MORPHOLOGY AND PATHOMORPHOLOGY

Pathomorphological Criteria of Arrhythmogenic Heart

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Translated from *Byulleten' Eksperimental'noi Biologii i Meditsiny*, Vol. 136, No. 7, pp. 106-110, July, 2003
Original article submitted May 23, 2003

Pathomorphological criteria of arrhythmogenic heart include structural compartmentalization with primary changes in the right ventricle and interventricular septum, fibro- and lipomatosis of the myocardium, and disseminated coronary obstruction. Ischemic focuses in the conducting system are the site of formation of arrhythmogenic substance promoting the development and progression of cardiac arrhythmias. Cardioneuropathy and pathological motility of the interventricular septum lead to systolic dysfunction and contribute to asynchronous excitation and contraction of ventricles in arrhythmogenic heart.

Key Words: *cardiosclerosis; cardiac arrhythmias; postmortem coronarography and cardioventriculography; morphometry; histopathology*

A considerable progress in modern arrhythmology was achieved toward an understanding of functional mechanisms and development of methods for the correction of cardiac arrhythmias [2]. However, pathomorphological changes in the myocardium during arrhythmias received little attention [1]. Studies of etiology, pathogenesis, and structural and functional criteria for arrhythmic activity of the heart contributed to the introduction of an independent nosological characteristic "arrhythmogenic heart" and its individual forms into clinical practice in recent years [13]. The incidence of this disorder constantly increases. However, there are no reliable pathomorphological criteria for the diagnostics of arrhythmogenic heart.

Here we evaluated most reliable pathomorphological criteria of arrhythmogenic heart.

MATERIALS AND METHODS

We examined the hearts from 120 deceased patients with various forms of cardiosclerosis (average age 56.8±0.2 years). These patients had severe arrhythmias over 8-10 years before death. Ectopic arrhythmias that included extrasystoles and paroxysmal (extrasystolic) tachycardia were revealed in 62 patients. Fibrillation and flutter of the atria and ventricles were observed in 18 patients. Forty patients had conduction disturbances, including sinoatrial (n=18), atrioventricular (n=12), and intraventricular blocks (n=10). The control group included 60 hearts from individuals of comparable age not having cardiac arrhythmias (accidental death).

The coronary system of the heart was examined by a modified method of postmortem contrast polypositional coronarography [6] alone or in combination with standard anatomical techniques recommended by the World Health Organization [6]. Due to specific topographic and anatomical characteristics, studies of the coronary system were performed in parallel with visualization of the conducting system by micropreparation under a MBS-2 microscope (×36 magnification). We examined sinoatrial and atrioventricular nodes and right and left branches of the His bundle. Specially dissected hearts were microprepared under a microscope to reveal additional conducting pathways [8]. The state and volume of cavities in the heart were determined by postmortem contrast polypositional car-

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0007-4888/03/1361-093$25.00

dioventriculography [6]. The degree of ventricular dilation was estimated by objective criteria (outer equatorial and internal diameters).

Postmortem examination was performed within 6-8 h after death. Myocardial samples were taken for histological, histochemical, and target histotopographic assays. Paraffin sections were stained with hematoxylin and eosin, van Gieson's picrofuchsin, and Weigert's picrofuchsin-fuchseline to reveal elastic fibers. Staining for muscle and connective tissue was performed with hematoxylin, eosin, and light green. The volume density of the vascular bed, conducting myocytes, and connective tissue was determined by the planimetric method [1]. The ratio between adipose and fibrous tissues and contractile myocardium was estimated morphometrically in individual cardiac zones.

RESULTS

Cardioventriculography revealed a considerable increase in the volume of the left ventricle, hypertrophy of its walls, and displacement of the interventricular septum toward the right ventricle. The right ventricle gained a triangular shape. It had loosened and thinned walls and small cavity. The volume of this ventricle was $^1/_3$ of the left ventricle volume. The internal diameter of the right ventricle decreased (Fig. 1). Despite a considerable decrease in the volume, the equatorial diameter of the right ventricle did not differ from the normal due to pronounced motility of its walls and variations in geometric and topographic characteristics of the septum. These changes reflect pathognomonic correction of true dilation.

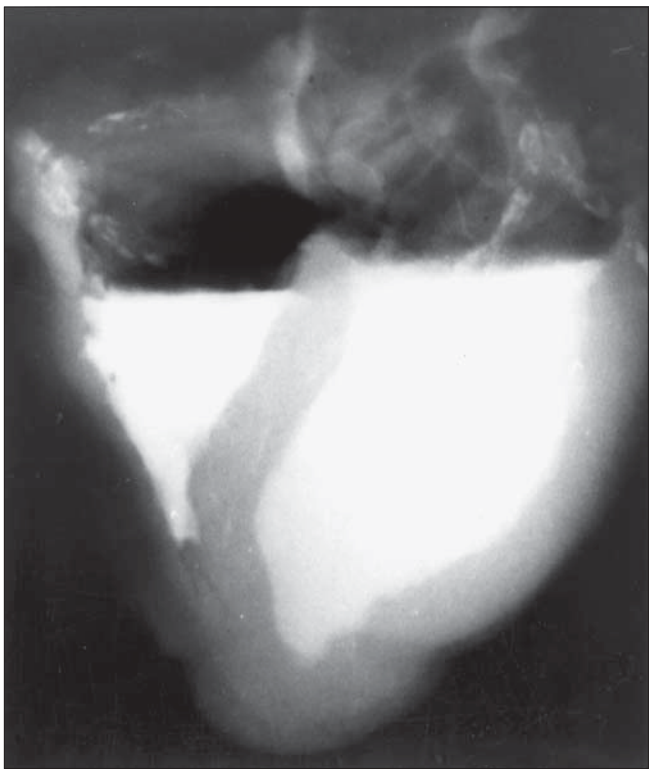

Fig. 1. Cardioventriculographic criteria of arrhythmogenic heart. Displacement of the interventricular septum toward the right ventricle gaining a triangular shape. Reduction in cavity, volume, and internal diameter of the right ventricle. Cardioventriculogram of patient A., 62 years.

Special dissection of the heart through the interventricular septum, point and triangle of Koch, and tendon of Todaro [8] visualized the atrioventricular node and revealed different scars in this zone. Transmural scars were revealed in 86 hearts (71.7%).

Histopathological examination found focuses of hemorrhages, calcification, and heterogeneous structure. Structurally heterogeneous focuses were especially pronounced in the zone of curvatures in the interventricular septum, which objectively reflects its pathological vital motility [7]. Pathological motility of the septum leads to systolic dysfunction and contributes to asynchronous excitation and contraction of ventricles. Interventricular asynchronism and conduction of electric pulses in branches of the His bundle determine variations in pathological movements of the interventricular septum [2]. The circuit of arrhythmogenic interrelations suggests that lengthening of electromechanical systole in the left ventricle and pathological movements of the septum are associated with the decrease in ventricular output [2,9].

The myocardium contained focuses of damage to cardiomyocytes and cardiosclerosis. We revealed diffuse and interbundle sclerosis of interstitial tissue and areas of fibrous scar tissue. Focal atrophy of cardiomyocytes and stromal infiltration with adipocytes were found in the sinoatrial anode, intraatrial tracts, atrioventricular node, His—Purkinje system, and additional conducting pathways (bundles of Kent, Mahaim, and James, Fig. 2, *a*). Fibromyocytic threads were present in areas of adipose tissue. The formation of adipose tissue represented the stage of degeneration and transformation of myocytes into adipocytes. The severity of changes increased from the endocardium to the epicardium. Myocardial threads were rarely found and substituted for adipose tissue and fibromyocytic threads (melange tissue) with a decrease in the distance to the epicardium [10,11].

Fibro- and lipomatosis were observed in 74 hearts (61.7%). These changes were most pronounced in the interventricular septum (Fig. 2, *b*) and walls of the right ventricle. The volume density of adipose and fibrous tissues and contractile myocardium varied in the anterior (62, 12, and 26%, respectively), posterior (48, 10, and 42%, respectively), and lateral wall of the right ventricle (44, 14, and 42%, respectively). Intersegment differences in the volume density of tissues

were revealed in the interventricular septum: proximal segment I, 30, 18, and 52%, respectively; median segment II, 58, 14, and 28%, respectively; and lateral segment III, 34, 20, and 46%, respectively. The observed volume relationships between adipose and fibrous tissues and contractile myocardium in the right ventricle and interventricular septum are typical of arrhythmogenic dysplasia of the right ventricle and arrhythmogenic heart, respectively [10,13-15]. The development of pathognomonic diagnostic changes was accompanied by progressive cardiac insufficiency.

Changes in heart architectonics were of particular interest. We revealed stenosis of main coronary arteries, collapse of distal regions in narrowed coronary branches, and reduction of coronary blood flow in myocardial scars. A considerable number of newly formed vessels with numerous collaterals were found. Most newly formed coronary vessels developed from collaterals of donor and recipient arteries. The main coronary arteries were characterized by a complex interbasin dissociation. Coronarography showed that obstruction of the left coronary artery was accompanied by dilation of the right coronary artery [6].

A special study of control samples visualized the sinoatrial nodal artery on coronarograms recorded polypositionally and in the right anterior lateral projection. This artery arose from the proximal regions of the right and circumflex coronary arteries. In 84 hearts of the main group (70%) changes in the sinoatrial nodal artery were manifested in curvedness, irregularity of the lumen, and aneurismal protrusions (Fig. 3). The artery was not visualized in 36 hearts (30%). The development of serious obstructive atherosclerotic changes in the coronary system suggests that the sinoatrial nodal artery was not absent, but various organic factors reducing its contrasting.

The data indicate that sinoatrial nodal arteries play an important role in arrhythmogenesis. Obstruction and obliteration of these arteries promote the development of various circulatory disturbances in nodal tracts and surrounding myocardium, which affects electrical stability of the heart. These changes contribute to insufficiency of the sinoatrial node and formation of stable ischemic focuses in the conducting system of the heart. Arrhythmogenic substances produced in these regions spread in the myocardium under conditions of residual blood circulation or reperfusion of ischemic tissue. Arrhythmogenic substances (primarily lysophosphoglycerides) determine arrhythmic activity of the heart during autonomic cardioneuropathy. This

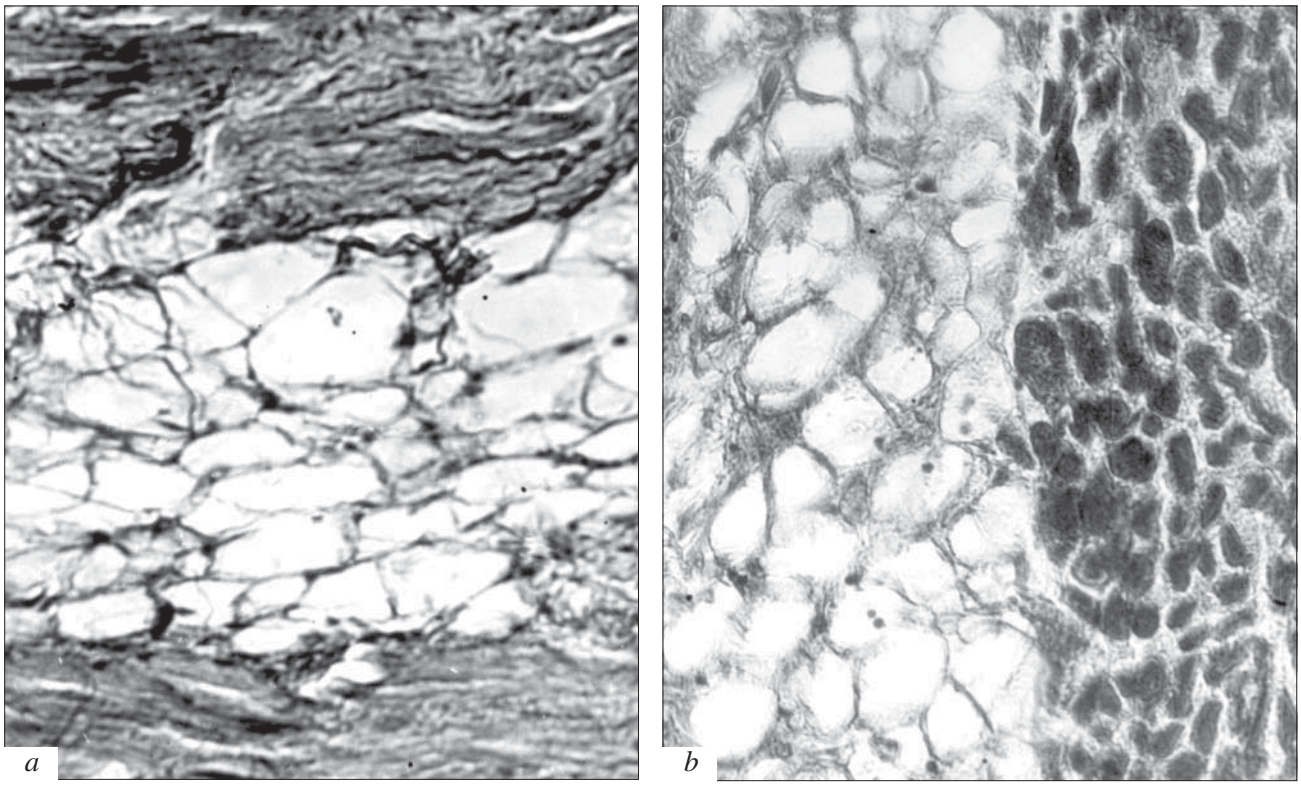

Fig. 2. Pathohistological changes in the myocardium of arrhythmogenic heart. Diffuse myocardial sclerosis, focal atrophy of cardiomyocytes, and pronounced adipocyte infiltration in the posterior wall of pathologically changed right ventricle in the zone of additional conducting pathways; staining with hematoxylin, eosin, and light green (patient G., 60 years, ×200, *a*). Pronounced lipomatosis in the proximal segment of the interventricular septum in the zone of the sinoatrial node; staining with hematoxylin, eosin, and light green (patient B., 64 years, ×160, *b*).

disorder is characterized by severe cardiac lipomatosis. Fibromyocytic threads serve as the zone for late potential generation [12,15] under conditions of myocardiocyte insufficiency [3-5].

Our findings show that pathomorphological criteria of arrhythmogenic heart are structural compartmentalization with primary changes in the right ventricle and interventricular septum and fibro- and lipomatosis of the myocardium. Diagnostic criteria also include disseminated coronary obstruction with interbasin dissemination, primary damage to the sinoatrial nodal artery, and formation of ischemic focuses in various zones of the conducting system producing arrhythmogenic substances. Arrhythmogenic substances contribute to the development and progression of cardiac arrhythmias under conditions of residual or reperfusion blood flow. Fibromyocytic threads serve as the zone for late potential generation during cardioneuropathy and lipomatosis. Pathological motility of the interventricular septum leads to systolic dysfunction and contribute to asynchronous excitation and contraction of ventricles in arrhythmogenic heart.

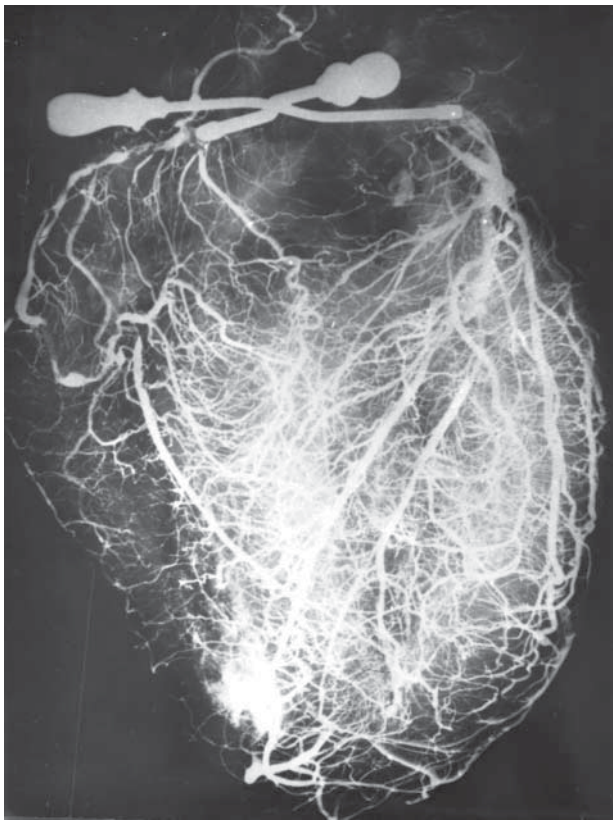

Fig. 3. Coronarographic criteria of arrhythmogenic heart. Sharply curved sinoatrial nodal artery with the irregular lumen, disseminated stenosis of main coronary arteries, and wide network of collaterals. Coronarogram of patient K., 58 years.

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