

# Peripheral Embolism in Nonamyloidotic Light-Chain Deposition Cardiomyopathy

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Nonamyloidotic light-chain cardiomyopathy is a paraproteinemia-induced disorder. Unlike amyloidosis, light chain deposition may be reversible with appropriate treatment. We report a case of fatal light chain deposition disease manifested as cardiomyopathy, which was complicated by arterial embolism despite the maintenance of sinus rhythm.

**Key words:** *Paraproteinemias. Cardiomyopathy. Embolism.*

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## Embolia periférica en la miocardiopatía no amiloidótica por depósito de cadenas ligeras

La miocardiopatía no amiloidótica por cadenas ligeras es secundaria a paraproteinemia. A diferencia de la amiloidosis, puede ser completamente reversible con tratamiento específico. Presentamos un caso fatal de enfermedad por depósito de cadenas ligeras libres manifestada por miocardiopatía y que presentó como complicación embolias arteriales en ritmo sinusal.

**Palabras clave:** *Paraproteinemias. Miocardiopatía. Embolia.*

## INTRODUCTION

Light-chain heart disease is caused by paraprotein deposition resulting from monoclonal plasma cell dyscrasia. Clinical symptoms resemble those of immune amyloidosis, which has specific histopathologic features.<sup>1</sup> The light-chain disease produces an infiltrate that is non-fibrillary and that can resolve after remission of the blood disorder.<sup>2-4</sup> We describe a lethal case of light-chain disease with peripheral ischemia that was probably caused by cardiac embolism.

## CASE REPORT

A 47-year-old woman was admitted to the hospital for acute ischemia of the left lower extremity. She had been followed for 8 months in an outpatient clinic for progressive symptoms of congestive heart failure. The outpatient work-up showed clinical and radiologic evidence of chronic congestive heart failure of unknown origin, and the patient was put on diuretics and vasodilators. Blood chemistry, including coagulation tests, was normal, except for the presence of proteinuria, which was 2.1 g/dL. An electrocardiogram showed si-

nus tachycardia and non-specific intraventricular conduction defects. An echocardiogram showed concentric ventricular thickening with highly echogenic myocardium and slightly depressed systolic function. Amyloid deposition was ruled out by rectal biopsy. The patient experienced sudden pain, pallor, and coldness of the left lower extremity with decreased distal pulses, and was admitted to the hospital for the first time and given anticoagulation treatment with heparin. She had dyspnea at rest and showed signs of severe congestive heart failure. Tests showed normal serum levels of thyroid hormones, ferritin, and angiotensin-converting enzyme. Serum creatinine was normal, and the erythrocyte sedimentation rate was 80 mm/h. Serum electrophoresis showed a monoclonal peak in lambda light chains and decreased levels of IgG, IgA, and IgM. There was Bence-Jones proteinuria, and urine electrophoresis confirmed the monoclonal peak in lambda light chains (3.5 mg/dL). Figure 1 shows the results of echocardiography. The patient evolved poorly and died from rapid progression of congestive heart failure over a 5-day period, despite aggressive medical treatment. Areas of non-amyloidotic lambda light chain deposition in the myocardium (Figure 2), kidneys, pancreas, and spleen were seen at autopsy. Recent renal infarction due to embolism was also noted, and mural thrombi were found in both atria, with partial organization in the right atrium. Bone marrow had a normal cellular appearance.

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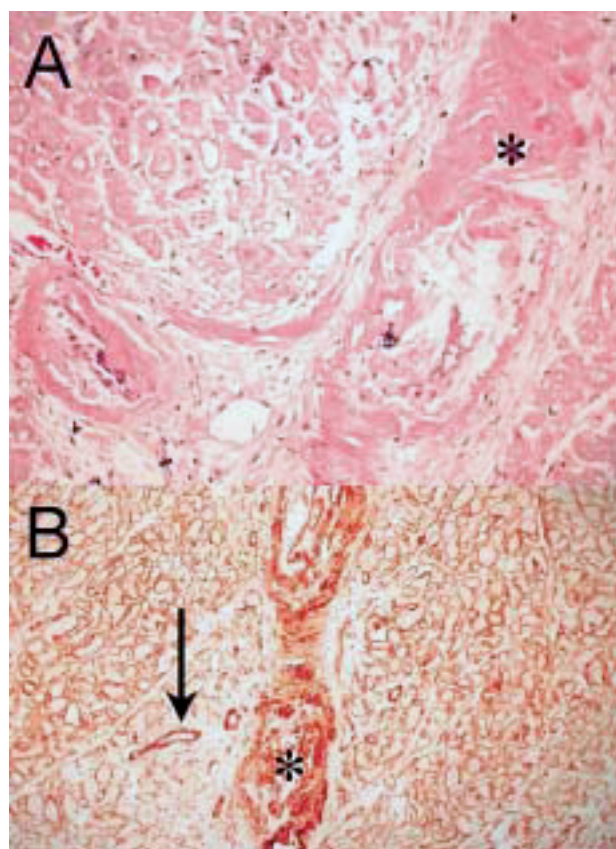
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**Fig. 1.** Transthoracic echocardiogram. A: Apical four-chamber view. Ventricular thickening with birefringence. Spontaneous echo contrast that was not seen in the static image. B: Pulsed Doppler showing mitral flow with little mechanical input from the atrium, despite sinus rhythm.

## DISCUSSION

Non-amyloidotic free light-chain deposition is a rare but underdiagnosed cause of restrictive cardiomyopathy resulting from paraproteinemia.<sup>2</sup> With specific treatment, the condition has a better prognosis than other depositional or infiltrative cardiomyopathies.<sup>3,4</sup> Cardiac problems can be the presenting symptoms of the disorder.<sup>4</sup> Thrombotic events during sinus rhythm have been described in patients with cardiac amyloidosis, but this is the first time they have been described in a patient with light-chain disease. Such events have been attributed to the physiological effects of restrictive myopathy and to atrial mechanical failure due to infiltrative disease, this being the reason anticoagulation therapy is recommended.<sup>5</sup> Clinical signs of peripheral ischemia, autopsy findings of renal infarction and atrial thrombi, and spontaneous echo contrast during sinus rhythm support the diagnosis of cardiac embolism. Light-chain cardiomyopathy should be foremost among the differential diagnoses of cardiomyopathy resulting from infiltrative or depositional processes, since treatment of the underlying blood disorder gives light-chain cardiomyopathy a better prognosis than the other



**Fig. 2.** Microphotographs (200) of myocardial samples. A: Extracellular deposit of amorphous eosinophilic material that was negative for Congo red (asterisk). B: The same specimen showing positive immunostaining (brown) for lambda light chains perivascularly (arrow) and in one extracellular deposit of amorphous material (asterisk).

conditions.<sup>3,4</sup> When performing routine tests in patients who present with these clinical symptoms, paraproteinemia must be ruled out, and possible involvement of other organs must be assessed. Light-chain-specific immunostaining should be used with all biopsy specimens. It is important to watch for the development of thromboembolism when atrial mechanical failure occurs despite normal sinus rhythm.

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