Giant diverticulum of the right atrium

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Abstract

Congenital diverticula of the right atrium are very rare malformations. Frequently they are asymptomatic and are detected accidentally on a chest radiograph or during the autopsy. We present a case of symptomatic giant diverticulum of the right atrium in a 3-month old female. Major symptoms were atrial fibrillations with low cardiac output leading to cardiac arrest. She was qualified for urgent operation. To avoid bleeding hazard abdominal vessels were cannulated and atrioplasty performed in deep hypothermia. In the post-operative period the patient remained in sinus rhythm and the heart size normalized.

Key words: congenital heart disease, right atrial enlargement, right atrial diverticulum, treatment.

Streszczenie

Wrodzony uchyłek prawego przedsionka jest bardzo rzadką wadą serca. Często jest bezobjawowy i przypadkowo rozpoznany na zdjęciach rtg. klatki piersiowej lub podczas badań pośmiertnych. Przedstawiamy przypadek 3-miesięcznej dziewczynki z olbrzymim uchyłkiem prawego przedsionka. Uchyłek ten zajmował całe śródpiersie oraz lewą jamę opłucnową. Głównym objawem klinicznym było migotanie przedsionków z szybką czynnością komór, prowadzące do niskiego rzutu i zatrzymania czynności serca. Pacjentka została zakwalifikowana do pilnej operacji. W celu uniknięcia wykrwawienia się chorej zdecydowano o kaniulacji naczyń brzusznych i wykonaniu operacji w głębokiej hipotermii. Pacjentka po operacji w stanie ogólnym dobrym, z rytmem zatokowym została wypisana do domu.

Słowa kluczowe: choroby serca u dzieci, przerost prawego przedsionka, prawy uchyłek przedsionka, leczenie.

Case report

A 3-month old female patient was admitted to the paediatric department because of asphyxiation. The patient was redirected to the Cardiology Department because of gross cardiomegaly on the chest radiograph (Fig. 1). An echocardiography and computer tomography scan revealed a giant diverticulum with a wide communication to the right atrium. Cranially the diverticulum extended above the aortic arch and caudally it covered the right ventricle to the apex of the heart and the whole left side of the thorax. The left lung was completely depressed (Fig. 2); furthermore an atrial septal defect of secondary type (ASD II) with bi-directional shunt and persistent ductus arteriosus (PDA) with l-r shunt was diagnosed. With the exception of mild tricuspid regurgitation no other cardiac abnormalities were noted. Clinically the patient was very unstable; atrial fibrillation with ventricular tachyarrhythmia was dominant. In blood gas checkup retention of carbon dioxide was noted; therefore the patient had to be supported by mechanical ventilation and continuous infusion of amiodarone. Two episodes of cardiac arrest due to low cardiac output qualified the patient for urgent atrioplasty.

The potential hazard of extensive bleeding on sternotomy and symptoms of congestive cardiac failure persuaded us to conduct abdominal vessel cannulation. The abdomen was opened through a longitudinal incision along the linea alba. In the retroperitoneal region abdominal blood vessels were prepared for the cannulation (Fig. 3). The descending aorta was cannulated with a 10 Fr cannula just above the bifurcation and vena cava inferior with a 12 Fr cannula above the junction of the iliac veins. These cannulations were sufficient for the cooling phase and a hypothermic cardiopulmonary bypass was initiated. The sternotomy exposed a huge thin-walled diverticulum sac occupying almost the whole thorax (Fig. 4). Blood was partly drained to the venous reservoir and the diverticulum sac was gently pushed

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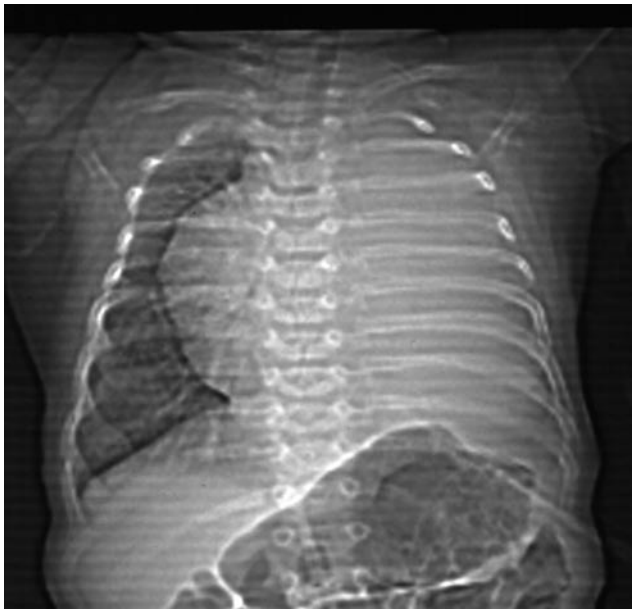

Fig. 1. Preoperative chest radiograph showing gross cardiomegaly

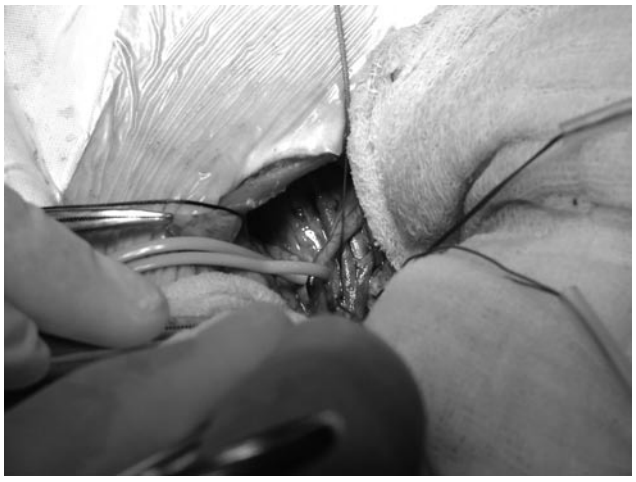

Fig. 3. Preparation of abdominal vessels for cannulation

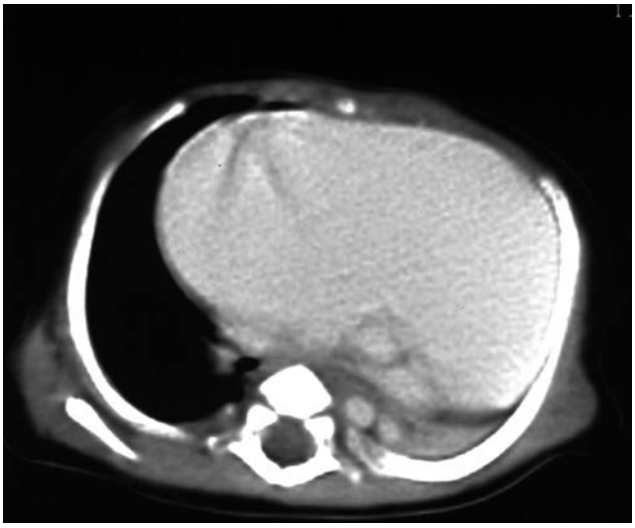

Fig. 2. CT scan of the same patient showing the extent of the diverticulum and the left lung depressed by it

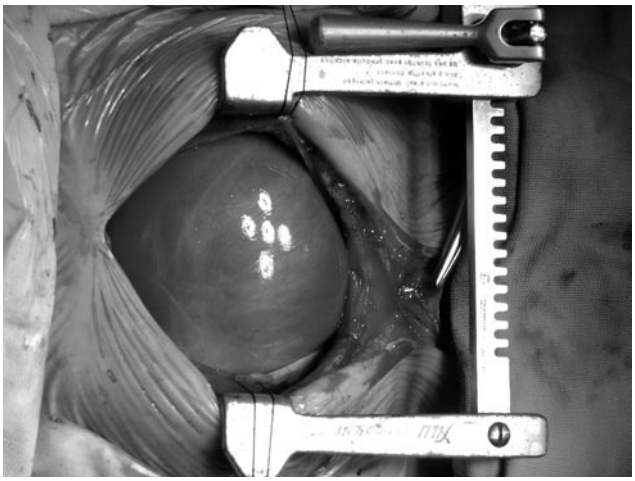

Fig. 4. Intra-operative picture of the diverticulum

aside to expose the great vessels, after which PDA was prepared and ligated. On reaching 18°C in the rectum, the ascending aorta was cross-clamped; secondly, cold blood cardioplegic solution was administered to the aortic root; and thirdly, blood was drained to the venous reservoir. Circulatory pumps were stopped and the "paper thin" diverticulum sac was opened, which revealed a finely trabeculated translucent atrial wall. Following this, ASD II was closed with a direct suture. The tricuspid valve was morphologically normal. The diverticulum sac was excised and direct suture was employed for the atrioplasty. Reperfusion and rewarming was commenced after cannulation of the ascending aorta and the right atrium; meanwhile cannulas from the abdominal vessels were removed. The patient was weaned off cardiopulmonary bypass with minimal inotropic support. Total cardiopulmonary bypass time was 173 min and aortic cross-clamp time (circulatory arrest) 28 min.

The patient was kept under sedation for two days because of deep hypothermia with circulatory arrest. The postoperative radiograph showed a normal size heart (Fig. 5). The circulatory system was stable with sinus rhythm.

Discussion

Diverticulum of the right atrium is a very rare congenital malformation. Bailey was the first to report such a case in 1955 [1] and surgical resection of the right atrial diverticulum was described by Morrow in 1968 [2]. Recently, a publication reported all cases in the literature from 1955 to 1998. Of 105 reported cases, only 60 were congenital diverticulum of the right atrium [3]; other cases were diverticulum of the coronary sinus and single or multiple saccular diverticulum of the right atrium. The incidence of such malformation is difficult to estimate, especially in asymptomatic patients as many of them (48%) remain asymptomatic. They are sometimes accidentally diagnosed because of gross cardiomegaly during a routine chest radiograph or discovered during the autopsy.

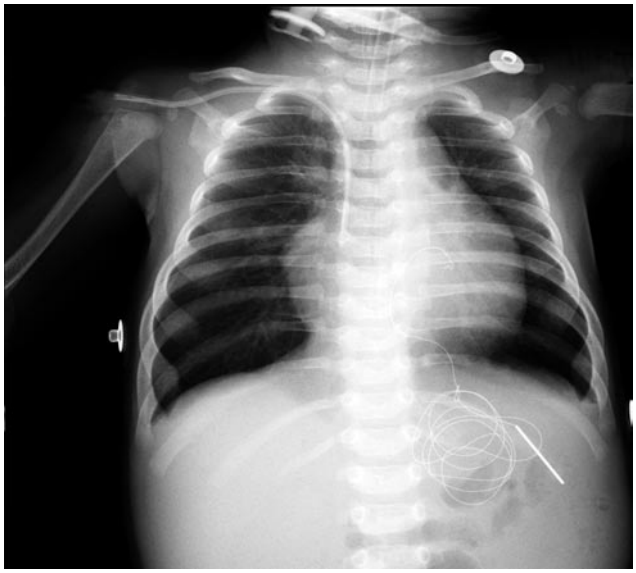

Fig. 5. Intra-operative picture of the diverticulum

Other cardiac conditions such as Ebstein anomaly, pericardial effusion, pericardial cysts and cardiac tumors can also mimic gross cardiomegaly on a chest radiograph [4]. Accurate diagnosis and differentiation from other cardiac anomalies should be considered before medical or surgical approach. In symptomatic patients, atrial arrhythmia, respiratory distress, jugular engorgement, oedema of the ankles and hepatomegaly are the major complaints [3].

Surgical atrioplasty is advised in symptomatic patients. The low operative mortality and high success rate justify the surgical approach [3] and patients with atrial fibrillation have been treated successfully with atrioplasty [5, 6]. Even in asymptomatic patients surgery should be considered because of the high risk of thrombus formation in the right atrium or the likelihood of developing atrial fibrillation. Some authors suggest that only symptomatic patients should be operated on; in the absence of symptoms anticoagulation as a precaution against thromboembolization should be considered [3, 7]. But these patients in future may develop atrial fibrillation even after surgical cryoablation [8]. Sudden deaths have also been reported [9].

In conclusion, right atrial diverticulum should be treated surgically to prevent rhythm disturbances, potential thrombus embolism, atrial rupture or congestive cardiac failure.

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