Primary Amyloidosis presenting as Cardiogenic Shock and Pseudoinfarction. A case presentation.

J Perez-Zaldivar, R Tamayo-Isla, J Kitinya

Abstract

Primary Systemic Amyloidosis is a rare disease characterized by the deposition of fibrils, amyloid, in multiple organs, leading to multisystemic dysfunction and death. The heart is frequently involved causing infiltration of the cardiomyocytes and specialized cells of the conducting system provoking heart failure, arrhythmias and blocks of various types, and electrocardiographic changes that resemble a myocardial infarction. We present a case with a previous history of hypertension that presented in cardiogenic shock with an electrocardiogram supportive of anterior myocardial infarction; a postmortem examination revealed Primary Systemic Amyloidosis.

INTRODUCTION

Primary Systemic Amyloidosis (PSA), also known as AL Amyloidosis, is a multisystem disease caused by a plasma cell dyscrasia, characterized by the extracellular deposition of fibrils composed of immunoglobulin light chains within various organs: heart, vessels, kidney, tongue, liver, peripheral nerves, and others. The extracellular deposition of fibrils causes dysfunction of affected organs that leads to death. Although a well known cause of restrictive cardiomyopathy, cardiac failure, syncope, pseudoinfarct pattern and conduction anomalies in adults, its diagnosis is frequently missed because the condition is rare, requires high index of suspicion and histological confirmation. We discuss a case that presented in cardiogenic shock after and episode of syncope with an electrocardiogram supportive of an old anterior myocardial infarction plus renal impairment and non-nephrotic range proteinuria; a postmortem examination revealed Primary Systemic Amyloidosis.

CASE PRESENTATION

A 60-yr-old-african-male was brought to casualty by his brother who gave the history that the patient lost consciousness for a few minutes while walking in his room; he did not show any involuntary movement but had urinary incontinence. On arrival the patient had recovered consciousness and gave the history that he had become very weak for the past 3 weeks, almost unable to stand; also complaining of shortness of breath, even at rest, dry cough and sweating. He denied chest pain, palpitations or fever.

He had been on treatment for hypertension for three years. He had few episodes of chest pain in the past; the last one just two months ago, he was assisted by his family physician as an out-patient. He was not diabetic and never had tuberculosis. He stayed in a rural village with his wife and two grandchildren. He used to be a carpenter. Never smoke or drink.

On physical examination he was found critically ill, bedridden, sweating.

Vitals: blood pressure: 88/44 mmHg, heart rate: 56/min, regular pulse, respiratory rate: 28/min, temperature: 36.0 ° Celsius. He had bibasal fine crackles. The apical beat was found at six intercostal spaces, one cm from the midclavicular line. A 2/6 midsystolic apical murmur and a fourth heart sound were heard. He also had bilateral ankle edema. Neck veins were not distended. The liver could be felt three cm bellow the costal margin. He spoke with difficulty in a low voice, but was well oriented and had no motor or sensory deficit. The tongue was normal.

His EKG (Fig 1) showed a sinus bradychardia, A-V block of first degree, delayed intraventricular conduction, QS in D1, D2, aVL, V1-V3, V4 and V6. A chest X ray showed early signs of pulmonary edema and cardiomegaly, no consolidation or pleural effusion. Other investigations results can be seen in Table 1.
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Table 1. Blood results

<table>
<thead>
<tr>
<th>Test</th>
<th>Day 1</th>
<th>Day 2</th>
<th>Day 3</th>
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<td>13.3</td>
<td>10.1</td>
</tr>
<tr>
<td>Hb</td>
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<td>10.2</td>
<td>9.2</td>
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<tr>
<td>Plat</td>
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<td>411</td>
<td>254</td>
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<tr>
<td>ESR</td>
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<td>18</td>
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<tr>
<td>CRP</td>
<td>49.5</td>
<td>78.3</td>
<td>106.0</td>
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<td>48</td>
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<td>656/13/2.0</td>
<td>335/10/3.1</td>
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Glomerular filtration rate estimated as 13.6 ml/min. An echocardiogram revealed concentric hypertrophy, diastolic dysfunction with restrictive pattern, mild mitral regurgitation. No wall motion abnormalities. And abdominal ultrasonography showed a congestive liver, kidney were normal in size with some how diminished corticomedullary differentiation but not loss of it. Rest of the organs was normal. Urine dipstick: 3+ blood, 2+ protein.

Figure 1

Table 1. Blood results

Figure 2

Fig 1. Electrocardiogram showing sinus bradycardia, undetermined QRS axis, prolonged PR, nonspecific intraventricular conduction disturbance, QS in D1, D2, aVL, V1, V2, V3, V5, V6.

Figure 3

Fig 2. Section of coronary artery stained with Congo red and visualized under polarized light, showing amyloid deposition as yellow fluorescence. X 400.

A diagnosis of Cardiogenic Shock was made, secondary to Ischemic and/or Hypertensive Heart Disease and an “old” Anterior Myocardial Infarction; Chronic Renal Failure due to Hypertensive Nephrosclerosis with an acute component due to shock.

He was admitted to ICU were despite IV fluids and inotropic support blood pressure never improved, renal function got worse and needed hemodialysis. He developed ventilator-associated nosocomial pneumonia and died four days after admission.

A post mortem examination revealed: amyloid infiltration of heart (Fig 2), kidney (Fig 3), liver, intestine, adrenal glands and lungs. No features of Multiple Myeloma were found. Immunohistochemistry revealed AL type of amyloid, kappa light chains; so the diagnosis of Primary Systemic Amyloidosis was made.
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Figure 4
Fig. 3. Section of kidney showing extensive hyalinisation of glomeruli and artery walls due to deposition of a homogeneous material that was shown to be amyloid by Congo red staining. H & E x400.

DISCUSSION
Primary Systemic Amyloidosis, or AL Amyloidosis, is characterized by the deposition of fibrils, which consist of immunoglobulin light chains polymers, in multiple organs, mainly heart, blood vessels, liver, tongue, kidney, skin, joints, and peripheral nerves. Cardiac involvement is recognized in about 50% of cases, usually as congestive cardiac failure, syncope, conduction anomalies or combination of them. Our patient presented after an episode of syncope, in cardiac failure of maximal severity, cardiogenic shock, probably as the last phase of a cardiac failure that was presented for months. This complication has been associated with very poor prognosis, with a survival rate of only four months after diagnosis.

What makes our case more interesting is the electrocardiogram which leads us to think in a previous myocardial infarction, even more since the patient reported a few episodes of chest pain in the last few months prior to admission that could explain undiagnosed episodes of myocardial ischemia.

Low voltage and pseudoinfarct pattern are the most frequently electrocardiographic signs found in cardiac amyloidosis, they have been reported in 50-65% of cases; also prolonged PR, intraventricular conduction delays and bundle branch block of all types and combinations have been described. Our patient electrocardiogram shows QS in D1, D2, aVL, V1, V2 V3 and V5, V6, with a minuscule r in V4, the ST segment was not displaced (Fig 1), all these findings are supportive of an anterior myocardial necrosis, not in acute phase. Low voltage was not present. The QRS duration was more than 120 msec, also the PR was prolonged (> 200 msec); all these findings of abnormal morphology and delayed conduction are just an indication of a wide infiltration of the conducting system by the amyloid substance. A restrictive pattern on echocardiography found in an adult with a failing heart should alert the physician about the possibility of cardiac amyloidosis; although it was found in our patient, the previous history of hypertension and the concentric hypertrophy mislead us to think in hypertensive heart disease, a condition of very high prevalence in South Africa. The combination of a thick septum and low voltage, that our patient did not show, has been reported by Rahman et al, and also by Carroll et al, as very characteristic of cardiac amyloidosis; the former reports a 72% sensitivity and 91% specificity for this condition.

This case shows that PSA, although rare, should be considered in the differential diagnosis of an adult patient with cardiac failure of uncertain cause which shows a restricted pattern with hypertrophy on echocardiography, without focal motion abnormality, and an electrocardiogram compatible with myocardial infarction, even if a low voltage is not present.

References
8. Rahman JE, Helou EF, Gelzer-Bell R, Thompson RE,
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