A Case of Sudden Death of Pregnant Woman with Complicated Endocardial Fibroelastosis

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Abstract
We present a rare case of thirty-two-year-old woman in her third pregnancy with a developed sudden death in the form of acute cardiac rhythm disturbance at 12 weeks of her third gestation due to complicated form of endocardial fibroelastosis of the left ventricle. The patient was followed by a cardiologist about congenital insufficiency of the mitral valve since childhood and periodically complained about frequent or rare pain in the heart, palpitations and fatigue. However, the death occurred suddenly and was not the result of pregnancy complications, but the result of severe irreversible pathological changes in the heart on the ground of congenital heart disease. The presented case is of a certain interest for cardiologists, obstetricians and gynecologists.

Keywords
Endocardial Fibroelastosis; Cardiomyopathies; Heart Diseases; Pregnancy; Pregnancy Complications; Maternal Mortality

Abbreviations
EFE – Endocardial Fibroelastosis
LV – Left Ventricle
LVHT – Left Ventricular Hypertrabeculation

Introduction
Endocardial fibroelastosis (EFE) is a pathologic condition of abnormal deposition of collagen and elastin within the endocardium of the heart [1]. Endocardial fibroelastosis is characterized by diffuse thickening of ventricular endocardium (with the help of collagen and elastic fibers) and associated myocardial dysfunction [2, 3]. It can be primary (idiopathic) or secondary to structural heart diseases [4], in infancy or early childhood, this progressive process, usually leads to a heart failure.

Primary EFE is characterized by a dilated left ventricle (LV) that typically has a high takeoff of the papillary muscles and thickening of the free edge of the mitral valve leaflets, in addition to diffuse thickening of the endocardium by aortic-like thick and parallel elastic fibers [2]. In the past, EFE was referred to rare cardiomyopathies, but in the latest classification of the American Heart Association (2006), EFE as cardiomyopathy was not mentioned [5].

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Endocardial fibroelastosis (EFE) is a unique form of fibrosis, which forms a de novo subendocardial tissue layer encapsulating the myocardium and stunting its growth, and which is typically associated with congenital heart diseases of heterogeneous origin, such as hypoplastic left heart syndrome [3].

Often, EPE is an important cause of congestive heart failure and, as a result, antenatal fetal death or infant death in the early neonatal period [6, 7]. Not infrequently, this pathology is associated with non-immune fetal dropsy [4]. Despite the fact that the prognosis for newborns with hypoplasia of the left heart and EFE over the last decade has significantly improved, the lethality still remains high [5, 8].

Progressive ventricular dilation and ventricular dysfunction are factors of unfavorable prognosis. Stable, unregulated ventricular tachycardia in these patients can be life-threatening complication that requires mechanical support and even heart transplantation [9].

Endocardial fibroelastosis is a disease detectable at a frequency of 1: 70,000, which is characterized by the presence of the diffusely thickened endocardium on the inner surface of the valves and ventricles. Death in this disease occurs as a result of decompensation of cardiac activity with myogenic dilatation of the heart cavities.

Case report

We observed a rare case of sudden death of pregnant woman D., 32 years old, mother of 2 children (cesarean sections for two full-term pregnancies in 2004, 2010), who had an endocardial fibroelastosis complicated by failure in the conduction system of the heart with mitral valve damage and the conduction system of the heart up to pathologic anatomical study.

D. since childhood was registered with a cardiologist about congenital insufficiency of the mitral valve: prolapse, cleavage of the anterior leaflet, prolapse of the posterior leaflet of the mitral valve, ventricle regurgitation. Periodically complained on frequent or rare pain in the heart, palpitations, fatigue; pain in the heart of aching or piercing nature, palpitations with attacks.

From a photocopy of the D.’s emergency case report: came to the office, suddenly fell down and turned cyanotic. The age of gestation - the first trimester of pregnancy.

While pathologic anatomical study - the heart of a regular shape, measuring $11 \times 10.5 \times 5.5$ cm, weighing 350 gram, the tip is moderately pointed. The extreme right type of arterial blood supply of the heart. The perimeter of the mitral valve is 14 cm. Leaflets of the mitral valve on its free edges, moderately deformed, tightly-elastic consistency. Posterior leaflet with minimal focal changes, in a small area is adjacent to the thickened anterior leaflet. At the same time, the modified mitral valve leaflets are unevenly thickened and indurated in their free margins, in some places in the middle part of the anterior leaflet, while in the base of the valve the walls are thin, the surfaces are grayish-pink, flat, shiny, smooth and resiliently-dense, without impositions and deformations. The posterior flap of this valve is structured correctly, without a tissue defect; no thickening and deformations are found on most part of the flap. The anterior flap of the mitral valve has a splitting from the free edge almost to the base of the leaflet in the form of an acute angle, with rounded and thickened edges, without impositions, chords are thickened. The anterolateral wall of the left ventricle from the base to the apex of the heart is thickened, indurated grayish-whitish and whitish-pinkish, shiny, without impositions. The thickness of the left ventricle’s wall at the base of the heart is 0.7 cm. The thickness of the left ventricle’s wall in the middle third is 1.7 cm.

On the sections of the left ventricle’s endocardium to 0.1 whitish, indurate, with the network of proliferated connective tissue that spreads from it into the muscular wall of the ventricle (see Figure 1).

Figure 1. Thickening of the endocardium of the heart’s left ventricle (b). A thin and transparent normal endocardium of the right ventricle of the heart to compare (b).

At histological examination, cardiomyocytes of the conducting system (Purkinje cells) with karyorexis and karyolysis are located in the thickness of the endocardial connective tissue, either subendocardially or immured in
the fibrous stroma. From the thickened endocardium to the subendocardial parts of the myocardium, are growing narrow and wide strands, spurs; fields of fuchsinophilic fibrous tissue grow to a considerable depth, in many places forming anastomosing focal-reticular branching, islets, often surrounding or occurring bundles of cardiomyocytes, single cardiomyocytes or their small groups (see Figure 2).

In some fields of view, cardiomyocytes are fragmented, in others - bundles of thinned or uneven thickness of muscle fibers are synchronously wavy, deformed, with areas of angular bends and kinks.

Figure 2. Markin “a” - sharply thickened endocardium with severe sclerosis and calcifications (arrows), hematoxylin-eosin ×250. Marking “b” - proliferation of connective tissue in the thickness of the myocardium, atriochrome dye “picroMallori” ×250, connective tissue stroma dark blue (arrow). Marking “in” - karyolysis in the Purkinje cells (arrows), compressed by a connective tissue, atriochrome dye “picro Mallory” × 400.

Analysis of the clinical and morphological picture of the disease D. says that the death occurred due to acute cardiac rhythm disturbance - ventricular fibrillation caused by progressive damage of the conduction system of the heart in the left ventricle, because of the compression of the Purkinje cells by connective tissue growing into the myocardium with a complicated form of endocardial fibroelastosis of the left ventricle. Pregnancy of the woman has not played a significant role in the tanatogenesis of the disease.

Discussion

We have found only one case of death of 25-year-old woman with cardiac arrest and dilated cardiomyopathy dating 8 weeks after delivering her second child. To our knowledge, this is the first case in which findings consistent with LVHT. Left ventricular has been found in a patient with dilated cardiomyopathy in the peripartum period [10].

In the present case, the death of a woman occurred in the first half of pregnancy. This fact suggests that the death of women with such severe cardiac pathology, as an endocardial fibroelastosis can occur in any period of pregnancy and in the postpartum period as well. Undoubtedly, the additional load on the cardiovascular system during pregnancy in such patients is becoming an essential risk factor and develops irreversible destructive processes, leading to decompensation of cardiac activity due to myogenic dilation of the heart cavities.

The uniqueness of this case seems to us also in the fact that a young woman aged 32 has already had two previous successful pregnancies with the birth of healthy children. Moreover those pregnancies proceeded without complications for both -fetus and mother behavior. But, the third pregnancy ended tragically-with the death of the woman.

Summing up the discussion of this case, it should be noted that women with severe congenital and acquired heart diseases at the stage of planning pregnancy need to conduct a thorough cardiological examination to decide on possibility of child bearing and assess the degree of risk to the life and health of the woman herself.

References

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