Single Ventricle Cardiac Defect

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Abstract

Single ventricle heart is defined as a rare cardiac abnormality with a single ventricle chamber involving diverse functional and physiological defects. Our case is of a ten month-old baby boy who died shortly after admission to the hospital due to vomiting and diarrhoea. Autopsy findings revealed cyanosis of finger nails and ears. Internal examination revealed; large heart, weighing 60 grams, single ventricle, without a septum and upper membranous part. Single ventricle is a rare pathology, hence, this paper aims to discuss this case from a medico-legal point of view.

Introduction

Single ventricle heart is defined as a rare cardiac abnormality with a single ventricle chamber involving diverse functional and physiological defects.[1-3] It has been reported to co-exist with anomalies like mitral valvular ring,1 and tricuspid atresia.[2] Single ventricle is a rare pathology, hence, this paper aims to discuss this case from a medico-legal point of view.

Case Report

A ten months old baby boy died shortly after admission to the hospital due to vomitting and diarrhoea. Baby was 64 cm in length and had a weight of 5680g. Autopsy findings revealed cyanosis of finger nails and ears on external examination. In the internal examination; heart was found to be large weighing 60g (Figure-1)

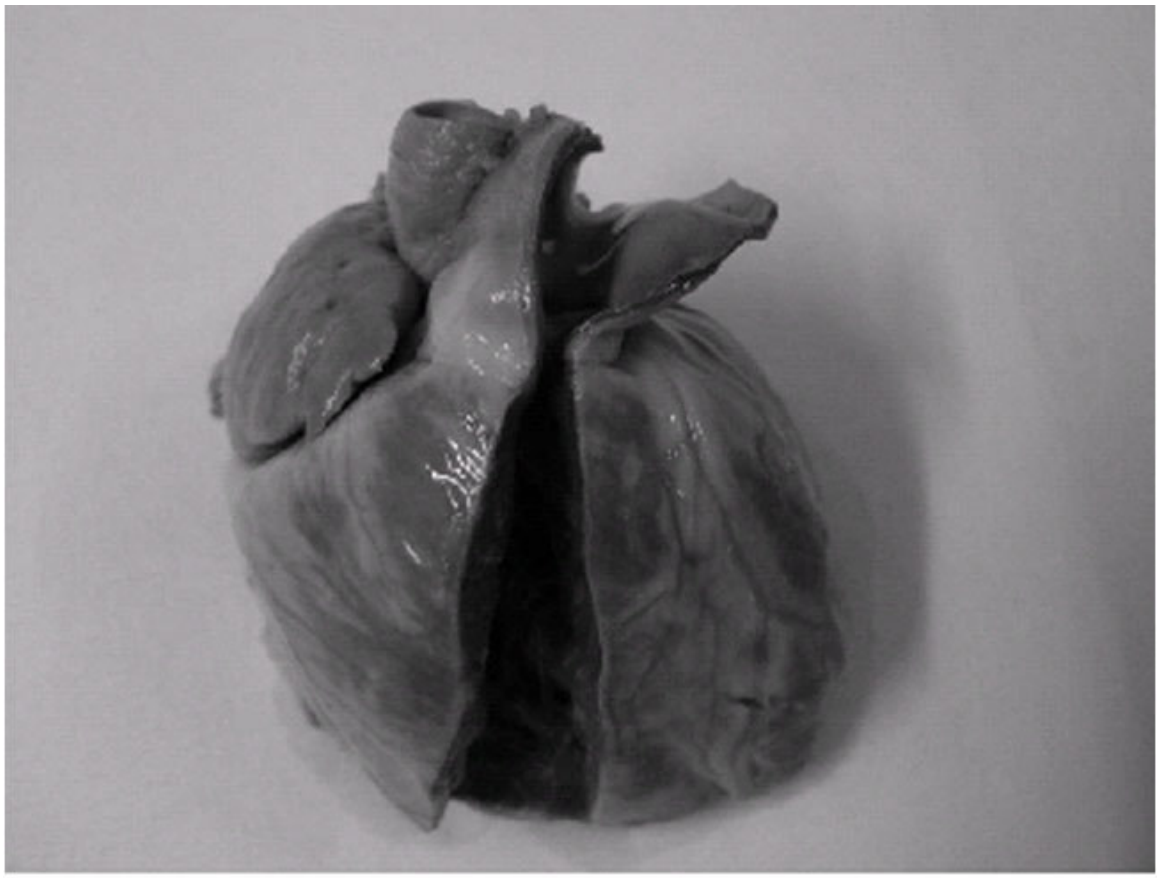

Figure-1: Macroscopic appearance of the heart.

with a single ventricle, and only upper membranous septum. Right atrium and auricula were prominent. Left atrium and auricula were hypoplastic. Dissection of the heart showed Atrial Septal Defect (ASD) separated by membranous band, and a single ventricle without ventricular septum except the upper membranous part was observed (Figure-2).

Figure-2: Single Ventricle.

Right ventricle was rudimentary, anterior tricuspid and septal valves were found, papillary muscles and tendineal cords extended into the left wall of the ventricle. Microscopic evaluations showed hypertrophic changes in cardiac muscles and oedema in the pulmonary tissue. Toxicological analysis of blood, organ tissues and stomach ingredients were negative. The cause of death was reported as heart failure due to cardiac abnormality.

Discussion

Single ventricle heart is defined as a rare cardiac abnormality with a single ventricle chamber involving diverse functional and physiological defects.[1-3] Weigel et al.[2] consider uni-ventricular heart cases in four distinct groups such as: (a) double-inlet left ventricle; (b) complex univentricular heart with single or common inlet, or with a ventricle of common or right ventricular morphology; (c) complex univentricular heart with asplenia; and (d) complex univentricular heart with polysplenia. When we evaluate our case according to Weigel's morphological classification, our case is in the complex univentricular heart with single or common inlet, or with a ventricle of common or right ventricular morphology group with no abnormality in the spleen. Besides, univentricular heart has been reported to co-exist with different anomalies like mitral valvular ring,[1] and

tricuspit atresia.[2] Despite the fact that there are contradictions on the classification of single ventricle heart defects; similar to our case, it was reported that instead of existence of a single ventricle, it is more likely to be rudimentary right ventricle with two atrioventricular entrances.[4] The studies regarding the relationship between univentricular heart and ASD among inter family marriages, show that multi-factorial genetics play a role, but no co-relation between these two anomalies has been demonstrated.[5]
Physiological changes in these cases include obstruction of pulmonary and systemic circulation, and also increased resistance in pulmonary vessels. Treatment plans comprise considerations and evaluations of physiological states.[3] Prognosis mainly depends on severity of pulmonary circulation, and surgical procedures such as Mustard, Senning, Norwood or modified Fonton operations are favoured.[3,6] In their experimental animal models Ricci et al.[7] support the hypothesis that cerebral injury in single-ventricle heart models are due to insufficient oxygenation of the brain.
From medico-legal aspect, careful evaluation and post-mortem detection of cardiac abnormalities in postnatal life is crucial in order to understand sudden cardiac deaths in newborns.

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