ORIGIN AND MODERN CLINICAL DIAGNOSIS OF INFILTRATIVE HEART DISEASES

[1]Ergashev Davlat

[2]Xolmatov Mehroj

[3]Boltayev Husan

[4]Mardonqulov Akmal

[5]Yavg'ashev Asomiddin

[1][2][3][4][5]Samarkand State Medical University, DKTF, Department of Internal Medicine, Cardiology and Functional Diagnostics, 2nd year clinical residents

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Introduction: To date, nine amyloidogenic proteins have been identified that can accumulate in the myocardium and cause cardiac amyloidosis. Three of them are common: serum amyloid A (AA amyloidosis, a consequence of chronic inflammation and infectious diseases), immunoglobulin light chains (AL amyloidosis, a consequence of monoclonal gammopathy), and transthyretin (two variants of transthyretin - ATTR amyloidosis: ATTRv amyloidosis, here ATTRv amyloidosis is associated with a non-hereditary type of transthyretin amyloidosis).

Research methods and materials: The prognosis of cardiac amyloidosis is very unfavorable. However, in recent years, certain progress has been made not only in diagnostics, but also in the development of drug therapy for cardiac amyloidosis, so that experts from the Working Group on Myocardial and Pericardial Diseases of the European Society of Cardiology have developed a consensus document on the diagnosis and treatment of cardiac amyloidosis [1].

A similar document was previously published by the American Heart Association [2]. Below we briefly review the main provisions of the European document.

First of all, the authors noted that the prevalence of cardiac amyloidosis may be higher than currently available data, which is usually associated with imperfect diagnosis of this disease.

In this regard, the following algorithm for diagnosing amyloidosis is proposed, consisting of two stages: 1. forming an assumption about the presence of cardiac amyloidosis 2. specific diagnosis of cardiac amyloidosis.

The following factors have been identified as factors that may indicate amyloidosis: left ventricular wall thickness of 12 mm or more, in combination with the following features: heart failure or aortic stenosis at age ≥65 years, hypertension, autonomic dysfunction, peripheral polyneuropathy, proteinuria, bilateral carpal tunnel syndrome, bilateral tendon syndrome, late gadolinium enhancement on cardiac magnetic resonance imaging (MRI), reduced left ventricular

longitudinal strain with preserved cardiac apex, low-voltage QRS complexes and pseudo Q waves on ECG, atrioventricular conduction disturbances, and family history.

In addition, both invasive and noninvasive diagnostic methods can be used to confirm the diagnosis. Invasive methods include biopsy of the myocardium or other organs, followed by staining of the biopsy material with Congo red and examination under polarized light, while noninvasive methods include echocardiography, cardiac MRI, phosphate scintigraphy, and blood tests for immunoglobulin light chains.

In addition, only ATTR amyloidosis can be diagnosed non-invasively. For this purpose, scintigraphy, blood tests for monoclonal gammopathies (to exclude AL amyloidosis) and echocardiography / MRI of the heart are performed. All other forms of amyloidosis are diagnosed only after confirming the presence of amyloid by biopsy (not necessarily myocardial) and during imaging studies according to certain criteria.

Results: No radiopharmaceutical uptake by the myocardium during scintigraphy, monoclonal protein studies are negative - the probability of ATTR and AL amyloidosis is very low.

Scintigraphy has revealed myocardial uptake of the radiopharmaceutical, monoclonal protein studies are negative - if the uptake level is 2/3 (the same as in bones or more), then the diagnosis of ATTR amyloidosis is considered confirmed. Next, genetic testing should be performed to determine its form. If the uptake level is 1 (less than in bones), histological confirmation of amyloid deposition is required.

There is no myocardial uptake of radiopharmaceuticals during scintigraphy, the study of monoclonal proteins is positive - AL amyloidosis should be excluded using cardiac MRI. If there are signs of amyloid on MRI, histological examination should be performed.

Scintigraphy revealed myocardial uptake of the radiopharmaceutical, monoclonal protein studies were positive - differential diagnosis between ATTR and AL amyloidosis should be made using histological examination.

Conclusion: Treatment of cardiac amyloidosis involves the use of specific drugs that should stop or slow the deposition of amyloid in the myocardium and therapy aimed at preventing cardiovascular complications. When discussing the latter, it should be noted that in general, only diuretics are used to treat heart failure in amyloidosis, and anticoagulant therapy is prescribed in atrial fibrillation, regardless of the number of points on the CHA 2 DS 2 VASc scale.

Pathogenetic therapy of amyloidosis includes chemotherapeutic drugs in AL amyloidosis, as well as several drugs that slow and stabilize transthyretin synthesis in ARRT amyloidosis:

patisiran, inotersen, tafamidis, etc. In the ATTR-ACT study, tafamidis was associated with lower cardiomyopathy in patients [3].

Thus, the available diagnostic methods allow for a reliable diagnosis of cardiac amyloidosis, as well as for determining its specific variant. However, the treatment of cardiac amyloidosis remains a difficult task, mainly due to the high cost of therapy, especially in relation to the most common type, transthyretin amyloidosis.

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