COMPLEX HEART MALFORMATION IN A SIX WEEKS CHILD. CASE REPORT

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ABSTRACT

The aim of our study is to present the case of a six weeks old infant, prematurely born, who was admitted to our hospital with systolic murmur and cyanosis. The diagnosis of cor triatriatum sinister associated with patent foramen ovale has been established by ultrasound examination. The infant died a month after admission. The necropsy confirmed the clinical diagnosis and also revealed the hypertrophy and dilation of the right heart, left ventricle hypoplasia, interstitial pneumonia and amniotic fluid aspiration bronchopneumonia. The possible cause of death in this case was right heart failure following pulmonary hypertension associated with acute respiratory failure. To our knowledge it is the first case with such a complex malformation reported in a premature neonate in our hospital.

Key words: *cor triatriatum sinister, patent foramen ovale, infant, heart defects.*

INTRODUCTION

Cor triatriatum represents less than 0,5 % of cardiac congenital defects, with a higher frequency in patients under the age of 6 months [1 Alphonso, 2005, 2 Werner, 2007 , 3 Schiller, 2012, 4 Ezeogu, 2013].

This malformation is characterised by a divided left atrium, the anterior part communicating with the left ventricle, while the pulmonary veins are connected with the posterior one, creating an obstacle at the level of pulmonary circulation [5 Slight, 2003, 6 Serban, 2006, 7 Su, 2008, 8 Gonzalez-Ramirez, 2012, 9 Mendez, 2013, 10 Bezgin, 2014, 11 Ríos-Méndez, 2015].

Depending on the size of the holes, the patient could be or not symptomatic, the evolution being favourable or leading to death [5 Slight, 2003].We present in this paper a case with a rare association of cor triatriatum sinister with hypoplastic syndrome diagnosed in an infant who was born prematurely.

CASE REPORT

A 6 weeks old, male patient was admitted in Clinical Children Hospital "St. Mary" Iasi with the following diagnosis: premature newborn, jaundice of prematurity. For 2 weeks he was hospitalized in the Premature Department. The clinical examination revealed a systolic precordial heart murmur and perioral cyanosis. The electrocardiography indicated sinusal tachycardia and ventricular repolarization. The ultrasound scan showed a well visible intra-atrial diaphragm, relative hypoplasia of left ventricle, foramen ovale patent, and possibly a small septal ventricular defect. The evolution was favourable and he was admitted to the Paediatric Recovery Department. After 2 weeks, the patient's status got worse. The evaluation of the vital signs revealed: perioronasal and subungual cyanosis, peripheral vasoconstriction with cold extremities, dyspnea with tachypnea, irregular breathing and the intensification of the systolic murmurs. After a week of treatment with oxygen therapy, there was no amelioration of the general status and since an intercurrent infection appeared he was transferred to the Intensive Care Department where the oxygen therapy continued and he received antibiotics. Five days after admission he died with the following clinical diagnosis: bronchopneumonia, complex congenital cardiac malformation (cor triatriatum, foramen ovale patent), cardiac failure, stage I prematurity. After the decease we performed the necropsy in the Pathology Laboratory of Clinical Children Hospital "St. Mary" Iasi.

The necropsy

The macroscopic examination

At the external examination, the cadaver's skin was cyanotic with a higher intensity at the level of extremities. By examining the brain, we found cerebral edema and haemorrhagic petechiae.

By opening the thorax we observed numerous haemorrhagic spots on the lung and heart surface, in the adventitia of pulmonary artery and on the posterior visceral pericardium. The heart was globally enlarged (Fig. 1) due to the dilation and hypertrophy of the right ventricle and the dilation of the right atrium with the enlargement of atrioventricular foramen (Fig. 2). The left ventricle was hypoplasic with a thin wall. Foramen ovale was permeable, with a large diameter of about 1 cm. The pulmonary veins drained into a venous retropericardiac cavity that communicated with the left atrium (Fig. 3). The examination of the abdomen revealed an accessory spleen of 1 mm in diameter.

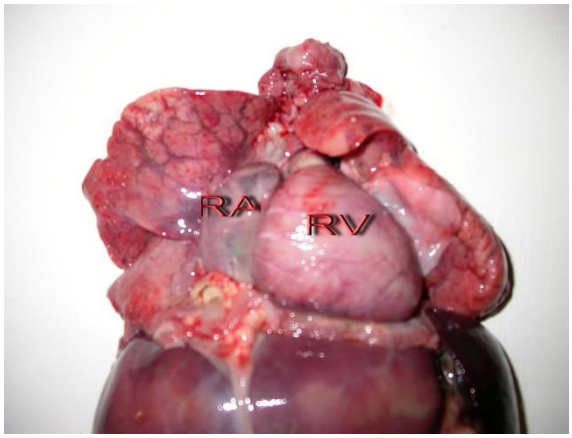

Figure 1. Cardiomegaly by dilation of right cavities

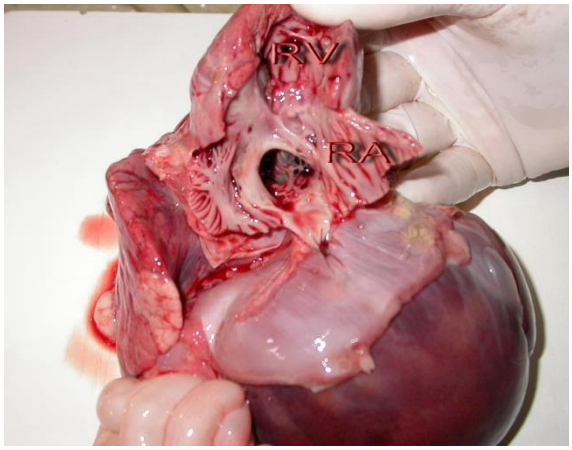

Figure 2. Dilation of the right cavities

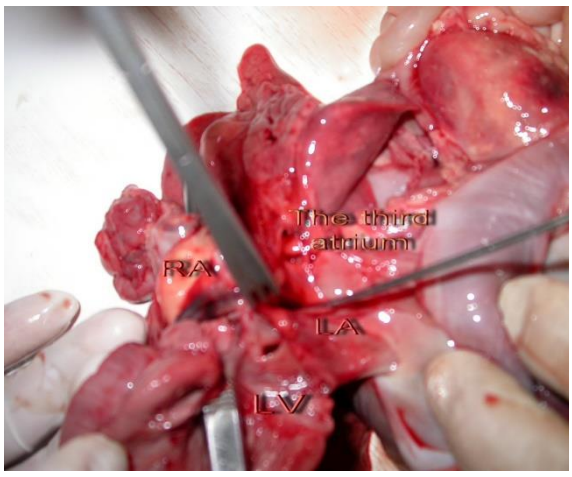

Figure 3. The third atrium

The microscopic examination

The fragments obtained during the necropsy were fixed in 10% buffered formalin and embedded in paraffin. The histologic sections were stained with haematoxylin-eosin. The microscopic examination revealed small arterioles with a thick adventitia and muscular layer (due to passive pulmonary hypertension). The lung showed alveolar macrophages, stasis and mononuclear inflammatory reaction along septa. Some alveoli contained amniotic liquid elements. We also found an accessory spleen included in the pancreas.

DISCUSSION

Cor triatriatum sinister was discovered for the first time by Church, in 1868 [1 Alphonso, 2005, 3 Schiller, 2012] and data from scientific literature have shown that it could be surgically treatable if it is diagnosed in time by the clinician [12 Ozyuksel, 2015]. It is a rare malformation, representing about 0,1 % of all the congenital heart malformations identified during life [1 Alphonso, 2005, 2 Werner 2007, 7 Su, 2003, 5 Slight, 2003, 13 Bassil, 2006, 4 Ezeogu, 2013, 9 Mendez, 2013] and 0, 4 % of the post-mortem malformations [13 Bassil, 2006, 3 Schiller, 2012]. It could appear isolated or in association with other cardiac anomalies such as: atrial septal defect (especially foramen ovale patent), mitral regurgitation, dissecting aneurysm [7 Su, 2003, 14 de Belder, 1992, 10 Bezgin, 2014], ventricular septal defect, partial anomalous pulmonary venous connection (15 Khan, 2012, 4 Ezeogu, 2013),

hypoplastic left heart syndrome [15 Khan, 2012], Fallot tetralogy [4 Ezeogu, 2013] or it could be associated with a left superior vena cava originating in the coronary sinus [16 Chen, 1999, 4 Ezeogu, 2013]. There are a few data in the scientific literature which can explain this congenital malformation from embryological development perspective. The common explanation is that the malformation appears due to the failure of incorporating the common pulmonary vein in the left atrium [17 Anderson, 1992, 5 Slight, 2003, 16 Chen].

In the case of our patient, the dividing of the left atrium into two cavities together with the communication between atria, led to right heart failure and pulmonary stasis by reduction of blood flow. On the other hand the incomplete development of the left ventricle gave rise to decrease in blood flow in the systemic circulation. The final result of this malformation was cyanosis, especially in the lower limbs and a decreased oxygenation of the vital organs: brain, kidneys. The overload of pulmonary circulation led to pulmonary hypertension associated with congestion and microhemorrhages in the lung and consecutive hypertrophy and dilation of the right cavities. Similar to other scientific studies [15 Khan, 2012], our patient died after a very short time due to heart failure and intercurrent pulmonary infection.

CONCLUSION

The division of the left atrium into two cavities with the inclusion of the pulmonary veins in one of them communicating with the real atrium causes an obstacle to the pulmonary circulation. These changes induce pulmonary hypertension, hypertrophy and dilatation of the right ventricle with increased pressure in it and also right-left shunt through the patent foramen ovale leading to cyanosis. On the other hand, interstitial pneumonia and bronchopneumonia due to amniotic fluid aspiration caused alveolar capillary block and acute respiratory distress.

Although at present there are methods of surgical treatment of this anomaly, the very young age, the prematurity and the accompanying complications (interstitial pneumonia, amniotic fluid aspiration bronchopneumonia) did not allow the surgical correction in this case

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