Cardiac arrest in a patient with a mobile right atrial thrombus in transit and amyloidosis

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Primary amyloidosis is a rare disorder in which insoluble fibers are deposited in tissue and organs, impairing their function. Cardiac involvement occurs in up to 50% of patients with primary amyloidosis. We describe a case of a 75-year-old admitted to our department after he had a sudden cardiac arrest due to massive bilateral thrombotic occlusion of the pulmonary arteries. The echocardiogram revealed many atrial thrombi swirling inside the right atrium and protruding into the tricuspid valve partially occluding it. Severe concentric hypertrophy of the left ventricle was also present with a preserved ejection fraction. The right ventricle was dilated, hypertrophic and ipokinetic with a severe tricuspidal insufficiency that permitted estimation of a severe pulmonary hypertension. All these characteristics were highly suggestive for an infiltrative form of hypertrophic cardiomyopathy. The final diagnosis was amyloidosis.

KEYWORDS
Amyloidosis; Thrombus; Pulmonary embolism

Introduction
Primary amyloidosis is a rare disorder in which insoluble fibers are deposited in tissue and organs, impairing their function.1,2 Cardiac involvement occurs in up to 50% of patients with primary amyloidosis. Diffuse amyloid deposits lead to an impairment of cardiac function with two different mechanisms: substitution of functional myocardium with amyloid protein that causes a systolic dysfunction and diffuse infiltration of the interstitium with stiff beta-plated proteins that reduce ventricle compliance, causing impaired relaxation and diastolic dysfunction. Diastolic dysfunction usually precedes systolic dysfunction determining high filling pressure with an abnormal enlargement of the atrium.

Atrial function is a key determinant of systolic function and prognosis. In the setting of a restrictive cardiomyopathy, left atrial contribution to left ventricular diastolic filling and stroke volume is critical. On the other hand atrial myopathy and systolic dysfunction may lead to thrombus formation even in the presence of sinus rhythm as previously reported in literature.3,4

Case report
A 75-year-old man who had been previously been healthy was admitted to our department after he had a sudden cardiac arrest successfully resuscitated by cardio-pulmonary manoeuvres.

The patient had no significant clinical history. At the time of presentation he was severely dyspnoeic. Initial clinical examination revealed blood pressure of 100/60 mmHg, markedly elevated jugular pressure, basal rales, split of S2.

The basal electrocardiogram showed sinus tachycardia, QRS low voltage and diffuse alteration of ventricular repolarization. Laboratory testing showed a highly positive D-dimer test. A bedside echocardiogram revealed many atrial thrombi swirling inside the right atrium and protruding into the tricuspid valve partially occluding it (Figures 1 and 2, Movie 1). Severe (IVS = 21 mm) concentric hypertrophy of the left ventricle was also present with a preserved ejection fraction (Movie 2). Left ventricle inflow showed a typical restrictive pattern at Doppler examination. Interventricular septum was also hypertrophic. The right ventricle was dilated, hypertrophic and ipokinetic with a severe tricuspidal insufficiency that permitted estimation of a severe pulmonary hypertension. All these characteristics, in the absence of valvular disease or a history of hypertension, were highly suggestive for an infiltrative form of hypertrophic cardiomyopathy.

The patient underwent a chest CT scan that revealed a massive bilateral thrombotic occlusion of the pulmonary arteries with atelectasia of the left inferior lobe. As an anticoagulation therapy heparin was added immediately.

As the clinical set stabilized an abdominal fat biopsy with Red Congo stain, showed amyloid deposition. Characteristic
‘apple green’ birefringence was clear under polarised light. No common biochemical abnormality leading to hypercoagulability was demonstrated in this patient.

Discussion

Cardiac arrest in patients with cardiac amyloidosis is mostly due to ventricular fibrillation or asystole.\textsuperscript{5,6} In this case a mechanical obstruction of the pulmonary arteries and tricuspid valve by massive floating thrombus is claimed to be the cause of an out-of-hospital cardiac arrest.

In cardiac amyloidosis, atrial myopathy infiltration consists in marked chamber enlargement, systolic dysfunction and atrial stand-still frequently in the more severe form of myocardial infiltration, even in patients with a stable sinus rhythm. Global atrial function is decreased significantly and much more than expected for the degree of ventricular involvement. These alterations joined to clotting factors might play a role in thrombogenesis even during sinus rhythm of primary systemic amyloidosis.

Cardiac amyloid predisposes patients to the development of atrial thrombosis and systemic embolization.\textsuperscript{3,4} There are limited data on mechanism of thrombosis in amyloidosis. Some authors proposed an impairment of the thrombin-antithrombin pathway.\textsuperscript{7} Therefore in literature the association between amyloidosis and bleeding abnormalities is well established.\textsuperscript{8,9}

We underscore in this case the utility and the rapid diagnosis by eco TTE that revealed right heart thrombus in transit and allowed/permitted a prompt therapy.

Supplementary material

Supplementary material associated with this article can be found in the online version.

References