

Multisystem dysfunction in advanced AL amyloidosis: A case report

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Abstract

Introduction: Systemic light-chain (AL) amyloidosis is a rare plasma cell dyscrasia characterized by the deposition of misfolded immunoglobulin light chains in multiple organ systems. Due to its nonspecific presentation, it is frequently under-recognized until late stages. Classically, AL amyloidosis presents in the sixth to seventh decade of life, and is more common in men. We present the unusual case of a 56-year-old woman presenting with cachexia, sepsis, recurrent pericardiac arrests, and multisystem decline who was ultimately diagnosed with advanced systemic AL amyloidosis.

Methods: This retrospective chart review presents a thorough clinical evaluation of a patient presenting with *Klebsiella pneumoniae* bacteremia on a background of progressive weight loss and functional decline. Diagnostic investigations included blood and urine cultures, computed tomography, echocardiography, cardiac MRI, and tissue biopsies. Congo red staining of fat pad and bone marrow samples, along with demonstration of clonal plasma cells, established the diagnosis of systemic AL amyloidosis with multi-organ involvement.

Results: A 56-year-old woman presented to the emergency department with abdominal pain, urinary frequency, severe watery diarrhea, hypotension, and hypothermia on a background of progressive weakness and 3-year history of weight loss of >50% from baseline. Over the month prior, she had declined from functionally independent to bed-bound. She was treated in ED for pyelonephritis, and subsequently admitted for management by the internal medicine team, where blood cultures showed *Klebsiella* bacteremia. Shortly after admission, she developed severe back pain, paresthesias, and intermittent slurring of speech. She was admitted to ICU with metabolic acidosis secondary to acute kidney injury (AKI) and sepsis. She subsequently developed bradycardia and experienced several episodes of cardiac arrest and resuscitation. CT abdomen revealed hepatomegaly, anasarca, and bilateral renal hypodensities. Transthoracic echocardiography revealed left ventricular hypertrophy with an apical sparing pattern, and cardiac MRI revealed a diffuse gadolinium enhancement. MRI brain showed pachymeningeal enhancement. Abdominal fat pad biopsy was Congo red positive, and bone marrow biopsy demonstrated 20% lambda-restricted plasma cells, confirming systemic AL amyloidosis with cardiac, renal, gastrointestinal, and autonomic involvement. Given her instability, she was not a candidate for chemotherapy, and goals of care were transitioned to DNR/DNI with palliative management. Prognosis was estimated to be less than 3–6 months.

Conclusion: This case highlights the diagnostic challenges and poor prognosis of advanced systemic AL amyloidosis. Multisystem involvement should raise suspicion for amyloidosis in patients with unexplained cachexia, cardiac dysfunction, renal impairment, and neuropathy. To ensure positive patient outcomes, early recognition and tissue diagnosis are critical, as therapeutic options are limited once hemodynamic instability and multiorgan failure occur.