**Cardiac AL Amyloidosis: A Case Report**

Vinod Krishnappa1, Shilpi Shah2, Mohit Gupta3, Rupesh Raina4

1Akron Nephrology Associates/Cleveland Clinic Akron General, Akron, Ohio, USA

2Northeast Ohio Medical University, Rootstown, Ohio, USA

3Department of Internal Medicine, Cleveland Clinic Akron General, Akron, Ohio, USA

4Department of Nephrology, Cleveland Clinic Akron General and Akron Children’s Hospital, Akron, Ohio, USA

**Background:** Amyloid light chain (AL) amyloidosis is a plasma cell dyscrasia in which misfolded immunoglobulin light chains deposit in organs and cause their dysfunction. The disease most commonly affects the heart, and also demonstrates involvement of the kidneys, peripheral and autonomic nervous systems, liver, and skin.The most common extracardiac involvement is severe proteinuria in the setting of nephrotic syndrome.

**Case:** A 70-year-old man presented to the hospital with progressively worsening painless bilateral lower extremity edema for 3 weeks. On review of systems, patient endorsed some generalized weakness, but denied urinary symptoms, orthopnea, paroxysmal nocturnal dyspnea, and cough. He had been seen at an outside institution for similar symptoms a month previously, and cardiac stress test and CT chest were normal. Physical exam was unremarkable except for the bilateral 2+ pitting edema in lower extremities. Initial labs showed hemoglobin of 10 g/dl, serum creatinine 2.3 mg/dl, albumin 1.7 g/dl and total protein 4.8 g/dl. Urinalysis showed trace hemoglobin and 300 mg/d protein. 24-hour urine protein was 10.2 g. A contrast CT abdomen and pelvis showed a 5.7cm hypodense mass within the posterior right hepatic lobe extending to the capsular surface and four other small hyper-enhancing lesions in the liver. Liver lesion was suggestive of hemangioma. Alpha-fetoprotein, CA 19-9, CEA, and a hepatitis profile were negative. Urine protein electrophoresis showed glomerular proteinuria, and serum immunofixation noted a polyclonal pattern. A renal biopsy showed glomeruli with randomly oriented, non-branching fibrillar deposits in the mesangia and loops. Congo Red preparation under polarized light showed apple-green birefringence, and immunofluorescence was positive for kappa light chains, which was highly suggestive of AL type amyloidosis. The patient was discharged on valsartan for proteinuria and advised to follow up with nephrology, cardiology, hematology and oncology, and also with general surgery for the incidental cavernous hepatic hemangiomas.

**Discussion:** AL amyloid deposits in walls of heart and the inter-atrial septum causing a restrictive cardiomyopathy-like effect, though it often presents primarily as severe right-sided heart failure**.** The median survival of patients with cardiac AL amyloidosis is 6 months from the onset of heart failure. Early diagnosis is unfortunately rare, as most clinicians have low suspicion for it**.** When AL amyloidosis is suspected, a histological diagnosis is essential which shows apple-green birefringence on Congo Red staining under polarized light. Serum-free light chain measurements, and serum and urine immunoelectrophoresis should be done. Oftentimes bone marrow biopsies required verifying plasma cell dyscrasia**.**

The management of AL amyloidosis with cardiac involvement includes symptomatic treatment of the heart failure and treatment of the underlying plasma cell dyscrasia. Diuretics and salt restriction are the best options for treatment. High dose chemotherapy with or without autologous stem cell transplantation can help stop or reduce production of amyloidogenic monoclonal Ig light chains and improves survival**.** Chemotherapy with melphalan with stem cell transplantation has been shown to result in a complete hematologic response**.** Despite hematologic response, patients with advanced cardiac amyloidosis often do not survive**.** Resolution of nephrotic syndrome is associated with the disappearance of light chains, though persistent amyloid deposition is often seen on repeated renal biopsies**.** Our patient has very poor prognosis due to his late presentation and heart failure symptoms.