

# Immunoglobulin Light Chain Amyloidosis (AL) Presenting as Periorbital Purpura in A 39 Years Female Patient

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## Abstract

*Immunoglobulin light chain amyloidosis (AL) is the most frequent form of amyloidosis in adults, being more common in the elderly and male patients. The cutaneous manifestations occur in about 30% and are important clue do diagnosis. We report a 39 year-old-female patient with periorbital purpura, peripheral polyneuropathy and cardiomyopathy diagnosed as amyloidosis and treated with melphalan and prednisolone.*

**Key words:** *Immunoglobulin light chain; amyloidosis; periorbital purpura; peripheral polyneuropathy; cardiomyopathy.*

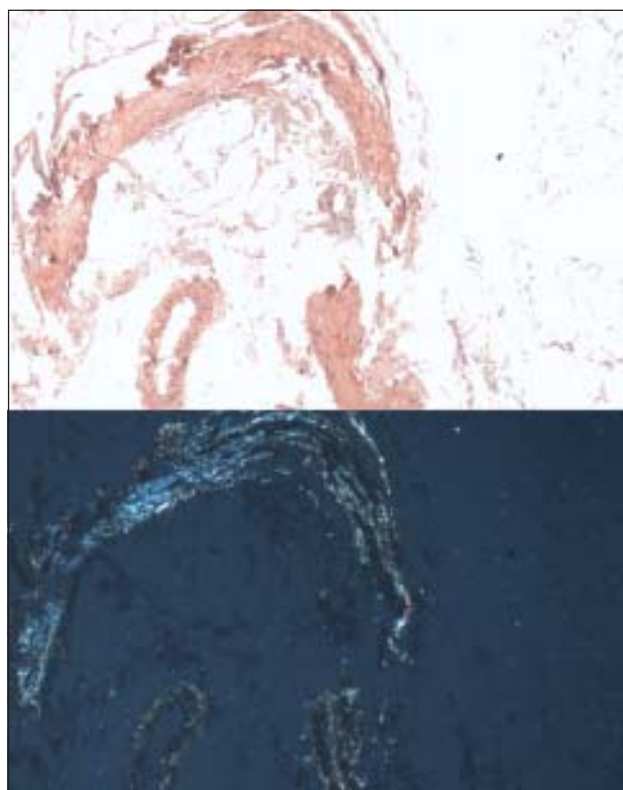
## Case Report

F.L.P.Q, 39 years, female, white, previously healthy, developed weight loss, fatigue, edema and periorbital purpura (Fig. 1), hoarseness, petechiae and easy bruising, symptoms that started 9 months before admission, evolving with painful paresthesia in ankles and feet, macroglossia,



**Fig.-1:** *Patient's face compatible with Amyloid purpura (Raccoons eyes)*

bilateral periorbital ecchymosis and postural hypotension. During investigation it was identified a monoclonal lambda protein in seric immunofixation and fat pad showed deposit of a pink, amorphous substance, stained with Congo Red, leading to green birefringence under polarized light (Fig. 2). The bone marrow aspirate was normocellular with mild plasmacytosis (5%) .



**Fig.-2:** *Abdominal fat pad (hematoxylin and eosin stained) presenting as a pink, amorphous substance, compatible with amyloid deposition stained with congo red leading to green birefringence under polarized light.*

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Electromyography showed distal and symmetrical axonal sensory polyneuropathy. On cardiac evaluation, electrocardiogram showed low voltage with the echocardiogram evidencing diffuse hypokinesia and diastolic dysfunction, compatible with cardiomyopathy by amyloid deposit.

The clinical picture associated with the absence of a family history of amyloidosis, the monoclonal lambda light chain component and the lack of chronic diseases that causes AA amyloidosis, strongly suggests the diagnosis of primary light chain amyloidosis, affecting cardiac, kidney, peripheral nervous system and skin. The patient was initially treated with prednisolone and melphalan, being referred for autologous bone marrow transplantation. Unfortunately she died of septic shock caused by lung infection.

### Discussion

Amyloidosis refers to the extracellular tissue deposition of fibrils composed of low molecular weight subunits of a variety of serum proteins, being the two most common causes Immunoglobulin light chain (AL) amyloidosis and AA amyloidosis in which the fibrils are composed of fragments of the acute phase reactant serum amyloid A.<sup>1</sup>

Light Chain Amyloidosis is considered a rare disease, with approximately 6 to 10 cases per million person-years and more common in older adults, with a slight male

preponderance. Less than 5% of the cases are in patients younger than 40 years.

The clinical manifestations are diverse, depending on the nature of the affected organs. Common clinical presentations include nephrotic syndrome, restrictive cardiomyopathy, peripheral neuropathy and non-specific symptoms, such as weight loss and fatigue.<sup>2</sup> Amyloid purpura appears in a minority of patients with amyloidosis. The purpura typically occurs above the nipple line and is often seen in the eyelids. Factor X deficiency, resulting from the binding of factor X to amyloid fibrils, is thought to be the one of the cause of the bleeding diathesis that may occur in patients with amyloidosis.<sup>3</sup>

In summary, we reported a rare case of AL amyloidosis in a young woman with fatal outcome. Systemic amyloidosis should be investigated in every patient with periorbital purpura, mainly in those with multiorgan symptoms, specially cardiac disease and peripheral neuropathy.

**Conflict of Interest:** None

### References

1. Comenzo RL. How I treat amyloidosis. *Blood*. 2009; 114:3147-3157.
2. Kyle RA, Linos A, Beard CM, et al. Incidence and natural history of primary systemic amyloidosis in Olmsted County, Minnesota, 1950 through 1989. *Blood*. 1992;79:1817.
3. Eder L, Bitterman H. Image in clinical medicine. Amyloid purpura. *N Engl J Med*. 2007;356:2406.