

# Acute Coronary Syndrome (ACS) with ST-Segment Elevation Revealing Cardiac Amyloidosis : A Case Report and Literature Review

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## Abstract

Cardiac Amyloidosis (CA) is an infiltrative cardiomyopathy characterized by the accumulation of fibrillar proteins in the myocardium. The deposition of these proteins disrupts the structure and function of the heart, leading to thickened walls, increased myocardial mass, diastolic dysfunction, and altered myocardial contraction, ultimately causing progressive heart failure. We report the case of a 65-year-old patient, S.L., admitted to our service with Acute Coronary Syndrome (ACS) with a posterior extended ST-segment elevation, Killip class I, semi-recent. She is hypertensive and diabetic; she previously underwent surgery for chronic constrictive pericarditis, presumed to be of tuberculous origin, and was treated accordingly. The echocardiogram revealed a hypokinetic left ventricle (LV), predominantly affecting the inferior, inferoseptal, and apical walls. The left and right atria were not dilated, and both ventricles were of normal size. Coronary angiography showed no significant anomalies. Cardiac MRI supported a diagnosis of localized amyloidosis. Through this case, we explore the various clinical forms of CA, referencing existing literature.

**Keywords :** acute coronary syndrome, cardiac amyloidosis, healthy coronary arteries.

## Introduction

Cardiac amyloidosis is the leading cause of restrictive cardiomyopathy in the Western world. Its diagnosis has been subject to codified recommendations, based on the recognition of syndromic entities where cardiac amyloidosis is frequently encountered (1-3). There are four major clinical entities that can lead to an early diagnosis: heart failure with preserved ejection fraction, myocardial hypertrophy, arrhythmias, and conduction disorders, and aortic stenosis (in patients over 65 years old). Although they are not highly specific, these are the most common cardiological presentations (Figure 1), encompassing the initial cardiac symptoms of patients with amyloidosis. The cardiac signs are not specific. You might find normal voltage or microvoltage discordant with ventricular hypertrophy on echocardiography, a pseudo-Q wave pattern in the anteroseptal region, conduction disturbances, or rhythm disorders. Echocardiography typically reveals hypertrophy (wall thickness  $\geq 12$ mm) or a restrictive pattern with a granular appearance of the myocardium, diffuse thickening of the ventricular walls extending to the valves and the interatrial septum, with a generally small pericardial effusion. Analysis of systolic function shows a discordance between a preserved left ventricular ejection fraction until late stages and

earlier impairment of longitudinal function, with alterations in tissue velocities, especially global longitudinal strain, displaying a typical target sign pattern. The association with aortic stenosis is common, particularly in low-gradient stenosis. Several diagnostic scoring systems based on echocardiographic criteria have been proposed recently (2-5). Cardiac biomarkers show an increase in NT-proBNP, but the level of troponins is higher, indicating myocardial involvement, which can initially mislead towards ischemic heart disease. Extracardiac signs can be found in systemic amyloidosis (4), which is not limited to AL amyloidosis: even moderate proteinuria, renal insufficiency, macroglossia, periorbital bruising, peripheral neuropathy, or dysautonomia. The presence of carpal tunnel syndrome, especially if it is bilateral in a man, is very suggestive of ATTRwt amyloidosis, often preceding cardiac manifestations by several years, as well as lumbar canal stenosis, biceps tendon rupture, or hearing loss. Certain factors will strengthen the diagnostic suspicion: the presence of hypotension or reduced need for antihypertensive treatment, and specific ECG and echocardiographic patterns. Once amyloidosis is suspected, a specific workup is needed to confirm or refute the diagnosis, with some forms representing a therapeutic emergency like AL amyloidosis (2) (Figure 2).

### Observation

We report the case of a 65-year-old patient, S.L., who has a history of hypertension and diabetes, and a previous pericardectomy due to chronic constrictive pericarditis, presumed to be of tuberculous origin. She was hospitalized in our department for acute coronary syndrome (ACS) with ST-segment elevation in the extended posterior region, semi-recent, Killip stage I, without rhythm complications. On clinical examination, there is a mid-thoracic surgical scar, and no murmurs or additional heart sounds. There are no signs of left or right heart failure, and her blood pressure is 120/75 mm Hg. The ECG shows an RSR pattern at 75 bpm, no low voltage, PR isoelectric at 180 ms, fine QRS axis in DI, ST-segment elevation in the extended posterior region, convex upward, with a mirror image in the basal and anterior regions. Laboratory results are unremarkable except for elevated troponin levels (554 times the normal value). Echocardiography shows the left ventricle (LV) is not hypertrophied, not dilated, with moderately impaired systolic function, hypokinesia in the inferior, inferoseptal, and apical regions. The left atrium is not dilated, and the right heart chambers are not dilated either. The right ventricle has good systolic function, there is no pulmonary hypertension, minimal pericardial effusion, and no signs of pericardial constriction (Figure 3). Coronary angiography revealed no coronary stenosis (Figure 4). Cardiac magnetic resonance imaging (MRI) is suggestive of localized cardiac amyloidosis (Figure 5).

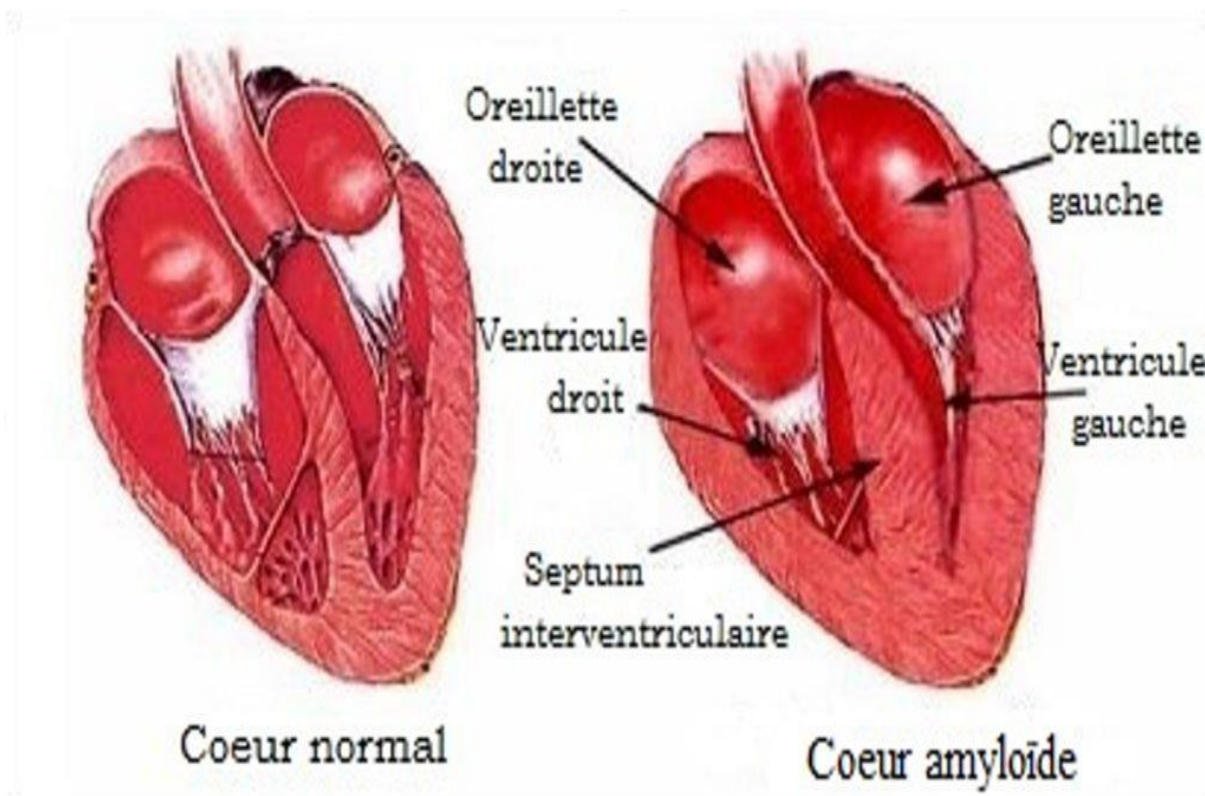
### Discussion

Cardiac involvement determines the prognosis of the disease. The symptoms of cardiac amyloidosis are heterogeneous and non-specific, making its diagnosis challenging. Chest pain can occur in rare cases, leading to a misdiagnosis of coronary artery disease (1), with the prevalence being uncertain; about 15% of cardiac amyloidosis patients have experienced angina-like pain. Three mechanisms can explain the occurrence of chest pain in cardiac amyloidosis (1-3): amyloid accumulation in the walls of small coronary arteries while the epicardial coronary arteries are normal; an extravascular cause, with perivascular and interstitial amyloid deposits potentially compressing vessels, thereby reducing diastolic perfusion time; and observed myocardial necrosis in areas surrounding vessels obstructed by amyloid infiltration, suggesting that cardiac amyloidosis could lead to myocardial ischemia. Another functional cause is mainly evidenced by PET scans, showing microvascular dysfunction (4, 5), with significantly

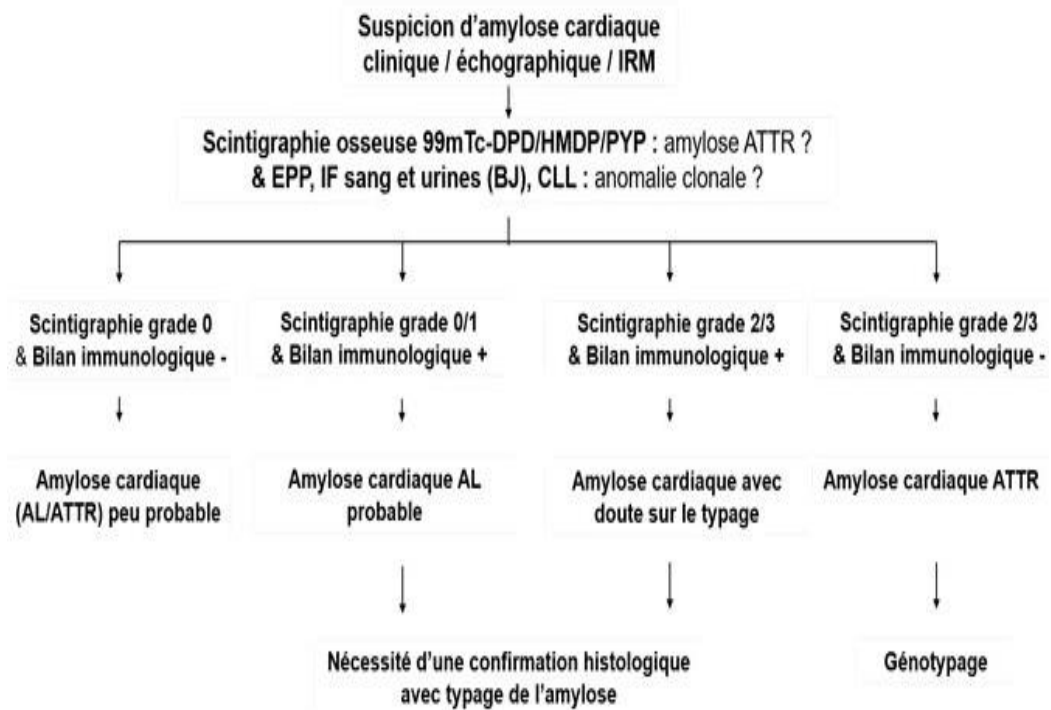
lower myocardial blood flow, lower coronary flow reserve, and higher coronary vascular resistance in cardiac amyloidosis patients compared to normal subjects. The gold standard for diagnosing cardiac amyloidosis is histological. To confirm AL amyloidosis before starting specific treatment, when histological evidence is required, the first sample is obtained through the least invasive methods: minor salivary glands or abdominal fat aspiration in experienced centers. The most recommended morphological examinations when cardiac amyloidosis is suspected are cardiac MRI and bone scintigraphy, the latter of which has gained a major role in the diagnostic algorithm for cardiac amyloidosis. Cardiac amyloidosis has long lacked specific treatment; the development of chemotherapies for AL amyloidosis and liver transplantation for ATTRv amyloidosis have opened new therapeutic avenues (6).

### Conclusion

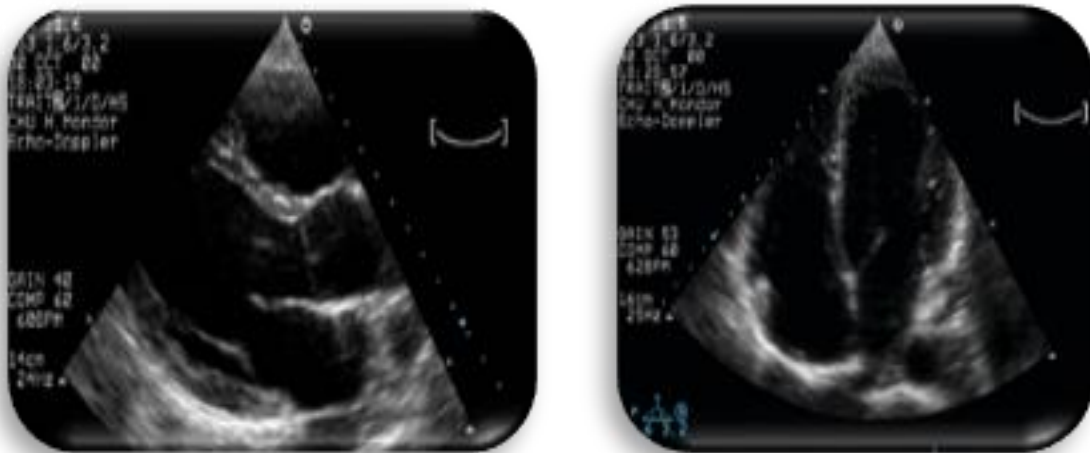
Cardiac amyloidosis is a rare and serious pathological entity. Diagnosis relies on a combination of clinical, biological, and morphological evidence. Epicardial coronary artery involvement is rare. Microcirculation impairment can mimic acute coronary syndrome, with healthy coronary arteries on coronary angiography. Cardiac MRI and histological studies help confirm the diagnosis and guide treatment adjustments.



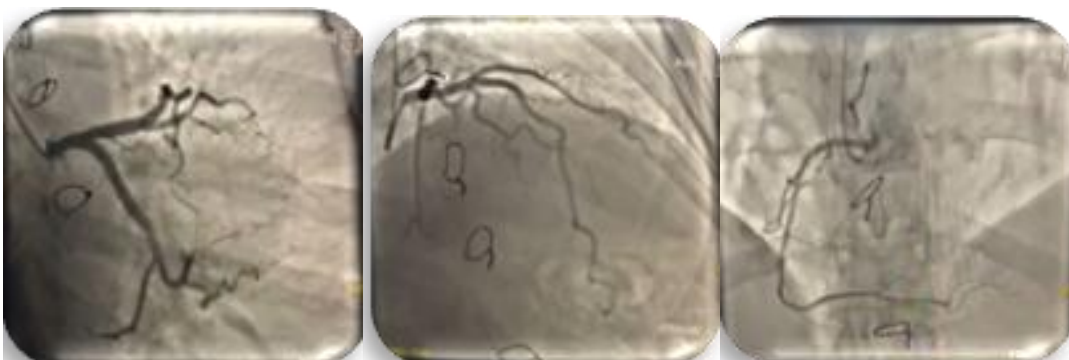
**Figure 1: Anatomical specimens comparing a normal heart with a heart affected by amyloidosis (5).**



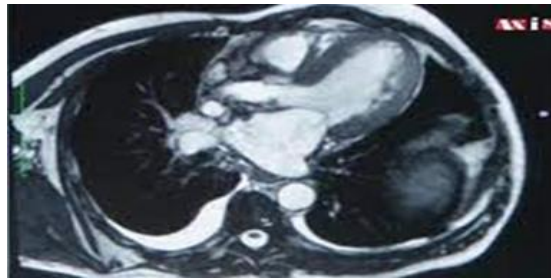
**Figure 2 : Diagnostic algorithm for cardiac amyloidosis (1,2).**



**Figure 3 : Echocardiography showing fibrous valves with normal-sized heart chambers.**



**Figure 4: Coronary angiography showing healthy coronary arteries.**



**Figure 5: Localized cardiac amyloidosis on the inferoapical wall of the left ventricle, diagnosed by cardiac MRI.**

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