

Image in cardiology

Diagnosis of Cardiac Amyloidosis by Skin Lesions



Diagnóstico de amiloidosis cardiaca por lesiones cutáneas

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Figure 1.

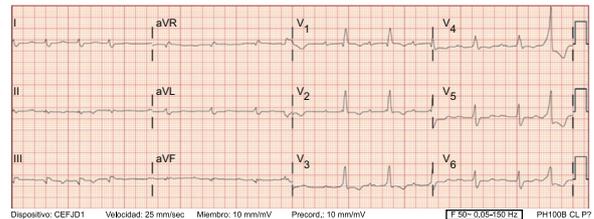


Figure 2.

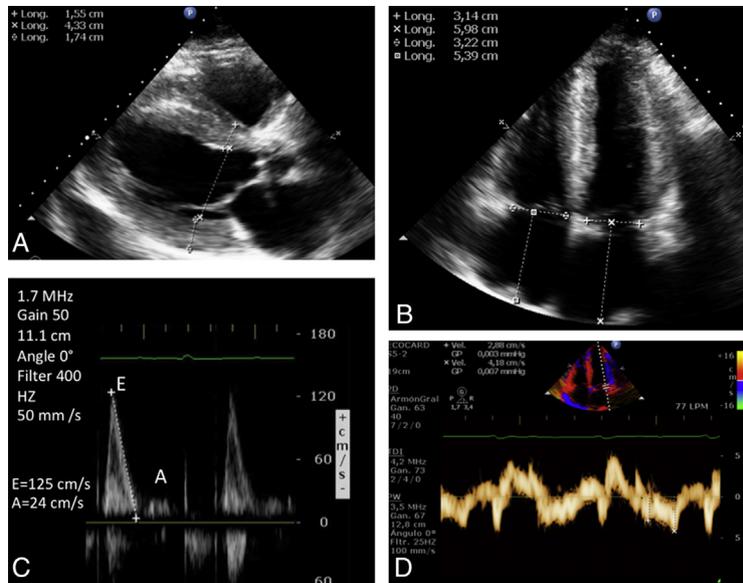


Figure 3.

In June 2011, a 74-year-old man was admitted for recurrent heart failure. He had hypertension, diabetes mellitus, renal failure due to nephrosclerosis and diabetic nephropathy, prostate cancer, tuberculosis, and gouty arthritis, and had received a parathyroidectomy. In 2009, he had received a mechanical prosthesis to treat aortic stenosis.

Cardiac auscultation was rhythmic, with no murmurs, and with normal prosthetic sounds. The patient presented petechiae, purpura, papules, and spontaneous ecchymosis at the eyelids, groin, neck, and axillas (Figure 1), with onset after initiation of acenocoumarol in 2009. The electrocardiogram demonstrated sinus rhythm with a long P-R interval, right bundle branch block with left anterior hemiblock, and low voltage, consistent with infiltrative cardiomyopathy (Figure 2). The echocardiogram demonstrated left ventricular hypertrophy with increased echogenicity (Figures 3A and B), ejection fraction of 45%, and a restrictive pattern on pulsed Doppler (Figure 3C: increased E/A ratio) and tissue Doppler (Figure 3D: decreased E' and A' peaks). Examination of skin biopsy specimens with Congo red stain showed amyloidosis secondary to monoclonal gammopathy, with positive immunohistochemistry for lambda light chains. He was treated with melphalan and dexamethasone, which were discontinued because of a favorable hematological response, with normalization of serum free light chain concentrations. Thereafter, he was hospitalized several times for heart failure and ultimately died of this cause in 2013.

The purpura, petechiae, and ecchymosis were due to intracutaneous bleeding, resulting from a fragile vascular status secondary to amyloid infiltration. This is sometimes the first and only sign of systemic amyloidosis. Although these features are not specific to this disease, they may be an aid to its diagnosis and prompt treatment in the appropriate clinical context.

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