

SUDDEN CARDIAC DEATH DUE TO HEREDITARY DISORDER

Siddikov Qutbiddin Marifjonovich

Andijan State Medical Institute, Uzbekistan

Abstract: This article presents a case from expert practice related to the sudden death of a teenager, which occurred within a few minutes after the appearance of the first symptoms. During his lifetime, this teenager had clinical manifestations of hereditary connective tissue disorders.

Keywords: hereditary disorder, cardiac arrhythmias, mitral valve prolapse, connective tissue, sudden death, cardiac death.

INTRODUCTION: According to modern concepts, hereditary connective tissue disorders are a heterogeneous group of diseases that are caused by genetic defects in the synthesis and/or breakdown of extracellular matrix proteins or disorders of connective tissue morphogenesis [1]. As an analysis of cases encountered in practice shows, recognition of hereditary connective tissue disorders is difficult, which is due to a complex of reasons.

MATERIALS AND METHODS: External examination revealed the corpse of a young man, body length 195 cm, asthenic build, satisfactory nutrition with deposition of subcutaneous fat of the female type. The muscular system is developed below average.

The head is of medium size, brachycranial in shape (wide, short), fused eyebrows, Mongoloid eye shape, ear

shells with a simplified curl pattern, earlobes are incremented.

The chest is narrow, flattened, the shoulders are narrow, the limbs and fingers are long and thin.

The skin is pale and cold to the touch. Cadaveric spots on the back surface of the neck, trunk and limbs, intense, diffuse, bluish-violet in color, practically do not fade when pressed. Muscle rigor is pronounced in the muscles of the face, neck, torso and limbs.

No bodily injuries were found during external or internal examination.

RESULTS AND DISCUSSION: Internal examination revealed the inner surface of the musculocutaneous flap and the temporal muscles without hemorrhages. The thickness of the bones of the cranial vault at the level of the sectional cut is 0.3-1.2 cm. The dura mater is tense, whitish-gray, and there is liquid blood in the sinuses. The soft meninges are swollen, smooth, translucent, with an expanded network of vessels, full of blood. The walls of the vessels at the base of the brain are thin and collapse. The hemispheres of the brain are symmetrical. The convolutions of the brain are flattened, the furrows are smoothed. On the lower surfaces of both cerebellar hemispheres there is a ring-shaped depression up to 1.5 cm wide and up to 1 cm deep. The brain is swollen, on a section with a visible border between the gray and white matter, the cerebellar hemispheres have a tree-like pattern, its tissue on sections is smoothed structure. The ventricles of the brain are not dilated and contain clear fluid.

On internal examination, the subcutaneous fat layer on the chest is 1.2 cm, on the abdomen – 3.0 cm.

The internal organs are located correctly; pathological changes are detected in the right common carotid artery, heart, thymus, spleen and lymph nodes.

At the site of the division of the right common carotid artery into the external and internal, a circular hemorrhage of a permeating nature is noted in its wall, spreading to the adjacent soft tissues with a maximum diameter of 3 cm. The inner lining of the aorta is yellowish, smooth. The width of the expanded aorta in the supravalvular section is 6.6 cm. The heart measures 13.5x12x7 cm, medium-medium with predominant hypertrophy of the left ventricle, weight 450 g. The outer lining

of the heart is smooth, transparent, shiny, with a small amount of fat. The cavities of the heart are not dilated; there is liquid dark red blood in the cavities of the heart and large vessels. The trabecular muscles of both ventricles have a complex pattern. The thickness of the muscle of the left ventricle of the heart is 1.7 cm, the right one is 0.5 cm, the thickness of the interventricular septum is 2.0 cm without obstruction of the outflow tract of the left ventricle. Ventricular index 0.5. The leaflet chords of the atrioventricular valves of the right and left ventricles are attached along the edge of the leaflets, are successively divided into rows, and are not shortened. The semilunar valves of the aortic and pulmonary artery valves are thin, smooth, transparent, and elastic. The mitral valve is multi-scalloped, redundant, with the presence of parachute-shaped bulges above the plane of the fibrous ring, the apex directed to the left atrium, the height of which varies from 3 to 5 mm. The left ventricle has a single diagonal median abnormally located chord with attachment points to the upper third of the interventricular septum and the head of the posterior papillary muscle. The inner lining of the heart is smooth and shiny. The heart muscle is dense, dark red and homogeneous on sections. The coronary arteries have ostia located centrally in the right and left sinuses of Valsalva, anatomically their course and division are correct, with a left type of blood supply to the myocardium.

The thymus gland weighs 80 g, flabby. Its left lobe is 7.5x4.0x1.5 cm, the right lobe is 7.0x4.0x1.0 cm, the isthmus is not defined. On sections, its tissue is light yellowish and homogeneous.

Spleen 21x15x8 cm, weighing 430 g, with a smooth capsule; When cut, its tissue is juicy, granular, dark red in color, and gives abundant scraping.

Lymph nodes of all studied groups are multiple, up to 1 cm in diameter, elastic, gray.

The spinal cord, respiratory, digestive, genitourinary and endocrine systems were also fully examined.

During the autopsy, in accordance with current instructions, blood, internal organs, brain and spinal cord were collected for forensic histological, forensic biochemical and forensic chemical studies.

A forensic histological examination revealed changes in the cardiovascular and nervous systems, thymus, and spleen.

In the heart there is moderate hypertrophy, atrophy of individual cardiomyocytes, stromal edema, mild lipomatosis and interstitial cardiosclerosis, in the coronary arteries there is subintimal hyperplasia and cystic medianecrosis. When stained according to Lee, there is positive staining over a large area. Intimal hyperplasia was noted in the right common carotid artery; in the middle layer - interstitial accumulation of extracellular basophilic matrix with the formation of slit-like cavities, degeneration of elastic fibers, around - bordering hemorrhage. When stained with MSB, there is no fibrin, plasmatic impregnation of connective tissue fibers. When stained for iron, the Perls reaction is negative.

In both hemispheres of the cerebellum there is congestion of the veins and venules, diapedetic hemorrhages, edema, and in some places a disturbance in the orientation of neurons.

CONCLUSION: Due to the fact that specific laboratory markers for a large group of hereditary connective tissue disorders have not currently been identified, and molecular genetic studies are inaccessible, priority is given to the formation of risk groups for the development of chronic pathology and the onset of sudden death has early detection of external phenotypic signs of hereditary connective tissue disorders and a set of clinical and functional disorders of organs and systems.

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