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An Extensive Atypical Systemic Light Chain Amyloidosis With Undetectable Plasma Cell Dyscrasia: A Case Report

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Abstract

Background: ***Light-chain amyloidosis (AL amyloidosis) is a rare, debilitating, heterogeneous, and multisystemic disorder that is often fatal in the absence of treatment. This is because of the conversion of light chains (LCs) produced by plasma cell dyscarasia from their soluble state into organized fibrillar aggregates, resulting in progressive organ damage and dysfunction. The clinical features of the disease are nonspecific and depend on the type of organ affected. Overall prognosis depends on several clinical, laboratory, and pathological parameters.*** ***Case presentation:*** ***A young Algerian man presenting with the diagnostic challenge of atypical extensive systemic amyloidosis initially presenting with severe neurological manifestations despite a lower burden light chain level and undetected plasma cell dyscarasia that developed over 10 years.*** ***Conclusion:*** ***Atypical clinical presentation, undetectable plasma cell dyscarasia, and unusual disease evolution led to delayed diagnosis and devastating complications in the patient at the time of diagnosis. A very high clinical suspicion with a combination of several laboratory and histopathological tests is required to confirm the diagnosis. Although biopsy is the gold standard for the diagnosis of AL amyloidosis and confirmation of the involvement of each organ, noninvasive techniques with high sensitivity and specificity have been used.***

Keywords: ***Systemic amyloidosis; Light chain amyloidosis (AL amyloidosis); Light chain burden level; Plasma cell dyscarasia; Monoclonal gammopathy.***

1. Introduction

Systemic amyloidosis is a group of disorders characterised by accumulation of insoluble proteins in various tissues[1]. Light chain amyloidosis (AL amyloidosis) is the most common form of systemic amyloidosis because of the conversion of these light chains (LCs) from their soluble states into organised fibrillar aggregates that are deposited in tissues, resulting in progressive organ damage and dysfunction [1], [2]. LCs are produced by monoclonal plasma cell dyscrasia

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[3], [4]. The clinical features of the disease are nonspecific and depend on the type of organ affected mostly the heart, liver, kidneys, nervous system, and the gastrointestinal tract. However, the overall prognosis depends on several clinical, laboratory, and pathological parameters [5]. Here, we report a case of extensive atypical systemic AL amyloidosis in a young patient with undetectable plasma cell dyscrasia that developed over 10 years.

2. Case

A 49-year-old Algerian male patient, a smoker 20 pack years (abstinence since 2020), was a former security man with no medical history or a history of similar familial cases.

In 2014, at the age of 39 years, the patient complained of fatigue associated with distal, symmetrical, ascending paraesthesia and a sensation of heaviness affecting the lower limbs without motor dysfunction. In 2018, the clinical picture worsened by the progression of the sensory disorder in intensity and topography associated with slight motor deficits affecting mainly the lower limbs. By 2020, the patient started complaining of autonomic affection manifested by motility-related diarrhoea, light-headedness, erectile dysfunction, voiding disorders, and aggravation of the motor disorder, leading to paraplegia by 2024.

Cutaneous examination revealed nail dystrophy, dried skin with lichenified lesions, macular hyperpigmentation, and purpura, which mainly affected the extremities (Fig. 1). Ophthalmological examination revealed periorbital purpura (Figs. 2 and 3), abnormal pupillary light reflexes, corneal dystrophy on slit-lamp examination with intact visual acuity in both eyes, and normal fundoscopic examination. Cardiac examination revealed distant heart sounds and first-degree AV block on ECG. Echocardiographic assessment revealed concentric, hyperechoic biventricular wall thickening, biatrial enlargement, and slight pericardial effusion (Figs. 4, 5, and 6). Impaired diastolic function, but preserved systolic function (ejection fraction (EF), 75%), as shown in Fig. 7. Notably, the patient had a normal cardiac evaluation (ECG and echocardiography) in 2022.

Standard laboratory analysis (Table 1) revealed non-regenerative, normocytic, and normochromic anaemia associated with prolonged prothrombin time, normal partial thromboplastin time, and plasma fibrinogen levels. Liver function analysis showed high levels of gamma-glutamyl transferase (GGT) with normal hepatic enzyme, bilirubin, and albumin levels. The patient has presented with slightly fluctuating hepatic enzyme levels since 2021. The patient also presented with elevated Nt-pro-BNP levels; however, normal troponin levels were associated with mild proteinuria with preserved renal function and normal urinary sediments. In contrast, the glycaemic and lipid profiles, copper, ceruloplasmin, thyroid-stimulating hormone (TSH), vitamin B12, B9, ferritenemia, and calcium serum levels were normal. Serum protein electrophoresis and immunofixation did not reveal any monoclonal gammopathy. Further specialised laboratory analysis revealed elevated lambda chain levels with a normal κ /λ ratio. The coagulation factor deficiency panel test demonstrated a deficit in factor X and anti-plasmin levels, with elevated VWF antigen levels.

However, viral serologies (HBV, HCV, HIV, and syphilis), cryoglobulinaemia, autoimmune panel test, complement level, screening tests for Gaucher disease, Fabry disease, and Neiman pick were negative. As shown in the Table 1.

A skeletal X-ray survey showed no bone lesions, whereas thoracoabdominal CT revealed homogenous hepatomegaly at 21 cm, heterogeneous splenomegaly at 16 cm, and normal-sized kidneys with cortical heterogeneous contrast enhancement. Hepatic elasticity was 26 kPa on the fibro scan. However, the upper and lower gastrointestinal endoscopies were normal.

Bone scintigraphy using ^{99m}technetium methylene diphosphonate (^{99m}Tc MDP) demonstrated slight tracer uptake in the heart in favour of cardiac amyloidosis classified as grade1 according to the Perugini score [6] without other sites of abnormal tracer concentration, as shown in Fig. 8.

A nerve conduction study and needle electromyography indicated peripheral axonal sensorimotor neuropathy with partial and complete denervation in the upper and lower limbs, respectively. The diagnosis of AL amyloidosis was confirmed by a positive Congo red stain coupled with a direct immunofluorescence assay of the salivary gland biopsy, as shown in Figs. 9 and 10. However, rectal and duodenal biopsies were normal, without evidence of amyloid deposition. Bone marrow biopsy and aspiration revealed hypoplastic bone marrow with no amyloid deposits or abnormal cellular proliferation or infiltration.

The patient was transferred to the haematology department for induction therapy with cyclophosphamide, bortezomib, and dexamethasone (CyBorD) but passed away o2 months later.

Table 1: Quantitative Laboratory Investigations.

Laboratory test	Value	Normal range
Haemoglobin	11g/dl	13-16 g/dl
Reticulocyte count	48000 ele/mm3	20-80
Creatinine	10 mg/l	8-13mg/l
ALT	2021: 60 U/L 2022: 65U/L 2023: 73 U/L 2024: 14 U/L	8 -35 UI/L
AST	12 UI/l	8 -30 UI/L
GGT	2022: 35 UI/l 2023: 79 UI/L 2024: 167UI/L	< 45 UI/L
ALP	2022:75 UI/L 2023:35 UI/L 2024: 50 UI/L	30 - 125 UI/L
Troponin	0.07 ng/ml	0.00-0,3 ng/ml
Nt-pro BNP	4288 pg/ml	
Proteinuria	370 mg /24hr	<150mg/24hr
PT	IN 2022: 53%	70-100%

	In 2023: 64% In 2024: 60%	
λ	47.7 mg/l	5.71-26.3 mg/l
ϰ	17.6 mg/l	3.30-19.4 mg/l
factor X	28%	70-130%
Anti-plasmin	35%	70-120%
VWF antigen	269%	50-200%

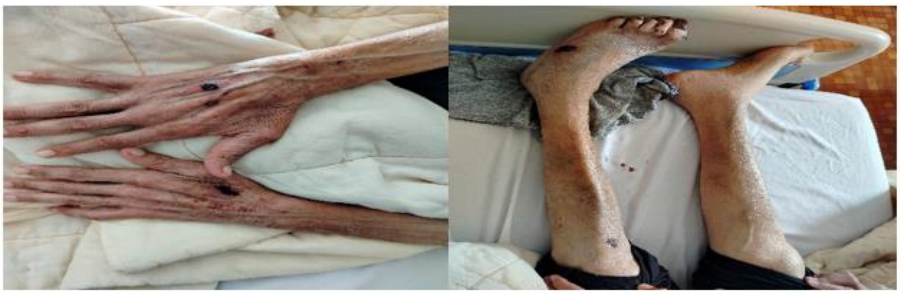

Fig. 1. Nail dystrophy, a dried skin with lichenified lesions, macular hyperpigmentation and purpura affecting mainly the extremities.

Fig. 2. Periorbital purpura.

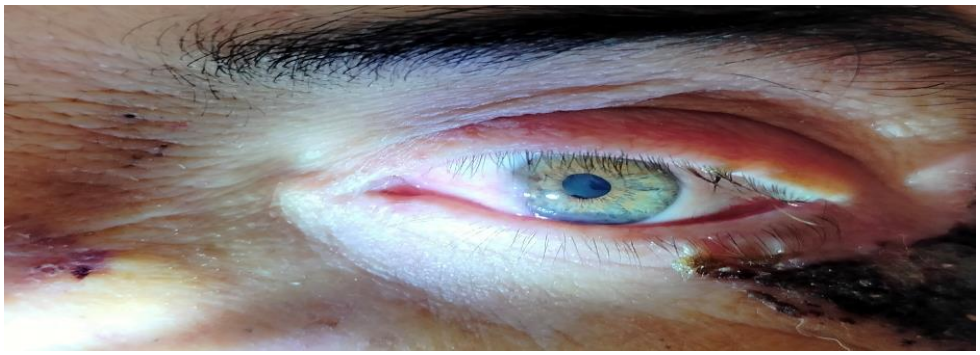

Fig. 3. Waxy lower eyelid papules with purpura and subconjunctival haemorrhage due to trivial trauma during hospitalisation.

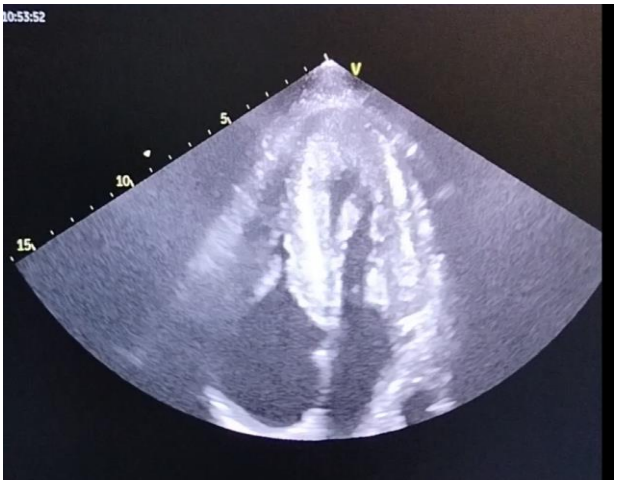

Fig. 4. Apical 4-chamber view demonstrating increased biventricular wall thickness with a sparkling texture of the myocardium.

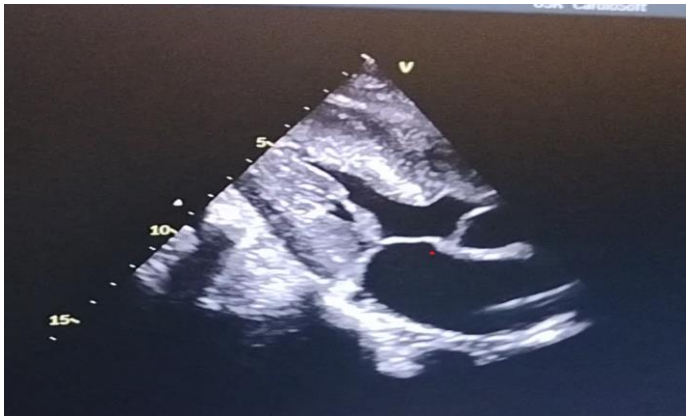

Fig. 5. Parasternal long axis view demonstrating ventricular wall thickness with a sparkling texture of the myocardium, slight pericardial effusion and an enlarged right atrium.

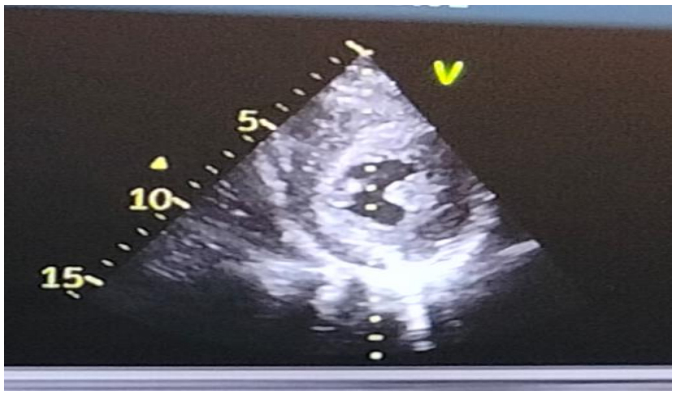

Fig. 6. Parasternal short axis demonstrates increased LV wall thickness with small pericardial effusion.

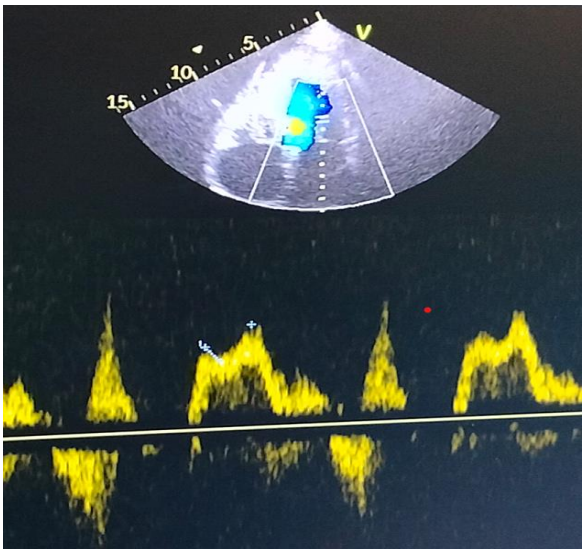

Fig. 7. Detection of early impaired diastolic function performed by measurement of trans mitral flow parameters through apical four chamber view with conventional pulsed wave Doppler showing: reduced amplitude of E velocity, increased A velocity, prolonged E-velocity DT with E/A at 0.8.

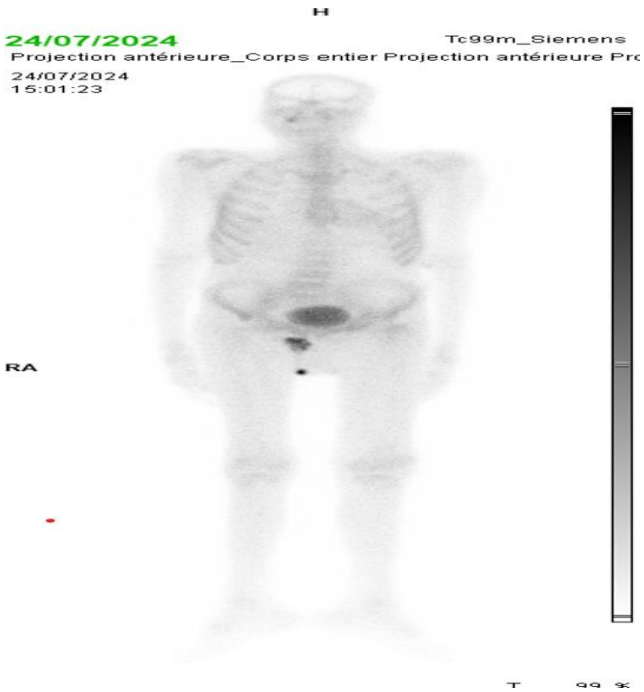

Fig. 8. Bone scintigraphy using 99mtechnetium methylene diphosphonate (99mTc MDP) demonstrated a slight increased tracer uptake in the heart in favour of cardiac amyloidosis classified as grade1 according to Perugini score without another site of abnormal tracer.

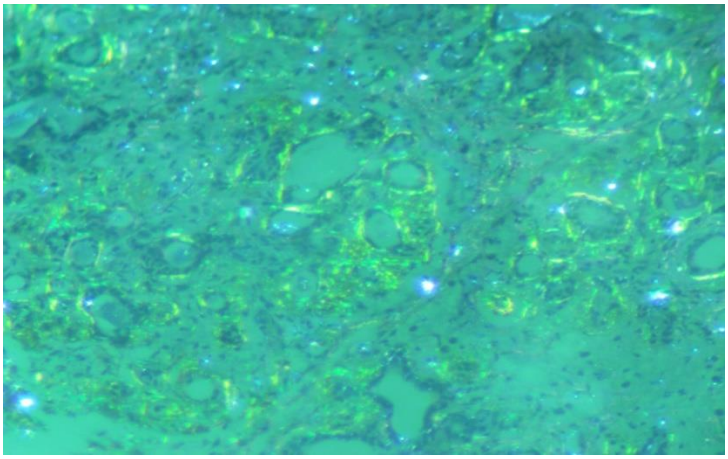

Fig. 9. Positive apple green refringence on Congo Red stain of salivary gland biopsy.

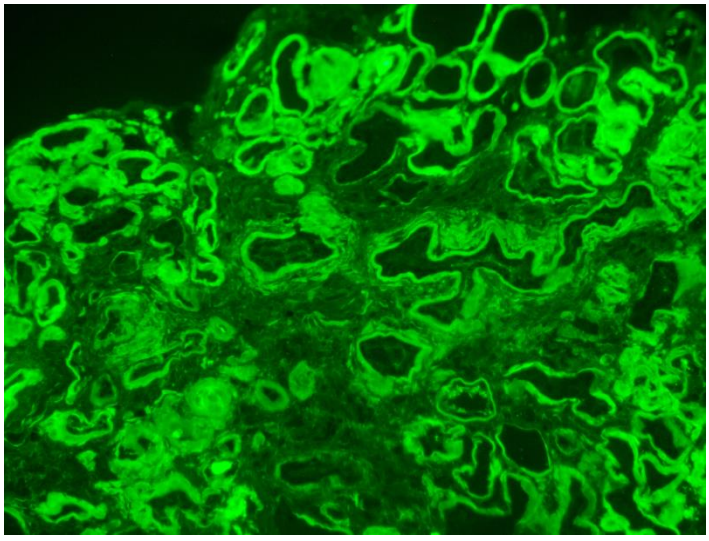

Fig.10. Direct immunofluorescence showing abundant lambda chain deposits in salivary gland but absent kappa chain deposits.

3. Discussion

Immunoglobulin amyloidosis is the most common form of amyloidosis, accounting for an annual incidence of 6 to 14 per million/year. Al amyloidosis is the most common subtype, with which 75% expressing λ chains [7], [8].

Our patient presented with initial symptoms of Al amyloidosis at an age of < 40 years, although, as reported by several studies, the median age at diagnosis was in the mid-60s[7]. However, a US study conducted on 1,722 patients with AL amyloidosis reported 45 (2.6%) patients being diagnosed at an age ≤ 40 years had better survival rate after treatment [9].

The clinical features of the disease depend on the type of organs affected which are usually nonspecific. Constitutional manifestations such as fatigue and weight loss are the most commonly reported symptoms [10], [11]. The other clinical features of the disease depend on the type of organ that mostly affects the heart, liver, kidneys, nervous system, and gastrointestinal tract [11]. However, the first reported manifestations in our case were in the form of fatigue and neurological disorders in the year 2014.

Neurological affection is not a typical prominent feature of Al amyloidosis as to cardiac and renal affection, mainly manifested by mixed sensory and motor peripheral neuropathy in 20% as to 15% with autonomic neuropathy [11, [12]. Only 15% of patients are presented with peripheral neuropathy at the onset of the disease were as 35%) during the disease progression [12]. Al amyloidosis initially presenting with neurological manifestations makes the diagnosis challenging and requires high suspicion to identify such patients[11,12]. Amyloid peripheral polyneuropathy is symmetrical, length-dependent, slowly progressive with a painful sensory-dominant character and early involvement of the lower limbs, whereas autonomic affection usually manifests as orthostatic hypotension, bowel and bladder dysfunction [12]–[14]. Nerve conduction studies and needle electromyography mainly showed axonal sensorimotor neuropathy and denervation changes, as reported previously [13]–[15]. Although our patient presented with typical neurological involvement according to the **updated** non-invasive consensus diagnostic criteria for amyloid–related organ involvement [16] however, the atypical onset of the disease made the diagnosis highly challenging at the early stage of the disease.

Heart is the most affected organ with systemic involvement, reported in 71% at the time of diagnosis, nonetheless it is one of the important prognostic factors accounting for 75% cause of death[10], [11]. The survival rate in the absence of treatment was less than one year [10]. Cardiac involvement mostly manifests clinically as heart failure or arrythmia (10-15% as atrial arrhythmia) and rarely by ischaemic manifestations [11]. NT-proBNP is the most important serological marker that reflects the degree of myocardial damage and the response to treatment [10], [11], [17]. ECG findings usually demonstrate low-voltage and slow atrial conduction due to amyloid infiltration [10], [17]. Cardiac amyloidosis usually affects diastolic functions, manifested by concentric cardiac hypertrophy and interventricular septum, reduced strain, and preserved ejection fraction in echocardiographic evaluations [10], [11], [17] . An endomyocardial biopsy is the gold standard for the diagnosis of cardiac amyloidosis [11]. However, non-invasive imaging modalities such as 1) cardiac MRI late gadolinium enhancement with a sensitivity of (80–100%) and a negative predictive value of 85–100%; 2) scintigraphy with 99mTechnetium (Tc)-bisphosphonate derivatives have also reported to have a high sensitivity and specificity for differentiating AL from transthyretin amyloid (ATTR) cardiac amyloidosis, of which (99mTc-pyrophosphate(PYP) has the highest sensitivity and specificity, could replace cardiac biopsy[10], [11], [18], [19]. However, our patient presented with a typical picture of stage II cardiac amyloidosis (Mayo 2012 Staging System) according to the **updated** non-invasive consensus diagnostic criteria for amyloid–related organ involvement and expert consensus recommendations for multimodal imaging in cardiac amyloidosis [16], [18]–[20]. The asymptomatic, delayed, and slowly progressive nature of cardiac involvement in our patient made it an unusual

disease evolution. Hepatic involvement in systemic amyloidosis accounts for 9% [21]. Clinical hepatic involvement accounts for 30% whereas, autopsies reported a higher incidence of 56-95% [21]. Hepatomegaly is the most reported manifestation accounting for 81-92% followed by borderline abnormal liver function tests, but rarely portal hypertension or hepatic failure [21], [22]. Hepatocytes are severely compressed by the extensive accumulation of amyloid in the parenchyma, along the sinusoids within the space of Disse, or in blood vessel walls, leading to an enlarged liver with a rubbery elastic consistency and eventual atrophy of hepatocytes [22]. Since hepatomegaly could be related to other causes, such as heart failure, hepatic biopsy remains the gold standard for confirming the hepatic involvement of amyloidosis but has a high risk of hepatic rupture and bleeding, especially with associated bleeding diathesis and advanced hepatic involvement [21]–[23]. According to the 2005 criteria for organ involvement and treatment response, liver involvement is defined as either hepatomegaly (defined as total craniocaudal liver span greater than 15 cm in the absence of heart failure) or when the serum alkaline phosphatase (ALP) value is above 1.5 × the upper limit with proven amyloidosis at another site, with a sensitivity and specificity of 50% and 74% but has never been validated systematically against the liver biopsy[16], [24]. SAP scintigraphy can be regarded as second substitute standard for liver involvement, with a sensitivity and specificity of 63% and 90% to detecting and quantifying amyloid deposits in the liver, spleen, and adrenal glands, but poor sensitivity for cardiac and neurological involvement [16], [24], [25]. Liver stiffness is another promising noninvasive modality with a cut-off value of 14.4 kPa and might be a good substitute to replace the liver span within the current consensus criteria for liver involvement [24]. Liver biopsy was avoided in our case because of the associated bleeding diathesis and increased risk of bleeding. However, the typical clinical features, such as hepatomegaly, slight alteration in hepatic enzymes, increased hepatic elasticity, absence of clinical features of right-sided heart failure, and elimination of other aetiologies, make it more of a hepatic involvement rather than a distinct hepatopathy.

The Kidney involvement is reported in 58% cases of Al amyloidosis with 25% of these patients progressing to end-stage renal disease (ESRD) and requiring renal replacement therapy [16], [25], [26]. It usually manifests as mild-to-nephrotic range of proteinuria with progressive impairment of renal function[16,26]. Although renal involvement has no significant impact on the survival rate compared to cardiac involvement, the development of ESRD increases morbidity and leads to treatment limitations [26]. ESRD development is mainly affected by disease duration and delayed diagnosis [25], [26]. The Pavia renal staging model stratifies patients based on their risk of progression to dialysis using the 24 h proteinuria (UPr) to eGFR ratio (UPr/eGFR ratio) [25], [26]. Unlike cardiac involvement, there are no imaging techniques that can replace the biopsy; however, certain radiological signs might aid in the diagnosis, such as enlarged kidneys, hyperechogenicity, and heterogeneous enhancement [25]–[27]. Although our patient presented with mild proteinuria, its occurrence in the context of systemic amyloidosis with elimination of other causes of proteinuria makes it a strong argument for renal involvement. However, despite the delayed diagnosis and prolonged disease duration of 10 years, the patient presented with early stage renal involvement (stage I according to the Pavia renal staging model) [26] .

Splenic involvement in amyloidosis accounts for 5-10% of systemic amyloidosis whereas primary splenic amyloidosis (PSA) is unusual [28], [29]. Amyloid in the spleen was identified in approximately 83% of patients at the time of autopsy, although it is usually asymptomatic [30]. It can manifest clinically as splenomegaly, splenic rupture, or hyposplenism with a high risk of infection [30]. Several nonspecific radiological manifestations have been reported on conventional contrast-enhanced CT scans, such as splenomegaly, calcification, and poor contrast enhancement of the splenic parenchyma on conventional contrast-enhanced CT scans [30]. As elaborated above, MRI and SAP scintigraphy have higher sensitivity for the detection of splenic amyloid deposits [28], [30]. Although SAP scintigraphy was not effectuated, splenic involvement in our patient was highly evoked due to the following arguments: 1) evidence of multiple organ involvement, especially in the liver; and 2) elimination of other possible causes of splenomegaly, such as infectious, vascular, neoplastic, inflammatory, autoimmune, and metabolic disorders.

Cutaneous manifestations are reported in 30-40% of systemic Al amyloidosis [31]. The most frequent mucocutaneous features are purpura, petechiae, and ecchymoses, due to increased blood fragility caused by amyloid deposition induced by Valsalva or trauma [31], [32]. Macular, nodular, lichenoid, papule (waxy/hemorrhagic), pigmentary changes, alopecia, scleroderma-like, and nail dystrophy have also been reported in certain studies as manifestations of either systemic or localised cutaneous amyloidosis [31]–[33]. Macroglossia is also a frequent presentation, accounting for 20% of systemic amyloidosis [31]. Sicca syndrome was also reported as one of the initial manifestations in 14 patients in a study of 16 patients with primary amyloidosis [34]. Cutaneous examination in our case revealed nail dystrophy and dry skin with purpura, lichenified lesions, and macular hyperpigmentation mainly affecting the extremities.

Ocular manifestations can be one of the initial manifestations of the disease and are usually overlooked by physicians, leading to late detection. Reynolds et al. found that, ocular involvement was reported in 11.8% patients with systemic amyloidosis[35]. Primary systemic amyloidosis affects various ocular structures (Table 2) [35]–[37]. As mentioned above, our patient presented with periorbital purpura which was strongly associated with systemic primary amyloidosis associated with corneal dystrophy.

Table 1: Ocular Manifestations in Primary AL amyloidosis [35]–[38].

Structure	Involvement
Eyelids ,Orbit & Adnexa	Waxy eyelid papules with purpura (strongly related to systemic primary amyloidosis), Proptosis, diplopia, Ptosis, accommodative paresis, Keratoconjunctivitis sicca, upper lid mass
Conjunctiva	diffuse yellow infiltrative masses
Cornea	Lattice Stromal Dystrophy
Anterior Chamber	Scalloped pupils, amyloid particles pupillary margin

Al amyloidosis is also associated with significant thromboembolic and haemorrhagic events. Fotiou D et al. conducted a retrospective study of 450 newly diagnosed patients with systemic AL amyloidosis, evaluating the thrombotic and

bleeding complications with their risk factors reported, 6% venous thromboembolic events occurred in 5% arterial embolic events [39]. They also reported that lower albumin levels, lower eGFR, higher bone marrow infiltration, soft tissue involvement, immunomodulatory drugs, higher platelet counts, and prior thrombosis are the main thromboembolic risk factors [39]. Whereas, Significant bleeding events accounted for 9% and were associated with a mortality of 19% [40]. The mechanism of bleeding diathesis is related to hepatic involvement, amyloid angiopathy, and the associated coagulopathies [11], [39]. Acquired factor X deficiency and increased fibrinolysis were the most frequently reported clotting abnormalities [39], [40]. Patel et al. reported that FX deficiency was significantly associated with an advanced stage of disease rather than a bleeding risk factor and that it is a biomarker of an advanced stage [41]. However, Fotiou et al. reported that VWF Ag levels were associated with an increased risk of bleeding, whereas Gertz et al. recognized it as a new risk factor for early mortality, which requires further validation [3], [40]. Although our patient did not present with any history of a major bleeding event but had significantly low levels of FX and anti-plasmin in with a high level of VWF Ag at stage II of the disease, which evolved over 10 years.

As reported by Gertz et al., overall prognosis depends on several clinical, laboratory, cytogenetic, and pathological parameters [3]. Milani et al. reported that patients with a lower burden light chain level, that is, the difference between involved and uninvolved FLC (dFLC) <50 mg/L, were significantly characterised by less common and less advanced cardiac manifestations, a better performance status, a lower bone marrow plasma infiltrate, an increased chance of complete response, and lower risk mortality but more frequent and more severe renal involvement[41]. Similar findings were also reported by Dittrich et al. [42] our patient presented with atypical systemic amyloidosis initially presenting with severe neurological manifestations, including cardiac, liver, splenic, cutaneous, ophthalmological, bleeding diathesis, and a lower performance status but slight renal involvement despite the lower burden light chain level and undetected plasma cell dyscarasia, that is, absence of monoclonal gammopathy on serum immunoelectrophoresis and absence of bone marrow plasma cell proliferation.

As reported in the literature, unlike multiple myeloma, Al amyloidosis is usually associated with subtle or rarely undetectable plasma cell dyscarasia which makes the diagnosis highly challenging, as experienced with our patient, and therefore requires a high clinical suspicion with a combination of several laboratory and histopathological tests to confirm the diagnosis. Staron et al. reported A variable clinical presentation and progression of the disease, but a modest improvement in end-organ involvement function in 6 patients with systemic Al amyloidosis with undetectable plasma cell dyscarasia [43]. They also suggested that this rare entity may be explained by the following hypothesis: (1) Splenic plasma cells are responsible for the generation of light chains instead of bone marrow plasma cells. 2) Heterogenous N-glycosylation leads to the dissimilation of proteins, and therefore, the absence of monoclonal gammopathy. 3)The disease is in its early stages; therefore, the population of the affected plasma cells is undetectable. 4) Spontaneous remission of plasma cell dyscarasia but persistence of amyloid deposits in the affected organs for several years [44]. However, this hypothesis requires further studies for the future validation.

4. Conclusion

Al amyloidosis is a rare, debilitating, heterogeneous, multisystemic disorder that is often fatal in the absence of treatment. The nonspecific clinical presentation reported in AL amyloidosis can lead to misdiagnosis and delayed diagnosis. These challenges are often associated with devastating complications for the patient at the time of diagnosis. A high clinical suspicion with a combination of laboratory and histopathological tests is required to confirm the diagnosis. While biopsy remains the gold standard for diagnosing AL amyloidosis and confirming organ involvement, evolving radiological techniques like scintigraphy and MRI offer high sensitivity and specificity for diagnosis and organ involvement. In this case report, we present the diagnostic challenges faced during the diagnosis of atypical extensive systemic Al amyloidosis with atypical clinical features and evolution. We also emphasise the non-invasive techniques used to confirm the involvement of each organ.

5. Ethical approval

All procedures involving this study followed the institutional and national research committee's moral standards, the 1964 Helsinki Declaration, and its later amendments or comparable ethical standards. All authors declare that consent was obtained from the patient to publish this study.

6. Consent for Publication

Written consent for the use of clinical information was obtained from the patient.

7. Data and Materials Availability

The data used in the current study are available from the corresponding author upon reasonable request.

8. Competing Interests

The author declares no competing interests.

9. Funding

This study received no funding from any sources. This work was done according to the CARE guidelines.

10. Authors' Contributions

Thouraya Soualah conceived the research concept, conducted the clinical examinations, monitored the patients, gathered laboratory data, and composed the manuscript.

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REFERENCES

1. Senecal JB, Abou-Akl R, Allevato P, et al. Amyloidosis: a case series and review of the literature. J Med Case Rep. 2023;17(1):1-9. PubMed| Crossref
2. Palladini G and Merlini G. How I treat AL amyloidosis. Blood. 2022;139(19):2918-2930. PubMed| Crossref
3. Gertz MA. Immunoglobulin light chain amyloidosis: 2024 update on diagnosis, prognosis, and treatment. Am J Hematol. 2024;99(2):309-324. PubMed| Crossref
4. Ikura H, Endo J, Kitakata H, et al. Molecular mechanism of pathogenesis and treatment strategies for AL amyloidosis. Int J Mol Sci. 2022;23(11):6336. PubMed| Crossref
5. Baker KR. Light chain amyloidosis: Epidemiology, staging, and prognostication. Methodist Debakey Cardiovasc J. 2022;18(2):27-35. PubMed| Crossref
6. Hutt DF, Fontana M, Burniston M, et al. Prognostic utility of the Perugini grading of 99mTc-DPD scintigraphy in transthyretin (ATTR) amyloidosis and its relationship with skeletal muscle and soft tissue amyloid. Eur Heart J Cardiovasc Imaging. 2017;18(12):1344-1350. PubMed| Crossref
7. Leung N, Nasr SH. 2024 Update on classification, etiology, and typing of renal amyloidosis: A review. Am J Kidney Dis. Am J Kidney Dis. 2024 Sep;84(3):361-373. PubMed| Crossref
8. Quock TP, Yan T, Chang E, et al. Epidemiology of AL amyloidosis: a real-world study using US claims data. Blood Adv. 2018;2(10):1046-1053. PubMed| Crossref
9. Verma K, Staron A, Zheng L, et al. Systemic AL Amyloidosis in Patients Younger Than 40 Years of Age: Clinical Presentation and Outcomes. Clin Lymphoma Myeloma Leuk. 2024;24(12):869-872. PubMed| Crossref
10. Stelmach-Gołdyś A, Zaborek-Łyczba M, Łyczba J, et al. Physiology, diagnosis and treatment of cardiac light chain amyloidosis. J Clin Med. 2022;11(4):911. PubMed| Crossref
11. Vaxman I and Gertz M. Recent advances in the diagnosis, risk stratification, and management of systemic light-chain amyloidosis. Acta Haematol. 2019;141(2):93-106. PubMed| Crossref
12. Namiranian D and Geisler S. Neuromuscular complications of systemic amyloidosis. Am J Med. 2022;135:S13-S19. PubMed| Crossref
13. Qian M, Qin L, Shen K, et al. Light-chain amyloidosis with peripheral neuropathy as an initial presentation. Front Neurol. 2021;12:707134. PubMed| Crossref
14. Matsuda M, Gono T, Morita H, et al. Peripheral nerve involvement in primary systemic AL amyloidosis: a clinical and electrophysiological study. Eur J Neurol. 2011;18(4):604-610. PubMed| Crossref
15. Adams D, Lozeron P, Theaudin M, et al. Varied patterns of inaugural light-chain (AL) amyloid polyneuropathy: A monocentric study of 24 patients. Amyloid. 2011;18 Suppl 1:93-95. PubMed| Crossref
16. Gillmore JD, Wechalekar A, Bird J, et al. Guidelines on the diagnosis and investigation of AL amyloidosis. Br J Haematol. 2015;168(2):207. PubMed| Crossref

17. Dima D, Mazzoni S, Anwer F, et al. Diagnostic and treatment strategies for AL amyloidosis in an era of therapeutic innovation. JCO Oncol Pract. 2023;19(5):265-275. PubMed| Crossref
18. Hanna M, Ruberg FL, Maurer MS, et al. Cardiac Scintigraphy with technetium-99m-Labeled bone-seeking tracers for suspected amyloidosis: JACC review topic of the week. J Am Coll Cardiol. 2020;75(22):2851-2862. PubMed| Crossref
19. Dorbala S, Ando Y, Bokhari S, et al. ASNC/AHA/ASE/EANM/HFSA/ISA/SCMR/SNMMI expert consensus recommendations for multimodality imaging in cardiac amyloidosis: Part 1 of 2 - Evidence base and standardized methods of imaging. Circ Cardiovasc Imaging. 2021;14(7):E000029. PubMed| Crossref
20. Kumar S, Dispenzieri A, Lacy MQ, et al. Revised prognostic staging system for light chain amyloidosis incorporating cardiac biomarkers and serum free light chain measurements. J Clin Oncol. 2012;30(9):989-995. PubMed| Crossref
21. Wang YD, Zhao CY, and Yin HZ. Primary hepatic amyloidosis: a mini literature review and five cases report. 2012;11(5):721-727. PubMed| Crossref
22. Shin YM. Hepatic amyloidosis. Korean J Hepatol. 2011;17(1):80. PubMed| Crossref
23. Duve RJ, Moga TG, Yang K, et al. Hepatic amyloidosis with multiorgan involvement. ACG Case Rep J. 2023;10(4):e00999. PubMed| Crossref
24. Brunger AF, Tingen HSA, Bijzet J, et al. Diagnostic performance of liver stiffness as marker of liver involvement in systemic immunoglobulin light chain (AL) amyloidosis. Ann Hematol. 2025;104(1):653-663. PubMed| Crossref
25. Fedotov SA, Khrabrova MS, Anpilova AO, et al. Noninvasive diagnostics of renal amyloidosis: Current state and perspectives. Int J Mol Sci. 2022;23(20):12662. PubMed| Crossref
26. Shafqat A, Elmaleh H, Mushtaq A, et al. Renal AL amyloidosis: Updates on diagnosis, staging, and management. J Clin Med. 2024;13(6):1744. PubMed| Crossref
27. Renal amyloidosis | Radiology Reference Article | Radiopaedia.org. Accessed December 19, 2024. [Online]. Available: https://radiopaedia.org/articles/renal-amyloidosis?lang=us
28. Dong W, Liu L, Wan Z, et al. Case Report Primary splenic amyloidosis: A case report. Int J Clin Exp Med. 2017;10(2):3963-3966.
29. Buzalewski J, Fisher M, Rambaran R, et al. Splenic rupture secondary to amyloid light-chain (AL) amyloidosis associated with multiple myeloma. J Surg Case Rep. 2019;2019(3):1-3. PubMed| Crossref
30. Monzawa S, Tsukamoto T, Omata K, et al. A case with primary amyloidosis of the liver and spleen: Radiologic findings. Eur J Radiol. 2002;41(3):237-241. PubMed| Crossref
31. Ramachandran V. Mucocutaneous manifestations of systemic amyloidosis in a black patient. QJM: An Int J Med. 2022;115(4):237-238. Crossref
32. Li Y and Jiang Y. Unusual purpura as a first sign of amyloid light chain amyloidosis. Lancet Oncol. 2022;23(10):e479. PubMed| Crossref

33. Mehrotra K, Dewan R, Kumar J V, et al. Primary cutaneous amyloidosis: A clinical, histopathological and immunofluorescence study. J Clin Diagn Res. 2017;11(8):WC01. PubMed| Crossref
34. Yoon SH, Cho JH, Jung HY, et al. Exceptional mucocutaneous manifestations with amyloid nephropathy: A case report. J Med Case Rep. 2018;12(1):241. PubMed| Crossref
35. Reynolds MM, Veverka KK, Gertz MA, et al. Ocular manifestations of systemic amyloidosis. Retina. 2018;38(7):1371-1376. PubMed| Crossref
36. Lee PH, Liao IC, and Lee WJA. Rare presentations of primary amyloidosis as ptosis: A case report. BMC Ophthalmol. 2022;22(1):1-4. PubMed| Crossref
37. Hashemian H, Jabbarvand M, Khodaparast M, et al. Ocular Presentations of Amyloidosis. Amyloidosis. Published online June 12, 2013. doi:10.5772/53910.
38. Mathew R and Zeppieri M. Ocular Amyloidosis. StatPearls. Published online May 28, 2024. [Online]. Available: https://www.ncbi.nlm.nih.gov/books/NBK603727/
39. Fotiou D, Theodorakakou F, Spiliopoulou S, et al. Thrombotic and bleeding complications in patients with AL amyloidosis. Br J Haematol. 2024;204(5):1816-1824. PubMed| Crossref
40. Patel G, Hari P, Szabo A, et al. Acquired factor X deficiency in light-chain (AL) amyloidosis is rare and associated with advanced disease. Hematol Oncol Stem Cell Ther. 2019;12(1):10-14. PubMed| Crossref
41. Milani P, Basset M, Russo F, et al. Patients with light-chain amyloidosis and low free light-chain burden have distinct clinical features and outcome. Blood. 2017;130(5):625-631. PubMed| Crossref
42. Dittrich T, Bochtler T, Kimmich C, et al. AL amyloidosis patients with low amyloidogenic free light chain levels at first diagnosis have an excellent prognosis. Blood. 2017;130(5):632-642. PubMed| Crossref
43. Staron A, Kataria Y, Murray DL, et al. Systemic AL amyloidosis with an undetectable plasma cell dyscrasia: A zebra without stripes. Am J Hematol. 2020;95(2):E45-E48. PubMed| Crossref

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