Amyloïdose cardiaque, à propos d'un cas atypique

Cardiac amyloidosis: a challenging diagnosis

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Résumé

Cas d'une patiente âgée de 59 ans présentant des lipothymies dans le cadre d'une amylose cardiaque AL . Ce cas Clinique permet de parcourir la littérature sur l'atteinte cardiaque du tissu conductif et la prise en charge spécifique de l'amylose cardiaque AL.

Mots-clés

Dysfonction sinusale , insuffisance cardiaque , echocardiographie, scintigraphie osseuse , dysfonction sinusale

Summary

A 59 year old patient with presyncope due to AL cardiac amyloidosis is described. This case offer the opportunity to review the literature with the cardiac involvement associated with conduction disease to review the literature on cardiac damage to conductive tissue and specific management.

Keywords

Sinus node dysfunction , heart failure , echocardiography, nuclear

scintigraphy

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CASE PRESENTATION

A 59-year-old woman with a history of Carpal tunnel release surgery and hypertension was presented to the emergency department with recurrent episodes of presyncope. Physical examination revealed blood pressure of 100/70 mmHg with a pulse rate of 55 beats per minute, jugular venous distension, cardiomegaly, loud B2, hepatomegaly, and Bilateral pitting edema.

A 12-lead electrocardiogram revealed low voltage in limb leads with isorhythmic AV and a pseudo infarct pattern with Q waves in the early precordial leads without signs of recent ischemic changes as compared with earlier records (Figure 1). Blood investigations revealed anemia, a mildly deranged liver, and normal renal function. The diagnosis of sinus node dysfunction was confirmed, and we opted for a Dual Chamber Pacemaker.

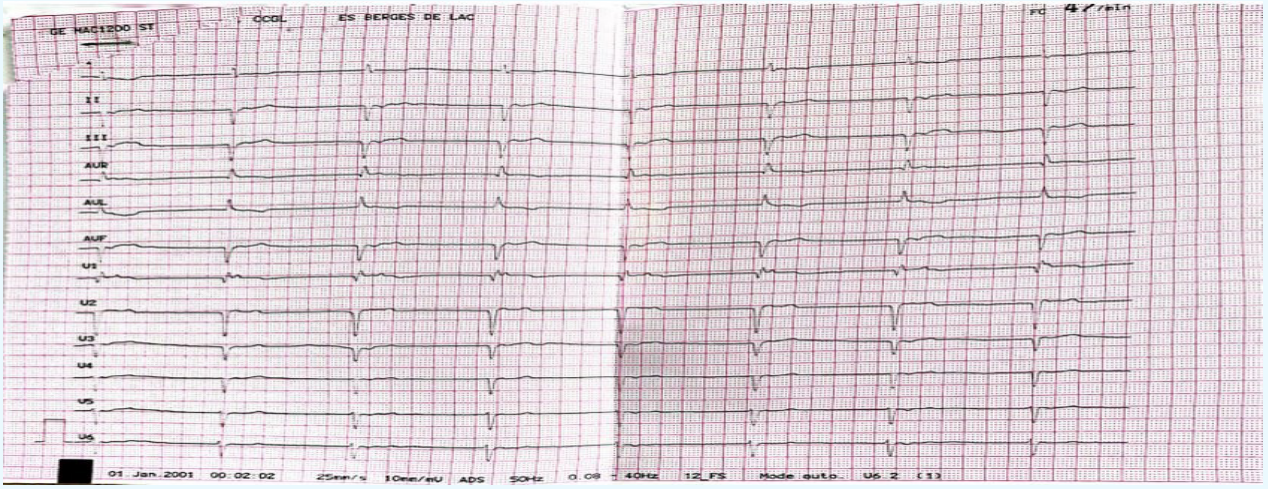

Figure 1. 12 leads electrocardiogram (EKG) showing low QRS voltage , isorhythmic AV Dissociation , left anterior fascicular block, QS-pattern in anterior leads

The echocardiography revealed marked thickening of the left ventricular (left ventricular posterior wall diastolic thickness 1.4 cm and right posterior wall, pericardial effusion a biatrial dilatation (Figure 2) , and restrictive filling pressure, LVEF was about 70 %, motion abnormalities in LV basal and middle segments, Global longitudinal strain (GLS) measured -12 .% (normal GLS = -18.6%), and regional strain values were significantly reduced in the basal and mid-LV, with relative apical sparing (Figure 3).Thus the diagnosis of Cardiac amyloidosis was suspected. Nuclear scintigraphy with the use of bone-seeking, phosphate-based radiotracer 99mTc-PYP showed Perugini Grade 1 myocardial uptake.

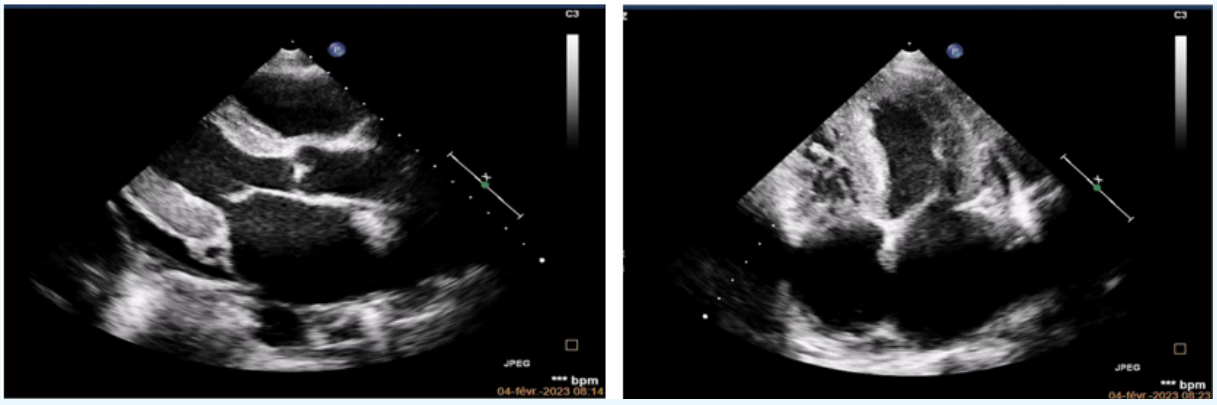

Figure 1. Transthoracic echocardiogram showing in apical four-chamber view fine granular appearance of the myocardium and biatrial enlargement. Left ventricular thickening in parasternal long axis view

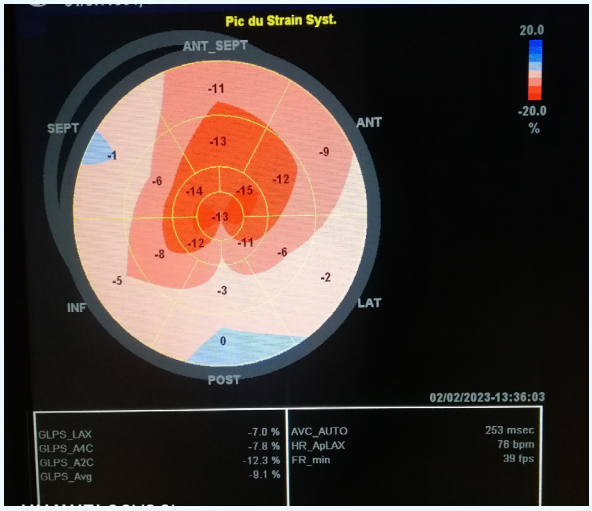

Figure 3. Echocardiographic strain analysis showing a relative sparing of the apex compared to the bases

We performed serum protein electrophoresis (SPEP) to investigate further, and we found a monoclonal spike in the gamma region with an estimated 15,6 g/l (normal reference < 12 g/l) kappa/lambda light-chain ratio was 0.07 (standard reference > 0,26 and < 1,65 g/l). Serum immunofixation showed a monoclonal spike in light chain lambda. Minor salivary gland (MSG) biopsy was negative, so we have done a kidney biopsy with immunofluorescence showing lambda light chains deposition in the glomerular mesangium. Thus the diagnosis of AL amyloidosis was established. During hospitalization, the patient presented a heart failure episode with Elevated Cardiac troponin at 692.35 ng/ml standard reference < 14 ng/ml) and NT pro-BNP 4839 pg/ml (standard reference < 125 pg/ml). She was treated with loop diuretics therapy and was addressed after decongestion to hematology department for chemotherapy.

DISCUSSION

This case highlighted many findings; our patient's atypical initial presentation, with syncope and isorhythmic AV Dissociation that is uncommon in cardiac Amyloidosis since Cardiac Amyloidosis is classically presented with clinical features of restrictive cardiomyopathy right ventricular failure, arrhythmias, and possible angina despite normal coronary angiograms and this is due to amyloid deposition in arterial vessel wall causing progressive luminal narrowing (3).

Conduction disorder is a potential complication of Amyloidosis. Despite its prevalence due to the predisposition for amyloid to deposit in the atria, Atrioventricular (AV) conduction delay or block appears to be more frequent than sinus node disease (4,5). SND in CA needs to be better studied. Most studies investigating conduction system disease in CA do not discuss SND. Available data suggest it is uncommon (4).

The second finding illustrated in our case was the isolated Sinus node dysfunction without another trouble, in contrast with literature where Sinus node dysfunction is associated with arrhythmias such as Atrial Fibrillation, Ventricular arrhythmias (5–7).

Thirdly in our case, the sinus dysfunction was associated with AL amyloidosis, which is less frequently observed in this form of amyloidosis.

In fact, sinus node dysfunction appears less common and relevant reports are mostly limited to isolated events in patients with ATTRm. In some reports, transient sinus node dysfunction was associated with autonomic dysfunction occurring spontaneously or during general anesthesia induction (8).

Similarly, In a recent single-institution retrospective study of 369 patients followed over 28 months, SND occurred in 7% of patients (9).

Although infiltrative conditions, including CA, have been considered classical causes of sinoatrial node pathology, published outcomes in CA are lacking

An intracardiac electrophysiology (EP) evaluation of sinus node function in 25 patients with AL amyloidosis revealed normal sinus node function in 88% of those examined (10).

CONCLUSION

AL amyloidosis is a rare disease that can be present with sinus node dysfunction screening for cardiac involvement should be considered in all patients with amyloidosis. We presented a case of a 59-year-female who presented with sinus node disturbance as the initial presentation and management by pacemaker implantation and was diagnosed with AL amyloidosis. This case highlights the importance of early recognition of AL amyloidosis manifestations in order to proper management and to improve patients' quality of life.

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