A CASE REPORT ON FREE LIGHT CHAIN AMYLOIDOSIS WITH CARDIAC AND EXTRACARDIAC MANIFESTATIONS AFTER YEARS OF DIAGNOSTIC DELAY

Manal Atef, MD
Dar El-Salam Cancer Center, , Cairo, Egypt



Introduction

- Free light chain (FLC) amyloidosis results from misfolded light chain fragments produced by a clonal population of bone marrow plasma cells. These fibrils infiltrate various organs, causing symptoms that vary based on the type and degree of organ involvement.

Objective

- To present a case of a rare disease with subtle, nonspecific symptoms often leading to misdiagnosis and treatment delays, resulting in progressive organ dysfunction and failure.

Patient

- A 70-year-old male patient, without diabetes or hypertension, was diagnosed with systemic λ light chain (AL) amyloidosis in November 2023. He had presented 2 years earlier with profound fatigue, unexplained weight loss, lower limb numbness and tingling, multiple hypotensive episodes, syncopal attacks, and watery diarrhea. The patient sought medical advice and was prescribed painkillers for peripheral neuropathy. A colonoscopy revealed nonspecific inflammation. By February 2023, the patient complained of dyspnea on exertion and lower extremity edema. An electrocardiogram revealed a premature ventricular contraction burden ≥10% and right bundle branch block. .

Patient

.The echocardiogram showed mild left ventricular hypertrophy with preserved systolic function. Troponin I and N-terminal pro-B-type natriuretic peptide (NT-ProBNP) levels were elevated (0.04 ng/mL and 2,030 pg/mL, respectively). The patient underwent cardiac catheterization and stent placement to open a blocked left anterior descending artery.

Interventions

-Hemoglobin:10g/dL ,Serum protein electrophoresis (PEP): hypoalbuminemia ,hypogammaglobulinemia with a monoclonal band in the gamma region, and abnormal α_2 region
,Proteinuria (3.5 g/24 h)
,Elevated λ FLC: 122 mg/L
,Abnormal κ:λ ratio: 0.23
,Difference in FLC (dFLC): 85 mg/dL
Low plasma cell frequency in bone marrow studies. -AL amyloidosis was confirmed by renal biopsy. No bone lesions were detected. Hepatomegaly was present, and alkaline phosphatase was 59 U/L. -The patient was deemed eligible for treatment with daratumumab-CyBORD (cyclophosphamide, bortezomib, dexamethasone).

Main outcomes

- After six cycles of therapy, the patient achieved a complete hematologic response, with improvement of all previous symptoms. The patient is currently receiving monthly daratumumab as a maintenance therapy.

Conclusion

Early recognition of AL amyloidosis reduces organ dysfunction and improves quality of life. Recognition of cardiac and extracardiac red flags is crucial for raising clinical suspicion of AL amyloidosis.

dr.manalatif@hotmail.com , +20- 1005644494

SOHO2025