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Case report

Immunoglobulin light-chain amyloidosis – Diagnosed through electrocardiographic and echocardiographic features

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ABSTRACT

Introduction: Immunoglobulin light-chain (AL) amyloidosis is a systemic amyloidosis characterized by extracellular accumulation of fibrillary deposits composed of monoclonal immunoglobulin light chain fragments. Detection of the amyloid requires a special dye. The gold standard is Congo red stain. Clinical manifestation of the AL amyloidosis is variable and nonspecific. Amyloid deposits in cardiac issues lead to biventricular wall thickening. On the other hand, the amount of cardiomyocytes decreases, which manifests as a low QRS amplitude. The standard electro- and echocardiography may assist in the diagnosis of AL amyloidosis.

Aim: The aim of the study is to present the possibilities of using a standard electrocardiography and an echocardiography for diagnosis of a rare immunoglobulin AL amyloidosis.

Case study: This is the case of a 60-year-old woman who suffered from progressive fatigue, weight loss, diarrhea lasting for two years and recurrent syncope, hypotension, and dyspnea for six months. Routine diagnostic tests did not explain the cause of her symptoms. Electrocardiography revealed a low QRS voltage. An echocardiogram showed thickening of the left and right ventricular walls. The histological examination with Congo red staining revealed the amyloid deposits. The primary λ-light chain amyloidosis of heart, kidneys, autonomic nerves and soft tissue was diagnosed.

Results and discussion: We presented a typical case of immunoglobulin AL amyloidosis, which was detected thanks to characteristic electro- and echocardiographic findings.

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Conclusions: The coexistence of multiorgan dysfunction and the thickening of ventricular walls as shown by echocardiography combined with the lack of hypertrophy electrocardiographical features enables us to diagnose amyloidosis intravitally.

1. Introduction

Immunoglobulin light-chain (AL) amyloidosis is the most common type of systemic amyloidosis characterized by extracellular accumulation of fibrillary deposits, which is composed of monoclonal immunoglobulin light chain fragments synthesized by clonal plasma cells in the bone marrow. In most cases it is a primary disease, but may be associated with myeloma or other B-cell malignancy. It is considered to be a very rare disease even though its incidence is similar to the better-known Hodgkin disease.[1] It is estimated that the disease affects 5–12 people per 1 million per year. The diagnostic process starts from proving tissue amyloid accumulation. For that purpose, a tissue (usually subcutaneous abdominal fat tissue) biopsy is performed. Detection of the amyloid requires a special dye. The gold standard test is the Congo red stain. Amyloid deposits bind with the Congo red dye and exhibit the pathognomonic apple-green birefringence when viewed with a polarizing microscope. The next steps are as follows: identification of the amyloid precursor protein (monoclonal immunoglobulin light-chain), detection and differentiation of plasma cells dysplasia (primary or myeloma-associated) and defining organ involvement.

The AL amyloidosis affects many organs (most commonly kidney, heart, liver, gastrointestinal tract, nerves, and soft tissues), therefore the clinical manifestation is variable and nonspecific, which make the disease difficult to recognize. The most common symptoms are weight loss and fatigue, chronic diarrhea, progressive dyspnea, peripheral edema, hepatomegaly, ortostatic hypotension and proteinuria. In some cases (about 15% of patients), more specific symptoms, called 'red flags,' such as macroglossia, submandibular edema and 'raccoon eyes' (peri-orbital purpura) are present. When amyloidosis affects the heart, congestive heart failure, conduction abnormalities, arrhythmias, angina, syncope and sudden cardiac death may occur. Cardiac involvement is a leading cause of mortality due to electromechanical dissociation. The long-drawn elevation of cardiac troponins and the N terminal-pro brain natriuretic peptide (NT-pro BNP) is regarded as a typical change in cardiac amyloidosis, even at an early stage, as a result of the toxic effect of amyloids on myocytes and microvascular changes.

Amyloid deposits in cardiac tissue lead to biventricular wall thickening without ventricular enlargement visualized by the echocardiogram. Additionally, myocardial echogenicity increases ('granular sparkling'). The left ventricular ejection fraction is normal for a long time and diastolic dysfunction is dominant, although systolic dysfunction can be proved by tissue Doppler techniques. Wall thickening is often incorrectly described as hypertrophy, leading to a misdiagnosis of hypertrophic cardiomyopathy or hypertensive heart disease. The QRS voltage on 12-lead electrocardiography in amyloidosis is low unlike true ventricular hypertrophy. The combination of these electro- and echocardiographic findings strongly suggests a cardiac amyloidosis.

2. Aim

The aim of the study is a case report of a rare AL amyloidosis unrelated to multiple myeloma (primary amyloidosis), diagnosed on the basis of standard electrocardiography abnormalities and an echocardiography.

3. Case study

This is the case of a 60-year-old woman who suffered from progressive fatigue, weight loss (from 102 kg to 60 kg), diarrhea for two years and recurrent syncope, hypotension, dyspnea on exertion (NYHA III) for six months. She was hospitalized four months before admission to our department due to syncope as a complication of a right femur fracture. At the same time, myocardial infarction was diagnosed based on elevated levels of cardiac troponin T; however, angiography was not

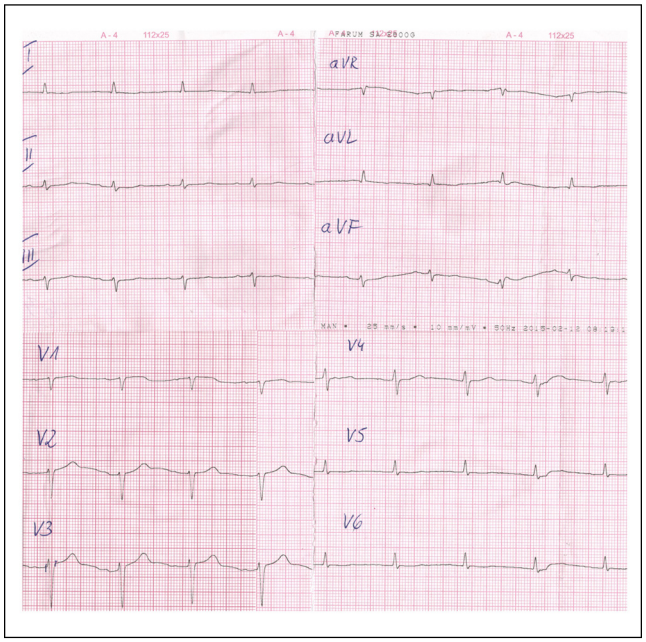

Fig. 1 – A 12-lead electrocardiogram of illustrating the low voltage limb QRS complexes, left anterior hemiblock and prolonged PR interval to 210 ms.