G41 Lymphogenic Cardiomyopathy: A Possible Cause of Non-Immune Fetal Hydrops

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After attending this presentation, attendees will gain knowledge of three particular cases of congenital and isolated cardiac lymphangiectasia manifested *in utero* with cardiac failure and hydrops.

This presentation will impact the forensic community by exploring how cardiac isolated lymphangiectasia might represent a new nosological entity that should be included among the primary cardiomyopathies (lymphogenic cardiomyopathy). Consequently, this entity should be investigated among the possible causes of non-immune hydrops foetalis (HF).

It is the intent of this presentation that cardiac isolated lymphangiectasia might represent a new nosological entity that should be included among the primary cardiomyopathies (lymphogenic cardiomyopathy). Consequently, this entity should be investigated among the possible causes of non-immune hydrops foetalis.

HF is an aspecific and terminal sign of many fetal diseases that could be observed at any time during pregnancy. In fully developed HF, there is subcutaneous oedema with fluid accumulations in peritoneal, pleural, and pericardial cavities. The umbilical cord and placenta are also oedematous and there is polyhydramnios. In the early stages of HF, the fluid accumulations are not present in all compartments. HF is caused by three main mechanisms: anemia, hypoproteinemia, and cardiac failure. Most cases fit within this classification, although some cases remain unsolved under the name of "idiopathic HF." Another classification divides HF into treatable (27%) and untreatable (73%) forms. The success of isoimmunization prevention programs demonstrated that most cases of HF are now non-immune and depend on cardiovascular diseases (22%), chromosomal abnormalities (13%), thoracic causes (10%), anemia (homozygous α-thalassemia), monochorionic twinning (6%), infections (5%), miscellaneous (16%), not determined (20%). Cardiovascular HF seems to be more frequently associated with structural and functional abnormalities that cause volume and/or pressure overload on the right atrium such as left heart syndrome, arrhythmias, myocarditis, cardiomyopathies, cardiac tumors, myocardial infarction, and arterial calcification.

Three unusual cases of congenital and isolated cardiac lymphangiectasia (ICL) manifested *in utero* with cardiac failure and hydrops will be presented.

Case1: A male hydropic fetus with a gestational age of 14.2 weeks without dysmorphia. The mother was 32-years-old and had four pregnancies, one of them resulting in miscarriage due to unknown causes. Ultrasound of the fetus and placenta showed regular heart rate with biventricular hypocontractility and without congenital cardiac and extra-cardiac defects and polyhydramnios. Amniocentesis revealed a normal karyotype.

Case 2: A male hydropic fetus with a gestational age of 19.5 weeks without dysmorphia. The mother was 29-years-old and had a previous miscarriage due to a premature rupture of the placental membranes (acute chorioamnionitis) at the 25th week of gestation. Ultrasound of the fetus and placenta showed regular heart rate with biventricular hypocontractility, without congenital cardiac, and extra-cardiac defects, and polyhydramnios. Amniocentesis revealed a normal karyotype.

Case 3: A female non-hydropic fetus with a gestational age of 22 weeks without dysmorphia, except for the presence of a single head held plica. The mother, 27-years-old, was at first pregnancy. Ultrasound of the fetus and placenta showed light pericardial effusions, regular heart rate with biventricular hypocontractility, without congenital cardiac and extra-cardiac defects, and polyhydramnios. Amniocentesis revealed Trisomia 21.

In all cases, fetal autopsies showed ultrasound findings conducted during pregnancy. At histology, the organs were normally structured except for the heart that showed a "moth-eaten" aspect in the ventricular walls, due to severe, diffuse and transmural lymphangiectasia and interstitial lymphedema. The interposed myocardium resulted compressed, distorted, trabeculated, and with multifocal patchy coagulative miofibrillolisis (contraction band necrosis). The morphological examination in situ of apoptosis highlighted in all cases the presence of frequent apoptotic events in the endothelia of small arteries and veins.

Discussion: ICL is an extremely rare entity and up-to-date the literature reports describe only one case characterized by septal localization of this lesion causing septal hypertrophy and left ventricular outflow obstruction mimicking hypertrophic cardiomyopathy. These cases represent the first report of ICL involving diffusely the heart, causing cardiac failure and hydrops of various degrees. The findings of a marked apoptosis in the endothelial cells of blood vessels suggest that interstitial lymphoedema and lymphatic overload is due to increase vascular permeability of the cardiac blood microcirculation.

Hydrops Fetalis, Lymphangiectasia, Cardiomyopathy

 ** Presenting Author*