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Myocardial amyloidosis combined with atrial giant thrombosis secondary to multiple myeloma: A case report

Jingbo Zhao, Zhu Ni, Yinhua Luo, Rui Huang, Ke Su, Yuanhong Li

ABSTRACT

Introduction: Multiple myeloma (MM) is a hematologic disorder that is relatively common in the elderly population. Given the rapid development, limited survival, and poor prognosis of the majority of patients, a definitive, prompt diagnosis is critical for treating individuals with multiple myeloma accompanied with cardiac amyloidosis. The incidence of myocardial amyloidosis with MM, relatively speaking, is low and early clinical manifestations are nonspecific. Atrial thrombi, with low detection, are a hallmark of poor prognosis of patients diagnosed MM with amyloidosis cardiomyopathy.

Case Report: Aiming to serve as useful feedback information for judging the prognosis of target patient, we now analyzed the clinical data of an elderly female with atrial thrombi as the main symptom and impaired cardiac function combined with echocardiography electrocardiography (ECG), laboratory data, cell Congo Red staining and other manifestations to diagnose amyloidosis combined with MM.

Conclusion: The formation of atrial thrombi is closely related to the prognosis of myocardial amyloidosis and in order to identify atrial thrombi early and to intervene early, transesophageal ultrasound should be included as part of routine investigations in patients with myocardial amyloidosis.

Keywords: Atrial thrombi, Echocardiography (ECG), Multiple myeloma (MM), Myocardial amyloidosis

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Jingbo Zhao[1], Zhu Ni[2], Yinhua Luo[1], Rui Huang[1], Ke Su[1], Yuanhong Li[1]

Affiliations: [1]Cardiovascular Disease Center, Central Hospital of Tujia and Miao Autonomous Prefecture, Hubei University of Medicine, Shiyan, Hubei Province 442000, China; [2]Department of Pathology, Central Hospital of Tujia and Miao Autonomous Prefecture, Enshi Clinical College of Wuhan University, Enshi Prefecture, Hubei Province 445000, China.

Corresponding Author: Yuanhong Li, Cardiovascular Disease Center, Central Hospital of Tujia and Miao Autonomous Prefecture, Enshi Prefecture, Hubei Province 445000, China; Email: lyh0101@vip.163.com

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INTRODUCTION

Amyloidosis cardiomyopathy is a rare disease with recurrent heart failure as the main clinical symptom, and is easy to be misdiagnosed and missed due to the insidious clinical manifestation at an early stage of the disease. Since it has no characteristic clinical manifestations and a low incidence rate, the diagnosis rate of myocardial amyloidosis following MM is low. In patients in sinus rhythm, the presence of atrial thrombi, especially if localized outside the appendages, is indicative of severe atrial stasis and also an independent marker of an unfavorable prognosis [1]. Thus, a search should be made for atrial thrombi, for it is helpful in judging the prognosis of patients diagnosed MM with amyloidosis cardiomyopathy. That's the reason that we reported a case of multiple myeloma with amyloid cardiomyopathy with primary presentation of left atrial appendage.

CASE REPORT

The patient, female, 54 years old, was hospitalized due to fatigue, bloating, and increased wheezing after activity for one month. No bone pain, no skin hemorrhage, and no fever during hospital stay.

Physical examination: blood pressure was 130/70 mmHg. Jugular venous distention, hepatojugular reflex (+), wet rales of bilateral lungs, and severe sunken edema in both lower limbs were found. Cardiovascular exam revealed that diastolic murmur of grade 5 or above can be heard in mitral valve auscultation area.

Laboratory results showed that elevated levels of cardiac enzymes, immunoglobulins (IgG, 7.05 g/L; IgA, 0.14 g/L; IgM, 0.08 g/L), immunoglobulin light chain (κ-light chains, 0.3 g/L; λ-light chains, 4.07 g/L), as well as elevated 24-hour urinary total protein (2218.6 mg/24 h). Electrocardiography (ECG) showed low voltage performance and junctional escape. During hospitalization, echocardiography displayed that the anteroposterior diameter of left atrium, left ventricle, right atrium, and right ventricle were 4.6, 3.9, 4.7, and 2.1 cm, respectively. Furthermore, echocardiography also revealed symmetrical thickening of the left ventricular wall and sparkling pattern of the myocardium (Figure 1). These clinical findings were highly suggestive of a diagnosis of myocardial amyloidosis combined with MM. Therefore, a bone marrow aspirate was performed for further evaluation. The bone marrow aspirate showed bone marrow changes typically seen in MM (Figure 2) and in combination with flow cytometry, plasma cells accounted for 12.5% of the total cell count. Immunofixation electrophoresis (IFE) revealed a positive result for λ-light chains (Figure 3). Cardiac magnetic resonance imaging (MRI) (Figure 4) shows thickening of the right and left ventricular walls during one cardiac cycle, with markedly increased myocardial signal and impaired filling of the myocardium, which is the typical characteristic of myocardial amyloidosis. Transthoracic echocardiography (TEE) showed a thrombus in the left atrial appendage, with a marked reduction of the left atrial appendage emptying velocity (Figure 5). According to these results and the diagnostic criteria, a clear positive diagnosis of myocardial amyloidosis combined with MM was finally made. In accordance with a statement issued by the Canadian Cardiovascular Society, we administered a combination chemotherapy regimen (DP chemotherapy regimen, doxorubicin + cisplatin) to this patient. The patient had recurrent malignant arrhythmias during treatment and was repeatedly readmitted for arrhythmias and heart failure at the 6-month follow-up.

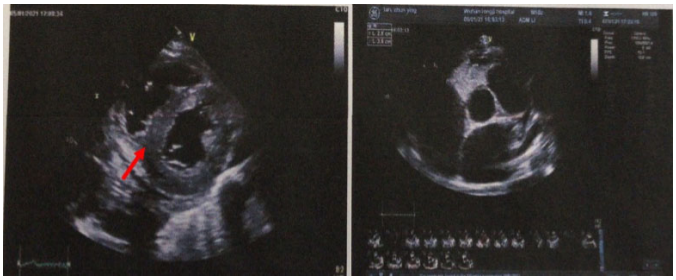

Figure 1: Echocardiography also revealed symmetrical thickening of the left ventricular wall and sparkling pattern of the myocardium.

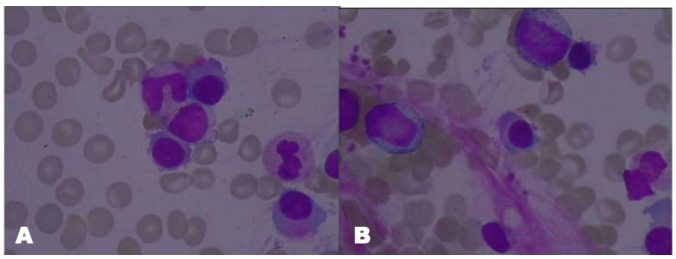

Figure 2: The bone marrow image showed an MM bone marrow change.

Figure 3: Immunofixation electrophoresis (IFE) revealed a positive result for λ-light chains.

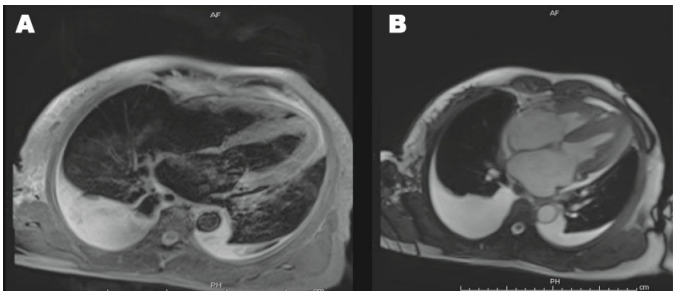

Figure 4: Diffuse tissue enhancement of the heart, involving the left ventricle, right ventricle, and left atrium, by gadolinium was shown on MRI.

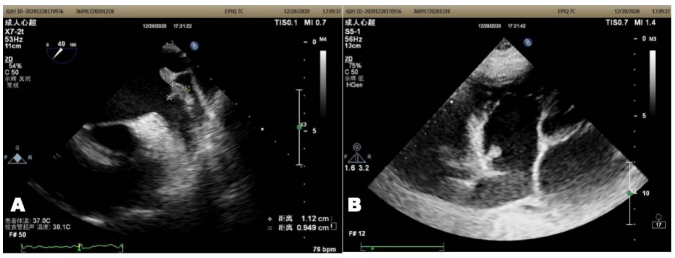

Figure 5: Thrombus in the left atrial appendage, with a marked reduction of the left atrial appendage emptying velocity.

DISCUSSION

Multiple myeloma accounts for 1% of all cancers and approximately 10% of all hematologic malignancies and is more prevalent in patients aged 65–77 years,

with a male prevalence [2–5]. Like multiple myeloma, amyloidosis also occurs in middle-aged and older adults (median age 64 years), with 99% of individuals aged 40 years or older. Amyloid deposits can occur in a variety of organs, including the heart, kidneys, liver, and peripheral nervous system, according to multiple literature reports. Kidney involvement is the most common among the above mentions followed by the heart. Cardiac amyloidosis often has no characteristic clinical symptoms initially, but eventually heart failure ensues [6]. And commonest presentation of amyloidosis is restrictive cardiomyopathy. The probability of developing amyloidosis in patients with multiple myeloma is about 10–15% [6] and the prognosis is extremely poor. Our patient was a 64-year-old female who admitted with cardiac insufficiency as the first manifestation, with no significant blood cell abnormalities or osteolytic symptoms, and who was diagnosed with multiple myeloma combined with myocardial amyloidosis based on the findings of bone aspiration and cardiac ultrasound and the revised diagnostic criteria for multiple myeloma.

The abnormal increase of monoclonal globulin caused by MM leads to the deposition of immunoglobulin in the heart and then contributes to myocardial amyloidosis which is classified as the immunoglobulin type of amyloidosis. According to the literature, myocardial amyloidosis often causes intracardiac thrombosis and often results in cardiogenic embolic stroke. It has been reported in the literature that in patients with sinus rhythm, the presence of atrial thrombus, especially if located outside the appendages, indicates severe atrial stasis, which is a characteristic of cardiac amyloidosis with restrictive diastolic dysfunction and a sign of advanced disease. However, the detection of atrial thrombi in patients with myocardial amyloidosis was mostly derived from autopsies, and there were bare of reports about giant atrial thrombi found in patients diagnosed MM with amyloidosis cardiomyopathy, as thrombi screening in target patients had not yet received sufficient attention. We reported firstly that a giant thrombus (approximately 1.1 × 0.9 cm in size) located in the left atrium of a patient diagnosed MM with amyloidosis cardiomyopathy aiming to serve as useful feedback information for improvements for thrombus screening. And it is important to routinely evaluate target patient with esophageal ultrasonography to avoid missing intracardiac thrombi or causing serious cerebral embolic events in patients.

Amyloid cardiomyopathy can predispose to left atrial thrombosis for multifactorial reasons and the mechanisms can be listed as follows: first, hemodynamic instability and intravascular turbulence formation in the cardiac chambers, leading to easy formation of thrombus at the location of the left atrium. Second, focal endocardial and subendocardial regions are continuously affected by amyloid deposition, leading to cell separation and deformation that can involve the myocardium, pericardium, small vessels, and conduction system, resulting in endocardial thickening, impaired ventricular wall motion. And the decreased ventricular wall compliance makes it easier for the formation of endocardial thrombi. Third, infiltration of amyloid into the endocardium may lead to myocardial ischemia, endocardial injury, appendicular thrombosis, and even cerebral embolism [6]. Finally, local amyloid deposition due to overload of circulating monoclonal components can cause vascular damage and lead to a hypercoagulable state of the blood, disrupting the blood clotting system, and thus predisposing to thrombosis. As in our patient, multiple myeloma leads to monoclonal protein overload in the blood system, which in turn deposits in the myocardium, pericardium, small vessels, and conduction system, causing restrictive diastolic dysfunction of the ventricular muscle and atrial blood pooling, leading to massive atrial thrombosis.

Although with the relatively high incidence of atrial thrombus in patients diagnosed MM with amyloidosis cardiomyopathy, the detection rate was still low and was mostly detected by autopsy. For conducting routine esophageal ultrasound evaluation for people diagnosed MM with amyloidosis cardiomyopathy was lost upon clinicians, and the main cardiac examinations were focused on two-dimensional ultrasound. However, the sensitivity of two-dimensional ultrasound is limited by the skills of ultrasonographers at all levels of hospital. The higher sensitivity of TEE in the detection of atrial thrombi recommends the introduction of this diagnostic tool routinely in patients affected by cardiac amyloidosis, at least in the advanced stages characterized by restrictive hemodynamics. We report a case of massive left atrial thrombosis in a patient with myocardial amyloidosis combined with multiple myeloma as an indication that transesophageal ultrasound should be included as part of routine investigations in patients with myocardial amyloidosis.

CONCLUSION

We reported a case of giant left atrial thrombosis in a patient with myocardial amyloidosis combined with multiple myeloma as evidence for the need for transesophageal ultrasound in patients with related disease.

REFERENCES

1. Santarone M, Corrado G, Tagliagambe LM, et al. Atrial thrombosis in cardiac amyloidosis: Diagnostic contribution of transesophageal echocardiography. J Am Soc Echocardiogr 1999;12(6):533–6.
2. Gerecke C, Fuhrmann S, Strifler S, Schmidt-Hieber M, Einsele H, Knop S. The diagnosis and treatment of multiple myeloma. Dtsch Arztebl Int 2016;113(27–28):470–6.

3. Falk RH, Alexander KM, Liao R, Dorbala S. AL (light-chain) cardiac amyloidosis: A review of diagnosis and therapy. J Am Coll Cardiol 2016;68(12):1323–41.
4. Saux A, Heroum C, Sportouch C, De Graeve F, Lequellec A, Pagès M. Amyloid cardiomyopathy: A rare cause of cerebral embolism. [Article in French]. Rev Neurol (Paris) 2006;162(11):1128–30.
5. Cho KH, Cho YM, Kim JS. Embolic infarction associated with cardiac amyloidosis. J Clin Neurol 2005;1(1):92–6.
6. Bøtker HE, Rasmussen OB. Recurrent cerebral embolism in cardiac amyloidosis. Int J Cardiol 1986;13(1):81–3.

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Author Contributions
Jingbo Zhao – Conception of the work, Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Zhu Ni – Acquisition of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Yinhua Luo – Acquisition of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Rui Huang – Acquisition of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ke Su – Acquisition of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Yuanhong Li – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.