułapki powietrznej przez istniejący ubytek osierdza w okolicy uszka lewego przedsionka. Zrosty osierdziowe mogły powstać na skutek komunikacji jamy opłucnowej lewej z workiem osierdziowym, która stanowiła potencjalną przyczynę odczynu osierdziowego na przebyte infekcje dróg oddechowych.

Słowa kluczowe: wrodzony ubytek osierdza, ubytek międzykomorowy typu Gerbode, wrodzone wady serca, kardiochirurgia dziecięca.

knowledge, we present the first case of a patient with congenital pericardial sac defect with congenital Gerbode type ventricular septal defect in an infant described in the literature. The case presented is twice as interesting because of preoperative diagnosis of Gerbode defect and necessary change of the operative strategy due to destabilizing pressure pneumothorax before cardiopulmonary bypass was commenced. Taking into account the patient's past medical history the pericardial defect could be responsible for pericardial adhesions as a reaction to recurrent pulmonary infections spreading via persistent pleuro-pericardial communication.

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Case report

An eight-month-old male infant; 7600 g birth weight, was admitted to the Department of Paediatric Cardiac Surgery, Mikotaj Kopernik Hospital in Gdańsk (Poland) with the diagnosis of ventricular septal defect (VSD) and hypothyreosis. The boy had a history of permanent respiratory tract infections prior to the admission with a remarkably bad clinical course. On admission his cardiac examination was remarkable for medial displacement of the apex right to the midclavicular line. Preoperative chest X-ray showed flattening and elongation of the left ventricular contour and right atrium enlargement (Fig. 1). ECG proved regular sinus rhythm with the right axis and partial right bundle branch block (PRBBB). Preoperative Echo examination showed 5.4 mm Gerbode VSD with left ventricle-right



Fig. 1. Preoperative chest X-ray showed flattening and elongation of the left ventricular contour and right atrium enlargement, with no symptoms that could indicate the presence of left-sided pericardial sac defects



Fig. 2. Preoperative Echo examination showed 5.4 mm Gerbode VSD (marked on the scan) with LV-RA communication and mild tricuspid regurgitation

atrium (LV-RA) communication and mild tricuspid regurgitation, the LV function and diameters of the heart chambers were normal (Fig. 2).

The patient was referred for scheduled ventricular septal defect repair. After classic median sternotomy and small thymus removal during the pericardiotomy unexpected pericardial adhesions in the area of ventricle body and right atrium (RA) and its appendage (RAA) were found. At the time of meticulous dissection there was growing left pressure pneumothorax with hemodynamic destabilization, bradycardia and pressure drop. The pneumothorax was relieved after insertion of an external suction line into the left pleura via the pericardial defect, further identified in the area of the left atrial appendage (LAA). The pericardial sac defect was functionally closed by the LAA causing air-trapping mechanism of the left pressure pneumothorax. The heart rotation (40 degrees) in the longitudinal heart axis was found with the left ventricle (LV) in the front and RA body situated deep in the pericardium and dorsally. The left pleura was opened with pleural adhesions as the remnants of recurrent pleural infections. The cardiac procedure was performed in cardiopulmonary bypass and moderate hypothermia with antegrade cardioplegic arrest, the heart was opened through RA incision. The Gerbode defect in the area of the tricuspid valve annulus with LV-RA communication was closed with running suture fresh autologous pericardial patch. The tricuspid valve inspection showed no dysfunction. The pericardial defect and pericardiotomy were left open. Postoperative course was uncomplicated. In the first postoperative echo after surgery there was a small LV-RA leakage that disappeared in further examinations. The child was discharged home on the 8th postoperative day in a good general condition. One year follow-up was uneventful.

Discussion

Congenital pericardial defects are rare findings with variable clinical presentation and can be complete or partial [1, 2]. These are still poorly known anomalies, sometimes overlooked, with a total number of less than 150 cases reported. Most frequently pericardial defects are left-sided (86%) and are related to premature atrophy of the left duct of Curvier during embryological development. Congenital complete pericardial absence is thought to be due to premature atrophy of the left common cardiac vein with insufficient blood supply to the pleuropericardium, which leads to its agenesis during pregnancy [1]. Pericardial sac defects appear three times more frequently in males than in females in a white-Caucasian population. Thirty percent of patients demonstrate associated congenital heart defects as the most frequent ones: atrial septal defect, bicuspid aortic valve, patent ductus arteriosus, tetralogy of Fallot, as well as others, like pulmonary sequestration, bronchogenic cysts, VATER syndrome (vertebral defects, anal atresia, tracheoesophageal fistula, radial and renal dysplasia), Marfan's syndrome, Pallister-Killian syndrome [1, 3-5]. Pericardial defects are more frequent in patients with congenital skeletal malformations (Holt-Oram syndrome) [2]. Complications

associated with pericardial defects depend on their extent. Complete absence of the entire pericardium or absence of the whole left or right side is usually associated with an excellent prognosis [6]. A small increase in preload may cause ventricular dilatation because of the loss of ventricular restraint, as it can be demonstrated in volume overload patients with pericardium left open after regular cardiac procedures. One case of severe tricuspid regurgitation due to cardiac hypermobility was reported and it required surgical intervention [6]. The partial absence is more dangerous. Entrapment of parts of the heart through defects may lead to strangulation of the atria, appendages and parts of the ventricles [7]. Congenital pericardial defect was reported with the presence of acute myocardial necrosis in an adult patient free of coronary artery disease, or there was evidence of impingement by a pericardial rim [8].

Clinical presentation of pericardial deficiency is non-specific. In most cases the defect is discovered incidentally in an asymptomatic patient [2]. There are reports of non-specific symptoms like mild cough, upper respiratory infection symptoms, brief chest wall throw during exercise usually not associated with chest pain, shortness of breath, palpitations, dyspnoea or respiratory distress leading to syncope [1, 8]. Sometimes the dyspnoea causes the "shifting heart" symptom, which is reported by selected patients. Physical examination may reveal significantly displaced apical pulse area, which may be palpated in the anterior or midaxillary line, as well as basal ejection murmurs, apical midsystolic clicks, or even systolic murmurs of undetermined origin. Patients may incidentally present some complications clearly related to the defect, like herniation and incarceration of the myocardium, predominantly the LAA and ventricles. All complications have been associated with presentations varying from chest pain to infarction, syncope, tricuspid regurgitation and sudden death [6-8]. The ECG is not pathognomonic and typically reveals bradycardia and right bundle branch block (RBBB), poor R-wave progression secondary to leftward displacement of precordial transitional zone is common, prominent P-waves in the mid-precordial leads denote right atrial overload. Echocardiographic findings are related to cardiac levoposition and increased mobility within the chest [8]. These include unusual echocardiographic windows marked change in cardiac position in the direction of the pericardial hole with changing patient position on the examination table (cardioptosis) and with abnormal cardiac cycle (swinging heart), with abnormal septal motion and false-positive appearance of the RV cavity dilation. Classic features of the chest X-ray include levoposition of the heart, confirmed by absence of the right heart border projecting on the right of the vertebral column and left cardiac border straightening and elongation (snoopy sign), as well as deep and very well-defined aortopulmonary window caused by the absence of the left pericardium and pleura allowing the left lung to invaginate into pericardial space [8]. Ultimately, the diagnosis is made or confirmed by CT scan or MRI. These diagnostic techniques not only prove initial diagnosis, but also deline-

ate an extent of the defect, which provides important information for further patient's management strategy [1, 9].

Therapeutic options are based on small, retrospective series which recommend surgical interventions only for patients with complications related to pericardial deficiency [1, 6]. These include both surgical closure or elongation of the pericardial defect with regard to initial problem caused by the pathology [2]. Asymptomatic left total defects usually do not require surgical treatment because of the small risk of circulatory complications, while the left-sided partial pericardial defects are more controversial. Small and moderate in size left-sided pericardial defects are considered for prophylactic surgery by some, while others suggest treating only symptomatic patients [2, 6, 8]. Otherwise there are many reports suggesting that both symptomatic and non-symptomatic patients should be followed by prophylactic operation to reduce the risk of death from cardiac structure herniation and incarceration. On the other hand, patients with partial absence of the pericardium (isolated); nowadays the focus is to manage the symptoms rather than prophylactic management. An interesting and still more and more available option is thoracoscopy intervention that provides safe and effective minimally invasive surgical intervention in the area of the pericardium as well as in the heart structures, possible even in a small patient [10].

In the presented case, we were challenged by the unique situation of a patient with Gerbode defect in a rotated heart and partial left pericardial defect, which was difficult to diagnose before surgery because of atypical position of the heart. Left-sided pericardial sac defects are usually seen with left cardiac border invagination to left pleural space in routine chest X-ray, although the heart borders were changed because of its irregular position. We also considered that both facts (partial absence of left pericardium and rightward heart axis change) might have not been related in embryonic development, thus we cannot exclude two independent factors causing right deviation and left-sided pericardial defect early in fetal life. Unexpected intraoperative complication made us modify the dissection technique nearby ECC. This complication gives another important argument in the discussion whether pericardial defect can be of special importance in selected cases.

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