WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 8.084
Volume 10, Issue 11, 95-101. Case Study ISSN 2277– 7105

COMPLETE ATRIOVENTRICULAR BLOCK IN CARDIAC AMYLOIDOSIS: UNUSUAL PATTERN OF REVELATION

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Article Received on
26 June 2021,

Revised on 15 July 2021,
Accepted on 05 August 2021

DOI: 10.20959/wjpr202111-21279

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ABSTRACT

Cardiac involvement is frequent in systemic amyloidosis and is a major determinant of treatment options and prognosis. It may occur in the three main types of amyloidosis, and include diastolic followed by systolic heart failure, atrial and/or ventricular arrhythmias, embolic events and sometimes conduction disturbances with sudden death. Complete atrioventricular block is an unusual pattern of revelation of this disease as described on this case report. This disturbance can be due to infiltrative destruction of the conduction system by amyloid; but also; it can result from diffuse or focal occlusion of the vessels supplying the specialized conduction system. The purpose of this work is to alarm the clinician about the significant risk of conduction disturbances in a patient with amyloidosis; as well as to highlight the interest of seeking, preventing and treating these disorders, and this by electrocardiogram, holters, or even endocavitary explorations.

KEYWORDS: Complete atrioventricular block; Cardiac amyloidosis; echocardiogram, pattern.

INTRODUCTION

Amyloidosis is due to extracellular tissue deposition of insoluble fibrils composed of a variety of serum proteins (amyloid), resulting in tissue involvement. Deposition of amyloid can be localized or systemic (in virtually all organs except the brain). Clinical manifestations are based on the site of the amyloid deposits and are related to the type of precursor protein involved.[1]

Cardiac amyloidosis (CA) describes clinically significant involvement of the heart by amyloid deposition, which may or may not be associated with involvement of other organs.[2]

The mode of revelation of this pathology can be atypical as it is described in this case and cardiologists should be aware of that, to allow early treatment that may change the prognosis of this disease.

CASE PRESENTATION

A 72 year old man presented to emergencies with left sided acute chest pain; which emerged after conducting heavy work at home. His medical history included hypertension with poor therapeutic compliance; and old smoking weaned. There was no past history of a prolonged fever or tick bite, and his family history didn't reveal sudden death or coronary artery disease. He has no medication based on digitalis, amiodarone or beta-blocker, and never had syncope or palpitations.

At physical examination, both systolic and diastolic blood pressures were elevated (Blood Pressure readings of 210mmHg/110mmHg). There was no significant pressure gradient between both upper limbs or upper and lower extremities. Cardiac auscultation revealed regular bradycardia at 45 beats per minute with a maximal ejectional systolic murmur at the left base of the sternum, with preserved B2. There was no sign of heart failure and pulmonary auscultation was normal. Electrocardiogram (ECG) shows a 3rd degree atrioventricular block with a ventricular escape at 41 beats per min **(Fig.1**). Troponin was moderately high and biological examinations didn't show any metabolic, ionic or renal perturbation.

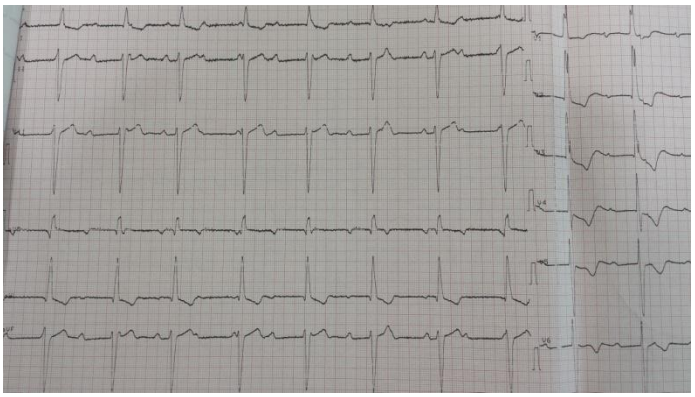
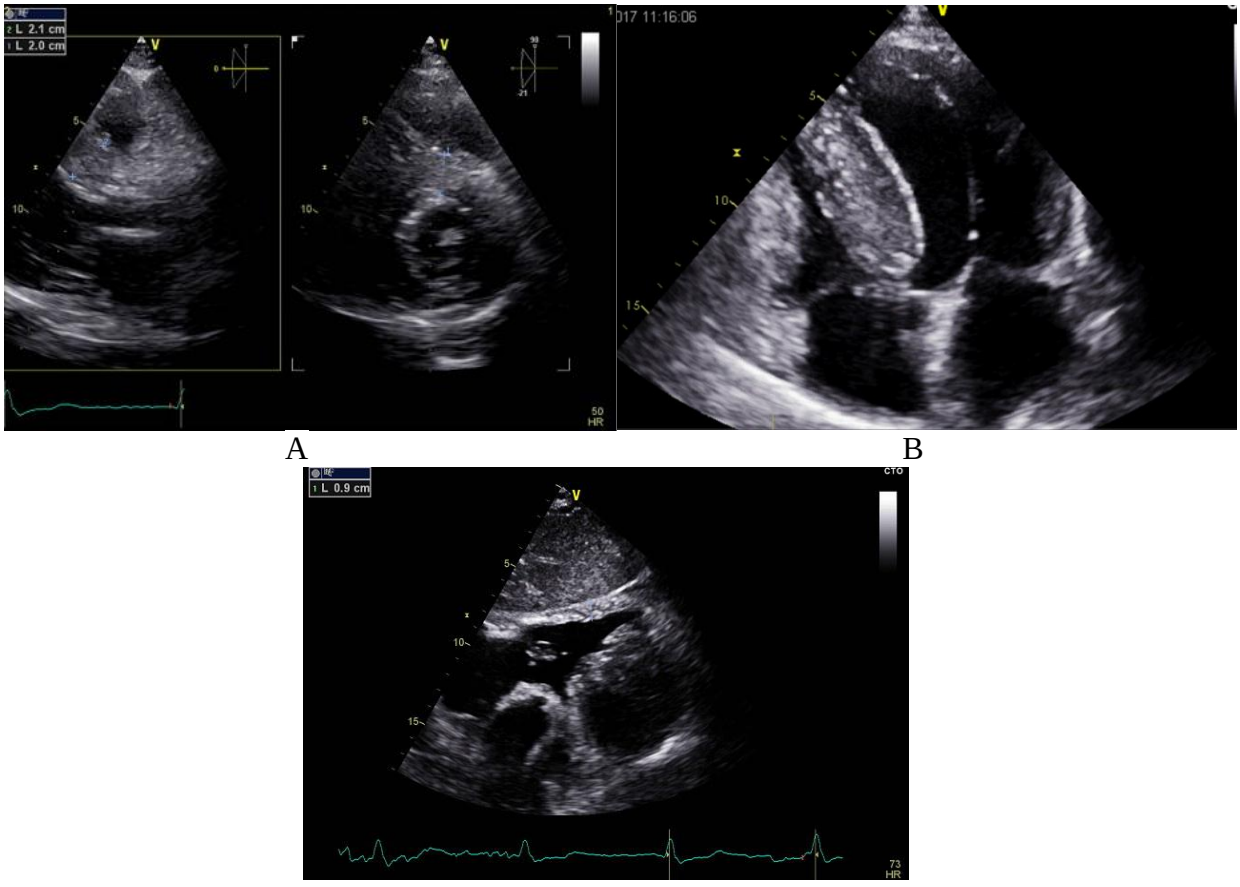


Fig. 1: ECG showing complete atrioventricular block with more P waves than QRS complexes, and no relationship between them. Narrow QRS complex escape rhythm is due to a blockage above the His bundle.

Trans Thoracic Echocardiography (TTE) revealed severe left ventricular hypertrophy with mild myocardial speckling, sparkled interventricular septum measuring 21mm, and thickened right ventricular free wall (9 mm) (**Fig.2**). Global LV systolic function appeared preserved;

ejection fraction (LVEF) by Simpson's biplane method measured 60%: no regional wall motion abnormality was identified. However, longitudinal myocardial systolic strain by 2D speckle tracking showed far more significant LV dysfunction. Global longitudinal strain (GLS) measured −12% (normal GLS = −18%), and regional strain values were greatly reduced in the basal and mid LV, yet preserved at the LV apex (see **Fig.3**). Longitudinal strain bull's eye plot patterns suggested CA **(Fig. 3**). Magnetic Resonance Imaging (MRI) confirmed the diagnosis. Coronarography shows non-significant lesions of the anterior interventricular artery. A pacemaker has been implanted in this patient .He remained asymptomatic after Pacing and was discharged three days later. A subsequent follow-up with characterization of his amyloid involvement by other explorations is planned for this patient.



A B

C

Fig. 2: A: Parasternal long and short axis views with with measurements of left ventricle dimensions, B: Apical 4 chamber view showing sparkled interventricular septum, C: subcostal 4 chamber view displaying right ventricular hypertrophy.

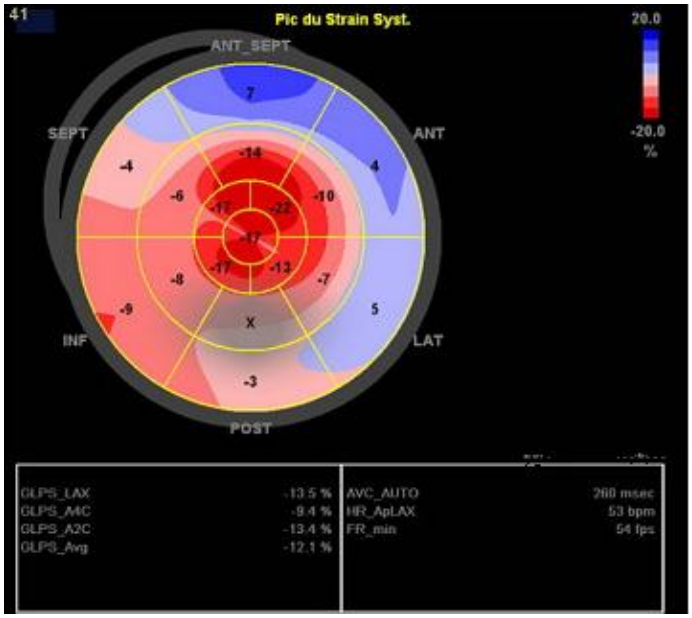


Fig.3: 'Bull's eye' display of regional LV longitudinal strain (with basal regions outermost, mid regions in the middle and apical regions innermost). Note the significantly reduced basal and mid LV longitudinal strain, with relative apical sparing in all ventricular walls.

DISCUSSION

The largest study that reported ECG findings in CA, consisted of 127 patients with AL and biopsy-proven cardiac involvement, and had confirmed that the two most common abnormalities were low voltage QRS complex and a pseudo infarct pattern on the precordial leads which were seen in roughly 50% of the patients included. Right and left bundle branch block are uncommon.[3] Other changes that may occur include conduction abnormalities, more frequently atrial fibrillation in about 15% of patients[4] and, rarely, ventricular tachycardia in about 5% of patients. Third degree AVB was found in only 3 patients (2%). Another study[5] that included a large number of patients with AL with and without cardiac involvement, reported that a fragmented QRS (notches and RsR' pattern in the absence of QRS prolongation) was significantly more frequent in patients with CA (28.5% vs. 11.7%; *P* =0.0008). Twenty-four-hour Holter ECG monitoring might help to identify asymptomatic ventricular/ supraventricular arrhythmias in > 75% of cardiac AL patients. Complex ventricular arrhythmias have been reported to be prognostic determinants for survival, but only couplets correlated with sudden cardiac death and were independent predictors of survival.[6] Regarding endocavitary conduction disturbance, a large study[7] that included patients with confirmed AL found that they all had normal sinus function. However, the infra-Hisian conduction time was significantly prolonged (79 ms) and was independently

associated with the occurrence of sudden death; the authors concluded that HV prolongation may be a marker of significant infiltration of myocardium by amyloid fibrils, which may be responsible not only for ventricular tachycardia, but also for severe atrioventricular conduction abnormalities with high degree AVB or asystole.[1]

Indeed, CA is associated with major electrocardiographic conduction disturbances (CD); but that these disturbances are due to infiltrative destruction of the conduction system by amyloid is unclear. In a study that reported conduction systems in 23 autopsy patients with CA[8], 21(91 per cent) of whom had abnormalities of conduction or rhythm during life, only three had extensive amyloidosis of the conduction system. In all three, ECGs showed first degree AVB and left anterior hemiblock. A more common morphologic abnormality of the conduction system was severe sinoatrial node fibrosis present in seven (30 per cent) patients (more frequent in patients with severe or moderate amyloid), and idiopathic atrophy and fibrosis of the bundle branches present in six (26 per cent) patients. However varying degrees of atrioventricular and bundle branch block were also present in six patients with no morphologic abnormalities of the conduction system. Thus, conduction and rhythm disturbances are frequent in CA, but direct amyloid infiltration of the specialized conduction tissue of the heart does not account for the majority of these disturbances.[8]

On the other hand, this case history illustrates that CA may simulate coronary heart disease. The patient had chest pain considered to be angina pectoris with troponin elevation, but the coronary arteries showed no significant stenosis. This has also been described previously in a minority of patients with CA.[9] The angina symptoms were not considered atypical in either ROBERT R. study[10] or a previous study. It appears that these clinical symptoms are the cumulative effect of progressive narrowing of the smaller coronary arteries with focal occlusions and localized myocardial “infarcts”**.** This focal necrosis and scarring eventually result in significant replacement of the myocardium by fibrous scarring. However; rarely amyloid deposition in these smaller intramyocardial coronary arteries may cause myocardial necrosis, ischemic heart disease and CD.[10] In fact; diffuse or focal occlusion of the vessels supplying the specialized conduction system can usually manifested by myocardial failure, functional impairment of vascular anastomoses or conduction and rhythm disturbances. It had been shown therefore that CA cause functional coronary insufficiency, presumably by involvement of small vessels with functional impairment of vascular anastomoses.[10] The elevation of troponin is also described in CA, and both NT-proBNP and cardiac troponin

have been used widely since2004 for assessing cardiac involvement severity and prognosis especially in Light-chain amyloidosis.[1]

CONCLUSION

CA may cause fatal disturbances in the heart rhythm and Conduction. A routine Holter monitoring in patient with CA is recommended. If it turns out normal; but despite this patient has syncope, repeated registrations have to be performed in hospital preferably by telemetry. Electrophysiological studies to look for sinoatrial and atrioventricular CD, and programmed stimulation to disclose tachyarrhythmias would probably be helpful in such cases.[9]

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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