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Complex Clinical Presentation of Late-Stage AL Amyloidosis: A Case Report

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INTRODUCTION

AL amyloidosis is a rare hematological disorder characterized by the deposition of amyloid fibrils in various organs, leading to multi-system dysfunction. Here, we present the case of a 61-year-old patient who exhibited a complex clinical presentation of AL amyloidosis, predominantly affecting the heart, gastrointestinal tract (GI), and nervous system.

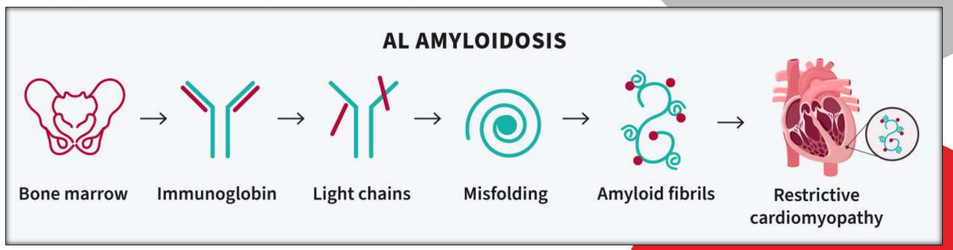
AIM

The aim of this case report is to present a case of AL amyloidosis that was successfully treated despite the challenges posed by its complicated manifestations and delayed diagnosis.

CASE REPORT

In October 2022, patient with hypercreatininemia (195 µmol/l) presented to nephrology department. Complains occurred from February 2021 and included fatigue, hypotension, dizziness, dysphagia, nausea, insomnia, and weight loss. Observation revealed diffuse macroglossia, leading to nocturnal apnea, impaired swallowing function, and malocclusion. Imaging investigations, namely CT scan and ultrasound, resulted: right kidney 8.2 x 4.2 cm, left kidney 10.7 x 4.3 cm with weak corticomedullary differentiation, hepatomegaly, and moderate pleural effusion. The patient was diagnosed with CKD, and was under observation for a year. Since May 2022, the patient's condition was worsening: lower limbs swelling, hypotension (90/60 mm Hg), and syncope. Newer imaging investigations: normal CT scan of the brain, reflux gastritis with GI barium contrast X-ray; left ventricular hypertrophy, reduced left ventricular cavity, type I diastolic dysfunction, IVS measuring 12.8 cm, EF between 45-50%, and slight free fluid in the pericardial cavity by Echocardiography; pathological Q waves in leads III and aVF, negative T waves in leads V4, V5, and V6, and decreased amplitude of the QRS complex by ECG. Hypertrophic cardiomyopathy was diagnosed, but despite treatment, the patient's condition continued to deteriorate progressively. Over the past year, the patient lost 15 kg. In August, experiencing GI complains, the patient was diagnosed with atrophic gastritis, erosive bulbitis, and a colon polyp. Histological examination of the GI biopsy material revealed amyloid deposition. Further immunohistochemistry revealed AL/λ amyloidosis, which was confirmed with additional analyses during hematology consultation. The patient received treatment with Daratumumab-VCD (bortezomib, cyclophosphamide and dexamethasone), and after 6 cycles had partial response. Maintenance with daratumumab was also completed.

Picture 1. AL Alyloidosis related cardiomyopathy (sounrced from: https://www.ottawaheart.ca/heart-condition/cardiac-amyloidosis)



CONCLUSIONS

This case underscores the importance of early recognition, comprehensive diagnostic evaluation, and multidisciplinary management in optimizing outcomes for patients with late-stage AL amyloidosis.

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