A CHANGE OF HEART: PROGRESSIVE EDEMA IN AN ELDERLY MAN

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LEARNING OBJECTIVES

- The systemic amyloidoses are a rare group of diseases caused by diffuse extracellular deposition of proteinaceous material
- Amyloid light-chain amyloidosis is characterized by multiorgan involvement
- Cardiac involvement carries a poor prognosis

CASE PRESENTATION

A 77-year-old man with a significant cardiac history presented to his cardiologist with new-onset lower extremity edema. Despite oral diuretics, edema progressed to anasarca and the patient was admitted for IV diuretics.

ROS: edema, orthopnea, fatigue, jaw claudication, neck pain, difficulty elevating his head

PMH: CAD s/p CAGB, paroxysmal atrial dysrythmia, diastolic heart failure, hypertension, hypothyroidism, CKD, anemia

PE: JVP 9cm, irregular irregular heart rhythm, decreased breath sounds at bilateral lung bases, severe pitting edema of the lower extremities and scrotum

DIAGNOSTIC STUDIES

Laboratory:
- Cr 2.03 mg/dL
- Hgb 10.1 g/dL
- BNP 2122 pg/mL Troponin <0.01

Imaging:
- CXR: pulmonary edema, R pleural effusion
- Echo: elevated PA pressure, biatrial enlargement, LVH, E/A 1.2 (Figures 1 & 2)
- Cardiac cath: elevated RA pressure, elevated RV EDP (Figure 3)

Tissue:
- Endomyocardial biopsy: Congo Red- positive, birefringent Mass spectrometry: kappa-type amyloid
- Bone marrow biopsy: plasma cell dyscrasia

CLINICAL COURSE

Cardiac catheterization was performed which suggested restrictive cardiomyopathy. Endomyocardial biopsy stained positive for amyloid. Bone marrow biopsy suggested light-chain amyloidosis. Despite diuresis, the patient remained fluid overloaded and renal failure progressed. As light-chain subtype was not confirmed until later in clinical course, the patient was not treated for amyloidosis. While in the hospital he developed wide complex tachycardia, which led to cardiac arrest. Autopsy revealed marked cardiomegaly (720gm) with biventricular hypertrophy. Histology showed amyloid deposition in the heart, kidneys, thyroid, temporal artery, and strap muscles of the neck.

DISCUSSION

AL amyloidosis is a rare plasma cell dyscrasia in which monoclonal immunoglobulin light chains form amyloid precursors; incidence is 8.9 cases per million person-years.

Cardiac involvement is present in approximately 60% of patients and can cause ventricular hypertrophy, pericardial and pleural effusions, granular-appearing myocardium on echocardiography, and low-voltage EKGs. Once heart failure develops, median survival is less than six months. Early diagnosis and treatment is paramount for reversal of organ damage. Treatment options include melphalan, dexamethasone, stem cell transplant, and heart transplant.

Endomyocardial biopsy proved amyloidosis as the etiology for heart failure in our patient. Autopsy suggested extensive systemic involvement (Figure 4).

Amyloidosis is an important component of the differential for heart failure when the etiology is uncertain, particularly in the setting of multiorgan dysfunction.

REFERENCES


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