

Cardiac Amyloidosis Revealing IgM-Kappa Waldenström Macroglobulinemia in an 84-Year-Old Man: A Case Report

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Abstract

Case Report

Background: Cardiac amyloidosis associated with Waldenström macroglobulinemia is rare and diagnostically challenging. In patients with a monoclonal gammopathy, low-grade uptake on bone-avid tracer scintigraphy is insufficient to confirm transthyretin amyloidosis; therefore, tissue confirmation through amyloid typing remains essential. **Case presentation:** An 84-year-old man presented with progressive heart failure and marked deterioration of his general condition. Cardiac assessment suggested a high probability of amyloid cardiomyopathy. Subsequent hematologic evaluation identified an IgM kappa monoclonal gammopathy, indicative of an underlying lymphoplasmacytic neoplasm. Bone marrow analysis confirmed Waldenström macroglobulinemia, while tissue biopsy demonstrated amyloid deposition. These combined findings established a diagnosis of systemic amyloidosis, with clinicopathologic evidence strongly supporting IgM-related kappa AL amyloidosis. **Conclusion:** This case demonstrates that cardiac amyloidosis can reveal an underlying IgM-secreting lymphoplasmacytic neoplasm. It emphasizes the importance of early multidisciplinary evaluation, careful interpretation of scintigraphy in the context of monoclonal gammopathy, and timely initiation of clone-directed therapy. Despite these interventions, the prognosis of cardiac involvement in frail elderly patients remains poor.

Keywords: Waldenström macroglobulinemia; AL amyloidosis; cardiac amyloidosis; lymphoplasmacytic lymphoma; case report.

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INTRODUCTION

Amyloidosis is a group of disorders in which misfolded proteins form insoluble fibrils that accumulate in the extracellular space of tissues and organs. According to the 2024 International Society of Amyloidosis update, 42 human amyloid fibril proteins have been identified, 19 of which are linked to systemic disease [1]. In cardiology, the main types are immunoglobulin light-chain (AL) amyloidosis and transthyretin (ATTR) amyloidosis [1, 2]. Notably, AL amyloidosis is the most common systemic form in Western countries, with an estimated incidence of 5–10 new cases per million population per year [3]. However, early symptoms are often nonspecific, leading to delayed diagnosis and worse outcomes [4, 5].

Cardiac involvement is the main determinant of prognosis in systemic amyloidosis. Cardiac amyloidosis

is usually suspected in patients with heart failure symptoms, otherwise unexplained increased left ventricular wall thickness, arrhythmias, elevated cardiac biomarkers, and suggestive findings on echocardiography or cardiac magnetic resonance imaging [2, 4]. In patients with a monoclonal gammopathy, however, tissue biopsy with amyloid typing remains necessary, because low-grade uptake on bone-avid tracer scintigraphy may also occur in AL amyloidosis. In this setting, a positive bone scintigraphy alone cannot establish ATTR amyloidosis, as it lacks specificity [2].

Waldenström macroglobulinemia is a rare IgM-secreting lymphoplasmacytic lymphoma that predominantly affects older adults. In the United States, its incidence is approximately 0.36 per 100,000 people, with higher rates in older adults, men, and White individuals [6]. Diagnosis relies on the identification of

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a monoclonal IgM protein and bone marrow infiltration by lymphoplasmacytic lymphoma, supported by immunophenotyping and, when feasible, MYD88 testing [7, 8]. Amyloidosis, though uncommon in Waldenström macroglobulinemia, is clinically significant because IgM-related AL amyloidosis accounts for 5–7% of all AL amyloidosis cases and has a distinct clinical profile [9, 10]. In this context, we present a case of cardiac AL amyloidosis that led to the diagnosis of Waldenström macroglobulinemia in an octogenarian patient, highlighting the importance of recognizing this association in clinical practice.

CASE PRESENTATION

An 84-year-old man with a 10-year history of hypertension treated with perindopril and indapamide and a remote history of inguinal hernia repair presented to the emergency department with New York Heart Association class III dyspnea, bilateral lower-limb edema, and general deterioration characterized by severe asthenia, anorexia, and unquantified weight loss. He was

initially admitted to the cardiology department for congestive heart failure and was subsequently transferred to the clinical hematology department after profound anemia was identified.

The admission electrocardiogram revealed sinus rhythm, first-degree atrioventricular block, left anterior fascicular block, and a QS pattern in the anteroseptal-apical leads (Figure 1). Initial laboratory tests showed severe normochromic normocytic anemia, with hemoglobin at 2.8 g/dL, mean corpuscular volume of 96.5 fL, mean corpuscular hemoglobin concentration of 34.1 g/dL, leukocyte count of $6.04 \times 10^9/L$, and platelet count of $148 \times 10^9/L$. Troponin was elevated at 52 ng/L and NT-proBNP at 3226 ng/L. Renal function was impaired, with a creatinine level of 136 $\mu\text{mol/L}$ and an estimated glomerular filtration rate of 44 mL/min/1.73 m². The markedly reduced reticulocyte count, together with the absence of folate or vitamin B12 deficiency, indicated hypoproliferative anemia, likely multifactorial in this clinical context (Table 1).

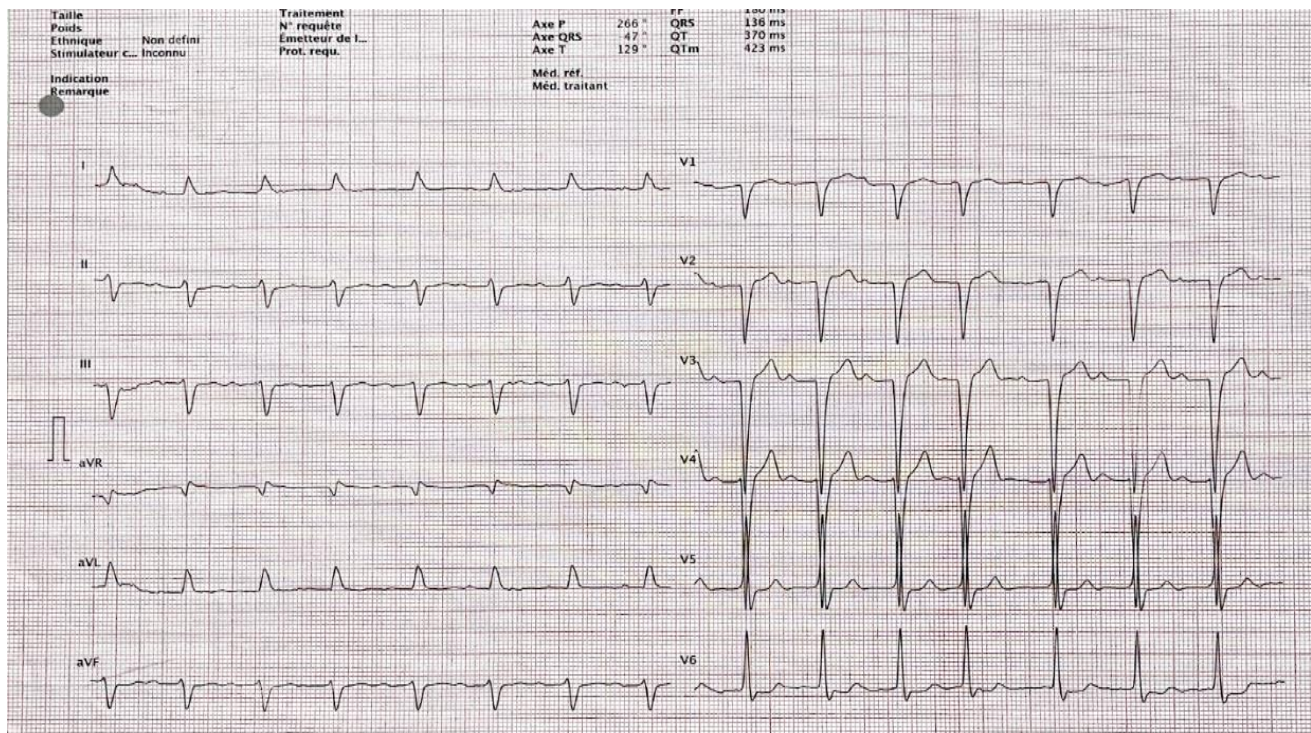


Figure 1: Twelve-lead electrocardiogram at admission showing sinus rhythm, first-degree atrioventricular block, left anterior fascicular block, and an anteroseptal-apical pseudo-infarct pattern

Table 1: Blood test results on admission

Parameter	Result	Reference range
Sodium (mmol/L)	136	136-145
Potassium (mmol/L)	4.5	3.5-4.6
Total protein (g/L)	87	64-83
Albumin (g/L)	33	35-50
Creatinine ($\mu\text{mol/L}$)	136	60-120
eGFR by CKD-EPI (mL/min/1.73 m ²)	44	90-140
Troponin (ng/L)	52	<14
NT-proBNP (ng/L)	3226	<300
White blood cell count (G/L)	6.04	4-11

Hemoglobin (g/dL)	2.8	13-18
Mean corpuscular volume (fL)	96.5	78-98
Neutrophils (G/L)	1.11	1.4-7.7
Lymphocytes (G/L)	4.29	1.0-4.8
Monocytes (G/L)	0.64	0.18-1.0
Platelet count (G/L)	148	150-450
Reticulocytes (G/L)	0.034	-
Fibrinogen (g/L)	2.2	2.00-4.00
C-reactive protein (mg/L)	4	<5
Serum iron (µmol/L)	36.8	10-28
Transferrin (g/L)	1.65	2-3.6
Total iron-binding capacity (µmol/L)	41.25	44.8-73.4
Transferrin saturation (%)	89.21	20-40
Ferritin (ng/mL)	583	30-400
Folate (ng/mL)	5.6	1.6-20.5
Vitamin B12 (pg/mL)	396	187-883

Transthoracic echocardiography showed findings strongly suggestive of infiltrative amyloid cardiomyopathy. The myocardium and valves had a granular, infiltrated appearance. The interventricular septum and posterior wall measured 14 mm and 13 mm, respectively, and the interatrial septum was also

thickened. Both atria were enlarged. The left ventricular ejection fraction was preserved at 59%. The right ventricle showed signs of infiltration and hypertrophy. Global longitudinal strain was markedly reduced at -10.6%, with a base-to-apex gradient indicating relative apical sparing (Figure 2).

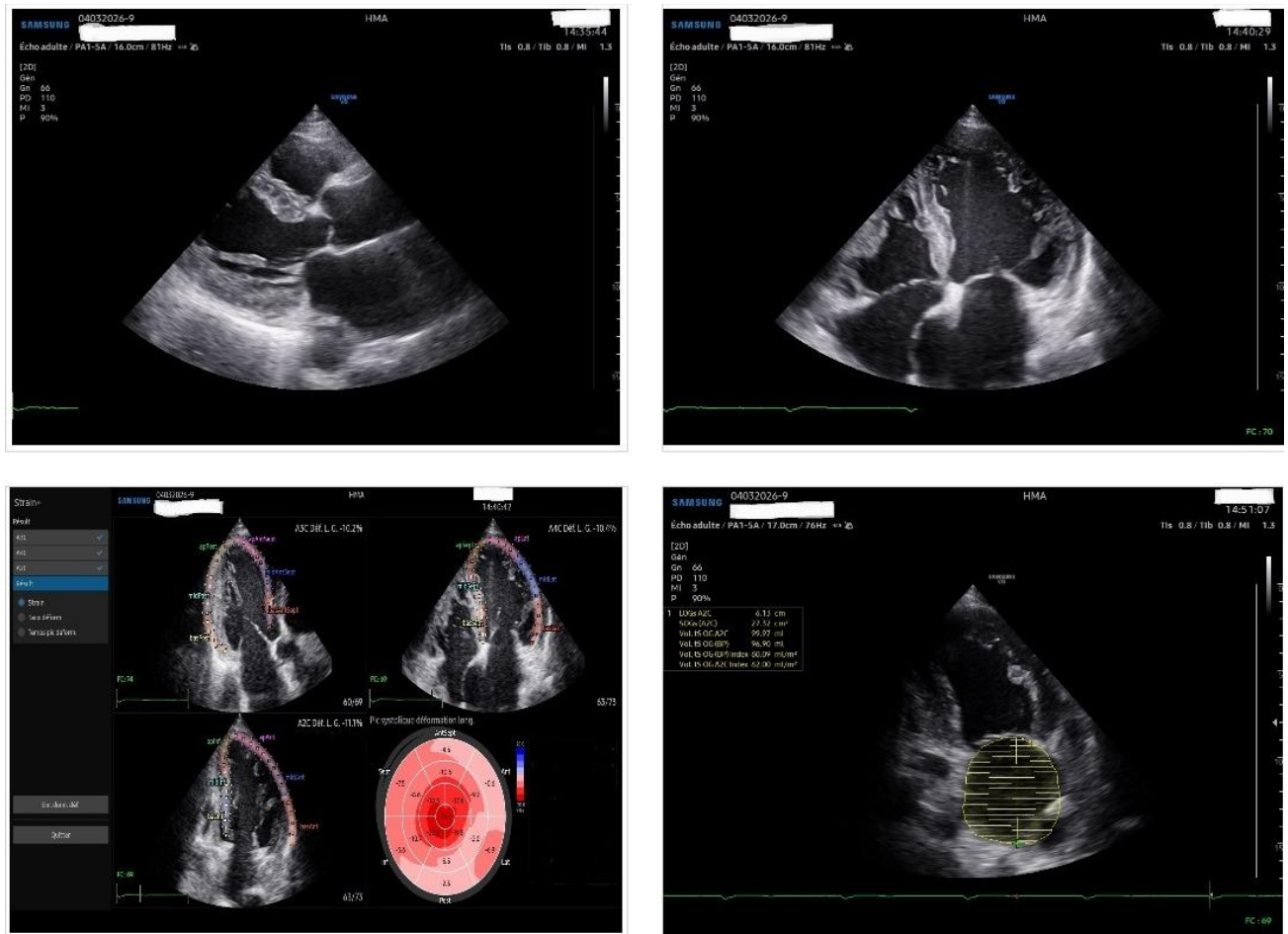


Figure 2: Transthoracic echocardiographic evaluation showing infiltrative cardiomyopathy and markedly impaired global longitudinal strain with relative apical sparing

Serum protein electrophoresis showed monoclonal hypergammaglobulinemia estimated at 38.8

g/L (Figure 3). Serum immunofixation confirmed the presence of an IgM kappa monoclonal protein. Urine

immunofixation detected small amounts of monoclonal free kappa light chains. The serum free light-chain assay showed an elevated free kappa light-chain level of

110.75 mg/L, a free lambda light-chain level of 11.76 mg/L, and an abnormal kappa/lambda ratio of 9.42.

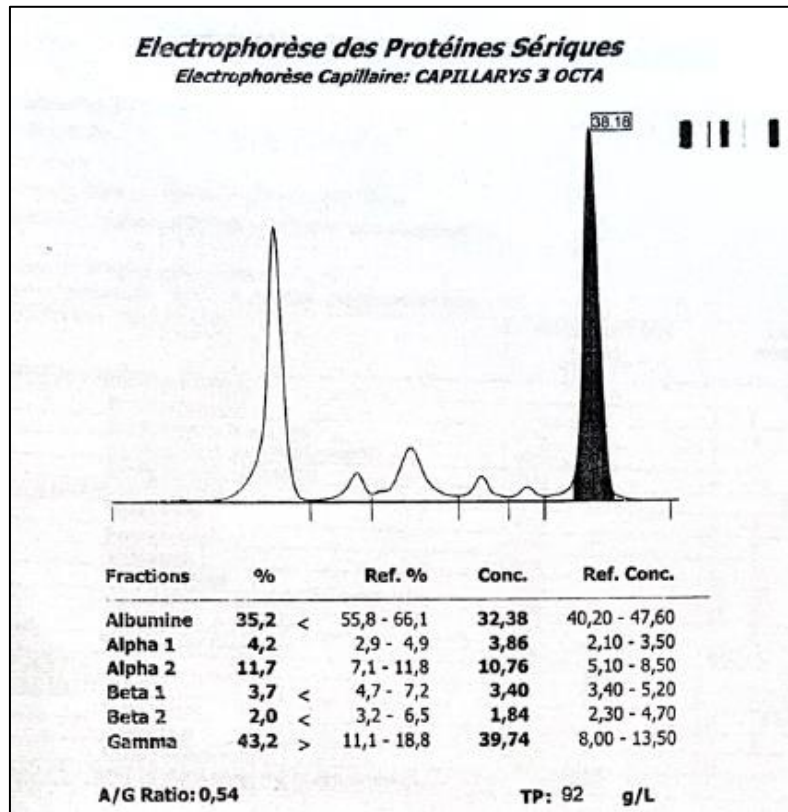


Figure 3: Serum protein electrophoretic profile suggestive of monoclonal hypergammaglobulinemia

The bone marrow aspirate was highly cellular, containing numerous megakaryocytes, 9% plasma cells, 22% lymphoplasmacytic cells, and 57% small to medium-sized lymphocytes characterized by a high nuclear-to-cytoplasmic ratio, regular nuclei with dense chromatin, and agranular cytoplasm. The other hematopoietic lineages were reduced, without significant cytologic abnormalities (Figure 4). Bone marrow biopsy

showed a kappa-restricted lymphoplasmacytic infiltrate; the B-cell component was CD20-positive, the plasmacytic component CD138-positive, and admixed T cells were CD3-positive. In the context of a serum IgM kappa paraprotein, these morphologic and immunophenotypic findings are consistent with a diagnosis of Waldenström macroglobulinemia.

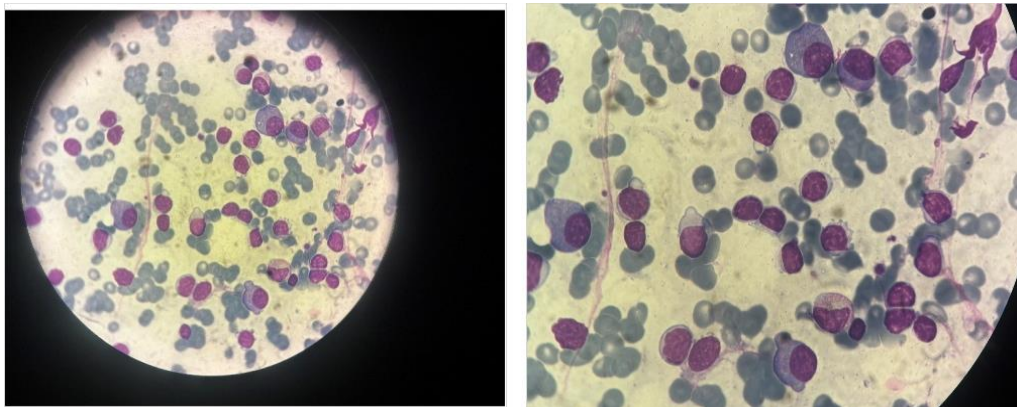


Figure 4: Bone marrow smear, May-Grünwald-Giemsa stain, showing infiltration by a lymphoplasmacytic population composed of lymphocytes and lymphoplasmacytoid cells

Congo red staining of a minor salivary gland biopsy revealed the characteristic yellow-green

birefringence under polarized light, confirming systemic amyloidosis. Immunohistochemical analysis identified

abundant kappa light-chain deposits, supporting IgM-related AL amyloidosis in this clinicopathologic context.

Technetium-labeled cardiac scintigraphy showed grade 1 myocardial uptake according to the Perugini visual score (Figure 5).

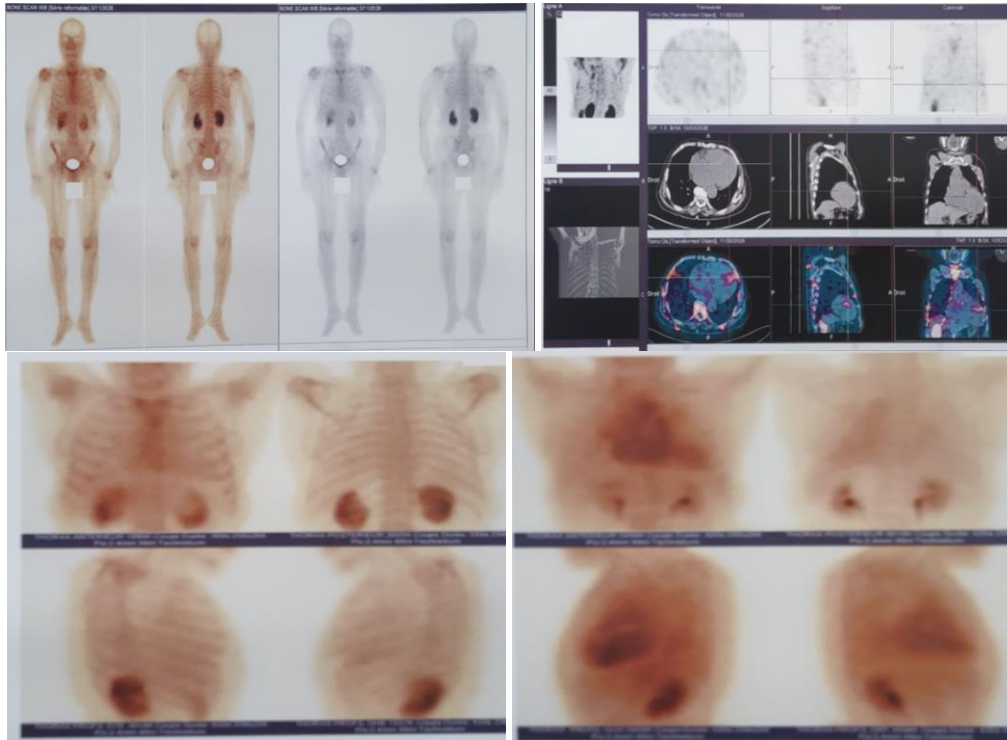


Figure 5: Technetium-labeled cardiac scintigraphy

Initial management included repeated transfusions of packed red blood cells and intravenous furosemide-based decongestive therapy. Subsequently, the patient received rituximab, cyclophosphamide, and dexamethasone every four weeks as individualized clone-directed therapy, given his advanced age, severe anemia, and cardiac involvement. Before chemotherapy, troponin I was 52 ng/L and NT-proBNP was 2050 ng/L. Cardiac monitoring included echocardiography and serial troponin I and NT-proBNP measurements. Over the first two cycles, the patient showed progressive improvement in symptoms, clinical signs, and laboratory parameters. His family later informed us that he had died suddenly at home, with no apparent precipitating cause.

DISCUSSION

This case combines advanced age, profound anemia, and heart failure; a thorough hematologic and pathologic work-up led to the diagnosis of Waldenström macroglobulinemia-associated AL amyloidosis. Published case reports indicate that this association frequently presents as restrictive or infiltrative cardiomyopathy, neuropathy, edema, chronic diarrhea, renal dysfunction, or unexplained general decline, rather than with typical lymphoma symptoms [11, 12]. This heterogeneous presentation contributes to diagnostic delay, which is clinically relevant because a longer time to diagnosis is associated with higher mortality in AL amyloidosis [5].

Several red flags were present in our patient. The electrocardiographic pseudo-infarct pattern and conduction delay, together with increased left ventricular wall thickness and preserved ejection fraction, elevated cardiac biomarkers, and a clearly abnormal strain pattern with apical sparing, strongly supported the diagnosis of cardiac amyloidosis [2, 4, 11]. Low QRS voltage is classically described but is not required, and its absence should not rule out the diagnosis when imaging and laboratory findings are suggestive [2, 4]. Bone scintigraphy was also informative: grade 1 myocardial uptake in a patient with a monoclonal gammopathy does not establish transthyretin amyloidosis and should prompt tissue confirmation and amyloid typing [2, 9, 10].

The hematologic evaluation was similarly conclusive. According to current guidelines, Waldenström macroglobulinemia is defined by the presence of serum monoclonal IgM and bone marrow infiltration by lymphoplasmacytic lymphoma [7, 8]. Our patient met these criteria, with an IgM kappa paraprotein, an abnormal free light-chain profile, and a marrow infiltrate composed predominantly of kappa-restricted lymphoplasmacytic cells. The minor salivary gland biopsy then confirmed systemic amyloidosis and pointed to kappa AL amyloidosis. While immunohistochemistry supported the diagnosis in this case, mass spectrometry remains the reference standard for amyloid typing when available and is particularly valuable when scintigraphic

or immunohistochemical findings are inconclusive [9, 10, 11].

IgM-related AL amyloidosis exhibits distinct biological and clinical characteristics compared with the more common non-IgM AL form. Representing only 5–7% of AL amyloidosis cases, it more frequently involves soft tissue, lymph nodes, lungs, and peripheral nerves, while cardiac involvement is less common but remains highly consequential [9]. Within Waldenström macroglobulinemia cohorts, amyloidosis is rare yet clinically significant. In a Mayo Clinic series, 7.5% of patients with Waldenström macroglobulinemia presented with coexisting AL or AHL amyloidosis, and synchronous presentation was associated with significantly shorter survival (median overall survival, 2.5 years) [13]. In a more recent cohort of 49 patients with Waldenström macroglobulinemia-associated AL amyloidosis, median overall survival reached 7.3 years; however, renal dysfunction and elevated natriuretic peptide levels remained predictors of poorer outcomes [14].

The management of Waldenström macroglobulinemia-associated AL amyloidosis focuses on rapid suppression of the pathogenic clone and preservation of organ function. Rituximab-based chemoimmunotherapy, proteasome inhibitor-based regimens, alkylating agents, and Bruton tyrosine kinase inhibitors are commonly used in Waldenström macroglobulinemia, while bortezomib-based combinations remain central to the treatment of systemic AL amyloidosis [8, 10, 16]. In the absence of a standard of care for IgM-related AL amyloidosis, therapeutic strategies should be tailored to patient age, frailty, organ involvement, and the urgency of hematologic response [9, 10]. In the present case, the use of rituximab, cyclophosphamide, and dexamethasone was justified by the patient's advanced age, severe anemia, and cardiac involvement. Despite short-term clinical improvement after two cycles, the subsequent sudden death underscores the persistent arrhythmic and hemodynamic risks associated with cardiac amyloidosis, even when early symptomatic benefit is achieved [4, 17].

Several features set our case apart and enhance its educational value: the patient's very advanced age, profound hypoproliferative anemia at presentation, diagnosis based on a minor salivary gland biopsy rather than cardiac tissue, and only low-grade myocardial tracer uptake despite strong clinical evidence of cardiac involvement. These elements highlight the need to integrate clinical, imaging, hematologic, and pathologic data, rather than relying on a single diagnostic approach.

The main limitation of this report is the absence of definitive proteomic amyloid typing, which prevents absolute certainty regarding fibril composition. Therefore, the overall findings strongly support, rather than definitively establish, IgM-related kappa AL

amyloidosis. We explicitly acknowledge this limitation in the interest of transparency.

CONCLUSION

Cardiac amyloidosis may be the presenting manifestation of Waldenström macroglobulinemia. IgM-related AL amyloidosis should therefore be promptly considered in older patients presenting with heart failure, increased left ventricular wall thickness, an IgM monoclonal protein, and systemic symptoms. In this setting, low-grade uptake on bone scintigraphy does not exclude AL amyloidosis, and tissue confirmation remains essential. Although early clone-directed therapy is required and may yield initial clinical improvement, cardiac involvement carries a persistently high residual risk.

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