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Postpartum sudden death by right atrial cyst

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Heart cysts are usually congenital. They are asymptomatic and thus found incidentally in relation to other conditions. Sometimes their size determines life-threatening complications. Surgical treatment is successful. We represent a case with a fatal outcome in a 28-years-old woman who had given birth by Caesarean section. The patient had been hospitalized 13 days after delivery in relation to sudden chest pain, fever and ultrasonographic (USG) examination that revealed pericardial effusion and a cystic formation of the right atrium. She had died suddenly on the second night of hospitalization before transfer into cardiac surgery department for further diagnostic procedures and surgical treatment. At autopsy a ruptured cyst of the right atrium and interatrial septum with relative tricuspid valve stenosis, pericarditis, myocarditis, haemorrhage in rupture and cardiac tamponade had been found.

Introduction

Giant Blood Cyst (GBC) [1, 2, 3, 4, 5] are usually congenital, asymptomatic affecting mostly valvular structures. They are rarely seen after the 2 years of age as they spontaneously regress over time, although in some cases they are found in adults [4, 5]. The most common size of GBC is microscopic [1], but sometimes their large size determines complications – valvular and ventricular dysfunction, thromboembolic myocardial infarction, pulmonary embolism or obstruction of the coronary arteries. The most common treatment is surgery [2, 3], after which there is a full recovery. [5]

Clinicomorphological case report

We present a case of a 28-y-o woman who had been admitted to Cardiology clinic with complaints of increasing during inhalation chest pain and fever up to 38,5°C that started suddenly the day before hospitalization. On the day of admission a cardiologist had found echocardiographic data of pericardial effusion and cystic formation in the

right atrium and directed her to hospital for further treatment. Thirteen days before that she had delivered a child by C-section for reasons of fetal distress. She had been taken L-thyroxine for hypothyroidism in Hashimoto's thyroiditis for a period of one year. At the time of admission her general condition had been satisfactory, with low-grade fever of 37,6°C and sounds of pericardial friction. The new echocardiography showed 300 ml of pericardial effusion and thin-wall cyst in the right atrium with a diameter of 65 mm, occupying about 50% of atrial volume, not participating in blood circulation, covering a large part of the interatrial septum near the tricuspid valve protruding into the left atrium, causing relative tricuspid stenosis. There had been serological evidence of adenovirus infection – anti-adenovirus IgM (+)/IgG (+), and exacerbated chronic pyelonephritis.

After the start of medication an improvement of symptoms had been noticed. Series of consultations had been performed. MRI and transfer to the Department of Cardiac Surgery had been planned, but prior to that, on the second night of the stay, the nurse on duty found the patient with no signs of respiration and heartbeat. Full CPR had been performed, but without success.

At autopsy 850 ml of bloody fluid and blood clots was found in the pericardial sac. The heart was enlarged, soft weighing 450 g. The right atrium was dilated, with a cystic formation with a diameter of 6.5 cm, 2-3 mm thick wall, located in the interatrial septum and front atrial wall, protruding to the atrioventricular tract. When opened, a 15 mm slit-like laceration on the right auricle wall was found, corresponding to a 7 mm laceration and haemorrhages on the outer surface of the auricula. This laceration communicates with the pericardial sac. No contents of the cyst was found. Histologically in the myocardium was found a diffuse lymphocytic inflammatory infiltrate in the rupture

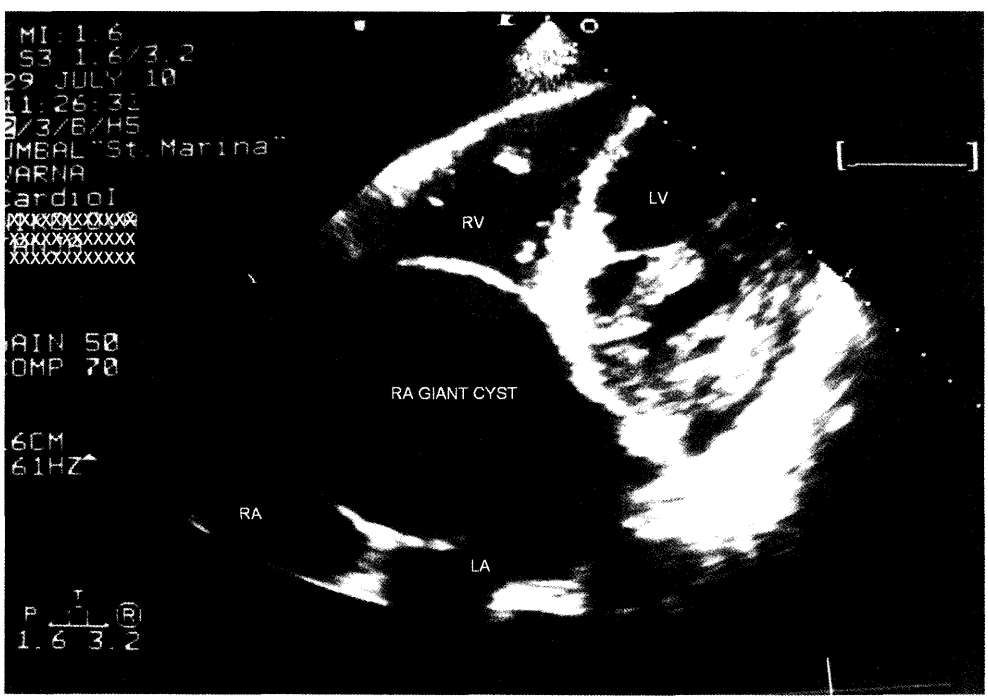

Fig. 1. Echocardiographic image of a cyst of the right atrium.

Fig. 2. Gross appearance of the cyst protruding over the tricuspid valve, which caused relative valvular stenosis.

Fig. 3. Gross appearance of the cyst – notice the granular inner surface.

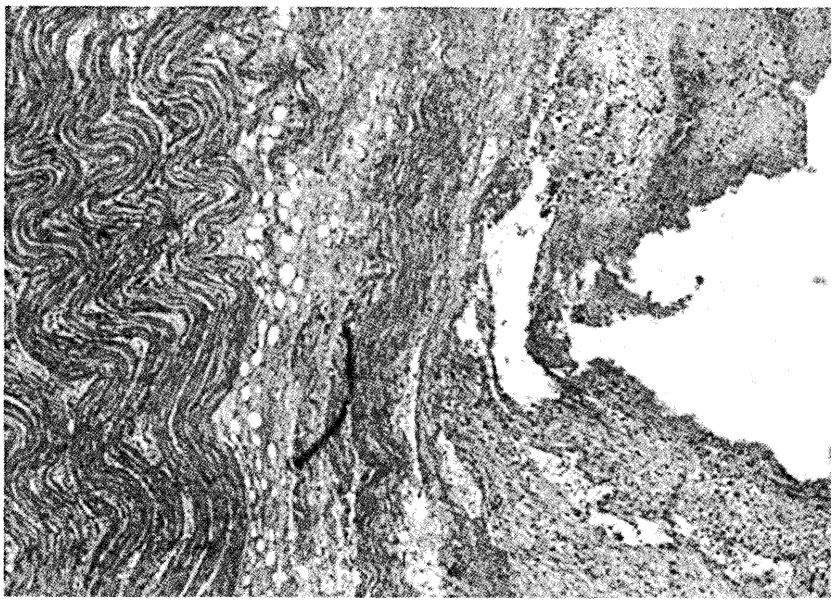

Fig. 4. Part of the cyst (right) area with a laceration and lymphocytic inflammatory infiltration between the myocardial fibres in the cyst wall.

area with formation of granulation tissue. The visceral pericardium was infiltrated with non-specific polymorphonuclear inflammatory cells.

Edema and single hemosiderin-laden macrophages were found in the lungs. The organs of the great circle of circulation were congested. Uterus showed early changes of birth delivery by C-section and a small, clinically insignificant myomatous nodule.

The cause of death was an acute heart failure after cardiac tamponade due to rupture of the cyst causing atrial haemorrhage and leakage of it's contents into the pericardial sac.

Another interesting fact is that the fetus had prenatal cardiac rhythm and conduction disorders. After birth tumor formations in the septum of the heart of the infant were found after USG examination, identified as rhabdomyomas, situated in the apical part of the interventricular septum on the right and the left ventricular surface, with maximum size of 12/5 mm, without hemodynamic significance. Bradicardic heart rate of 70-80 bpm had been found, due to blocked extrasystolic bigemeny. Reduction of the extrasystoly and normalization of heart rate had been achieved with age. Transfontanel USG have not shown any changes in the brain. The follow-up physical development of the child was satisfactory, without heart failure and cyanosis, persisting supraventricular extrasystoles and reductoin of cardiac tumors' size. At last follow-up examination the relatives reported of absence seizures, staring and temporary loss of consciousness. The child had been consulted with a pediatric neurologist and had been hospitalized for suspection of tuberous sclerosis and performance of brain CT and EEG.

Discussion

This case raises interesting issues for discussion – whether it is a hereditary heart abnormality or damage to the heart of the mother and the fetus in relation to the adenovirus infection found before. A positive IgM means an active infection which was confirmed morphologically, but positive IgG implies chronic viral disease which could be associated with both mother and child's heart conditions.

Another interesting question is whether there is a link between heart disorders of mother and infant to the clinical manifestation and treatment of Hashimoto's thyroiditis. It could be assumed that the thyroid hormonal imbalance (due to the lower than conventional dose of L-thyroxine that the mother had been taking) may lead to changes in the heart morphology and function of the fetus. A fact, indirectly supporting this assumption, is the regression of cardiac formations in the baby with age.

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