Poster C29

Cardiac Amyloidosis Case Review

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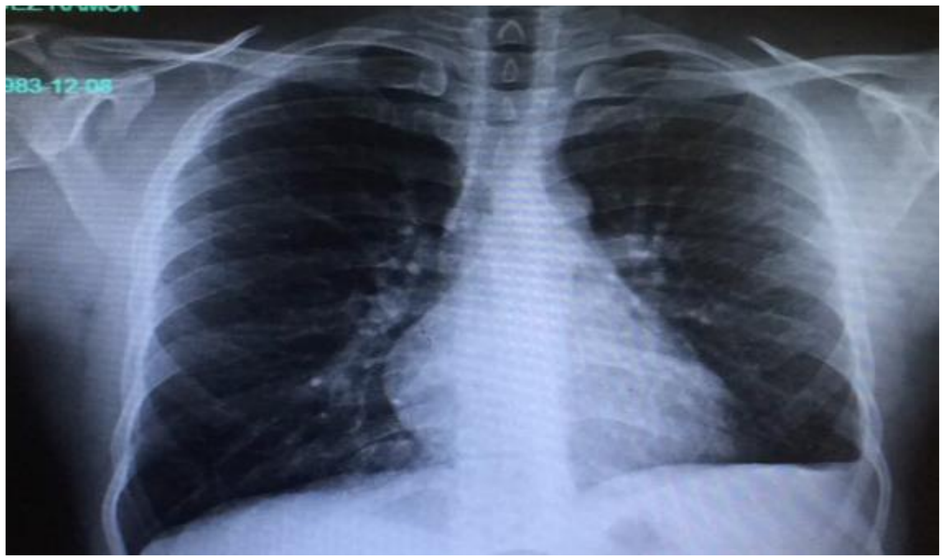
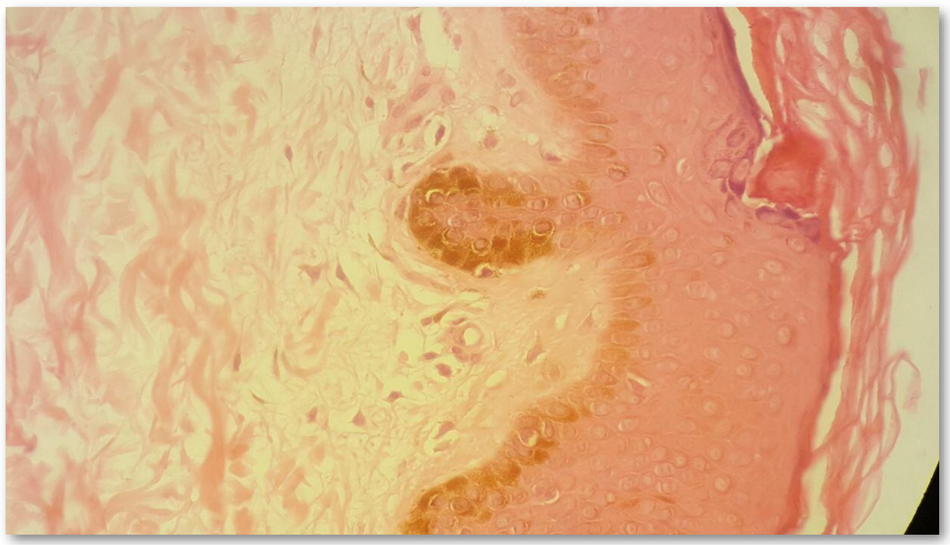
Clinical Case

A 33-year-old man with medical history of asthma arrived at the emergency room complaining of shortness of breath cough and consulted to Pneumology service. On X-ray (figure 1), left-sided pleural effusion was observed, laboratory work showed polycythemia (HgB 17.4, Hct 50.3%). Consulted with Hematology/Oncology service; bone marrow biopsy performed, myelofibrosis and hypercellularity observed. Case consulted with cardiology services due to dyspnea, left pleural effusion with EKG findings of ST-segment elevation on V1 V2. Echocardiogram showed left ventricular hypertrophy with associated hyperechogenicity and granular aspect more pronounced in the interventricular septum with ejection fraction in 67%. Cardiology services suspected infiltrative cardiomyopathy. Skin biopsy showed amyloid material (fig. 2).





Discussion

Cardiac amyloidosis is a rare form of cardiomyopathy, overall frequency in the population is not well described. Among patients with the condition, approximately 95% of cases caused by deposition of the transthyretin (ATTR amyloidosis) presents at age > 60-70, and immunoglobulin light chains (AL amyloidosis) presents at age >40 years commonly affects the liver, kidneys, autonomic and peripheral nervous system, lung as well as the heart, infiltration in the latter is present in 50-70% and is the primary determinant of prognosis. Clinical manifestations are diverse and depend on the pattern of the involved organ. Cardiac amyloidosis should be suspected in patients with unexplained left ventricular hypertrophy with or without heart failure, aortic stenosis with associated features (low flow, low gradient aortic stenosis, and echocardiogram detection of impaired longitudinal strain), and conditions highly associated with cardiac amyloidosis (systemic AL amyloidosis, ATTR-related peripheral neuropathy, or ATTR mutation carrier state). For this reason, cardiovascular magnetic resonance (CMR) imaging is the test of choice recommended. If CMR suggests the diagnosis, it is recommended to perform evidence of monoclonal protein, and if detected, hematology referral and tissue biopsy are indicated. If CMR is not diagnostic, cardiac amyloidosis is unlikely.

References

Uptodate Cardiac amyloidosis
JACC 2010;55:1769
Circ 2011;124:1079