ejpmr, 2025, 12(1), 458-460

SJIF Impact Factor 7.065

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

A RARE CASE OF YOUNG PATIENT WITH REFRACTORY HYPOTENSION WITH NEPHROTIC RANGE PROTEINURIA DIAGNOSED TO BE AMYLOIDOSIS

***[1]Dr. Anand Prasad, [2]Dr. Navya Jaiswal, [3]Dr. Apoorv Sharma, [4]Dr. Srikanth Sakhamuri**

[1]Assistant Professor of Nephrology, Swami Vivekananda Subharti University, Subharti Medical College. MBBS, MD, Dr.DNB Nephrology, MRCPUK (LONDON).

[2]Associate Professor Pathology, Subhart Medical College.

[3,4]Junior Resident Department of Medicine Subhart Medical College.

***Corresponding Author: Dr. Anand Prasad**

Assistant Professor of Nephrology, Swami Vivekananda Subharti University, Subharti Medical College. MBBS, MD, Dr.DNB Nephrology, MRCPUK (LONDON).

Article Received on 29/11/2024 **Article Revised on 19/12/2024** **Article Accepted on 09/01/2025**

ABSTRACT

This case report describes a patient presenting with facial puffiness, bilateral pitting edema, dizziness, Refractory hypotension and postural hypotension. Initial evaluation revealed dyslipidemia, hypothyroidism, and nephrotic range proteinuria. Further investigations, including fat biopsy and renal biopsy, confirmed the diagnosis of amyloidosis. Treatment with midodrine was initiated for postural hypotension. This case highlights the importance of considering amyloidosis in patients with nephrotic range proteinuria, Refractory hypotension and postural hypotension.

KEYWORD:- Amyloidosis, nephrotic syndrome, Refractory hypotension, postural hypotension, proteinuria, renal biopsy.

INTRODUCTION

Amyloidosis is a rare but serious disorder characterized by the extracellular deposition of insoluble amyloid fibrils in various tissues and organs. This abnormal protein accumulation can lead to significant dysfunction in affected organs. Among the various types of amyloidosis, the most common forms include AL (primary) amyloidosis, associated with plasma cell dyscrasias, and AA (Secondary) amyloidosis, often related to chronic inflammatory conditions.[3]

Renal involvement is particularly common in amyloidosis, frequently presenting as nephrotic syndrome, which is characterized by proteinuria, hypoalbuminemia, and edema.[8] The kidneys are often affected due to their high blood flow and the unique characteristics of the glomerular capillary wall, which makes them a prime target for amyloid deposition.[1] This renal manifestation significantly impacts patient morbidity and mortality, as progressive kidney damage can lead to chronic kidney disease and end-stage renal disease.[7]

While nephrotic syndrome is a well-recognized complication of amyloidosis, less common manifestations such as postural hypotensionand refractory hypotension can also occur. Postural hypotension, or orthostatic hypotension, is defined as a significant drop in blood pressure upon standing and can lead to dizziness, fainting, and falls.[4]

This symptom arises due to autonomic nervous system involvement and is frequently under-recognized in the context of amyloidosis.[2] The deposition of amyloid in the autonomic nerves leads to autonomic dysfunction, which can manifest as gastrointestinal motility disturbances, urinary retention, and Refractory hypotension.[5][7]

Early recognition of these diverse manifestations and appropriate management are crucial for improving outcomes in patients with amyloidosis. Diagnostic delays are common due to the protean nature of the disease and the nonspecific symptoms that often accompany it.[6] This case report highlights the occurrence of Refractory hypotension alongside nephrotic syndrome in a patient with amyloidosis, emphasizing the importance of comprehensive clinical evaluation and a high index of suspicion in these patients.

CASE PRESENTATION

A 35-year-old female presented to opd with complain of facial puffiness, bilateral pitting edema, and dizziness, She also denied of any history of smoking or alcohol/drug abuse and was not on any medications. On examination, She had a blood pressure of 80/60 mmHg

for which inotropoc support was started (noradrenaline @5mcdrops/min). Laboratory investigations revealed dyslipidemia, hypothyroidism on thyroxine supplementation, urine albumin-to-creatinine ratio >2gm, and 24-hour urinary protein excretion of 7.2gm. Patient was having refractory hypotension as blood pressure is not raised on inotrope support and nor there is an fall of blood pressure on stopping inotropic support. So, patient was suspected to be a case of adrenal insufficiency and early morning serum cortisol was done which came out to be normal. Serum ACTH was also done which was unremarkable. Patient was started on steroids but even after that blood pressure is on the lower side. So, further research was done in search of cases including hypotension with Nephrotic range protienuria and the following differential diagnosis was suspected and workup was done. Further workup for nephrotic range proteinuria included differential diagnosis of focal segemental glomerulo nephritis, minimal change disease, membranous nephropathy, HIV, hepatitis B and C, SLE, collagen vascular disease DKD and amyloidosis. Then patient was counselled for abdominal fat biopsy which revealed the patient to be a case of amyloidosis.

Diagnostic assessment

Fat biopsy revealed Congo red stain positivity with apple green birefringence on biopsy demonstrated amyloidosis with negative kappa lambda and serum amyloid A (SAA) but positive ATTR (transthyretin) (Figure 1). Additionally, A 12-lead electrocardiogram (Figure 2) revealed left ventricular hypertophy, A transthoracic echocardiogram (Figure 3) revealed moderate concentric left ventricular hypertrophy, Ejection fraction exhibited significant beat-to-beat variation between 50% and 55%. indicative of cardiac involvement in amyloidosis.

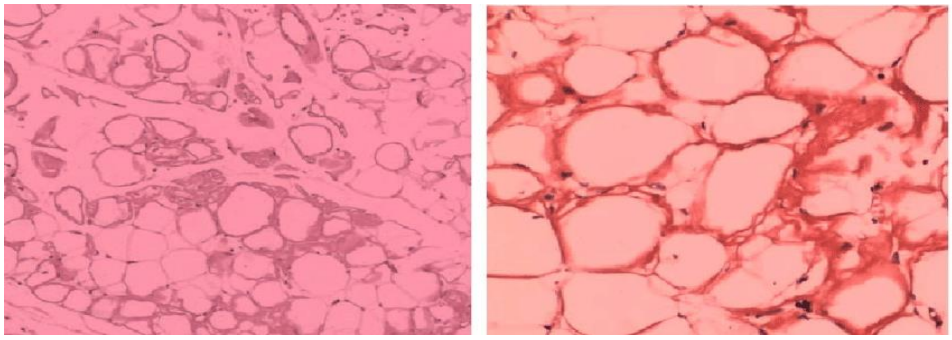

Figure 1

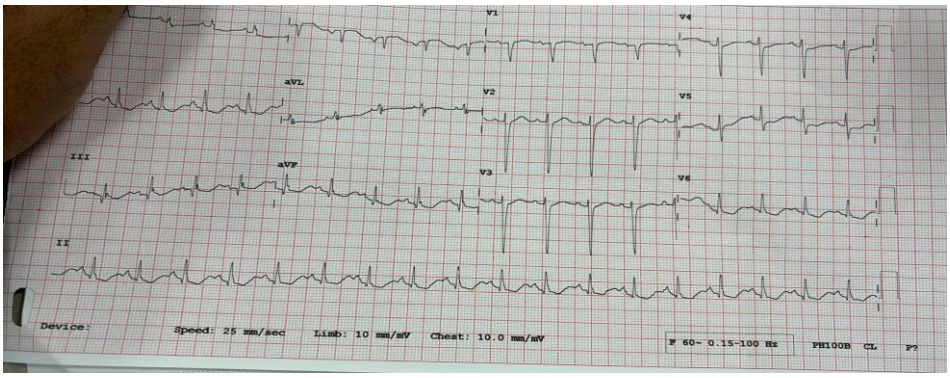

Figure 2

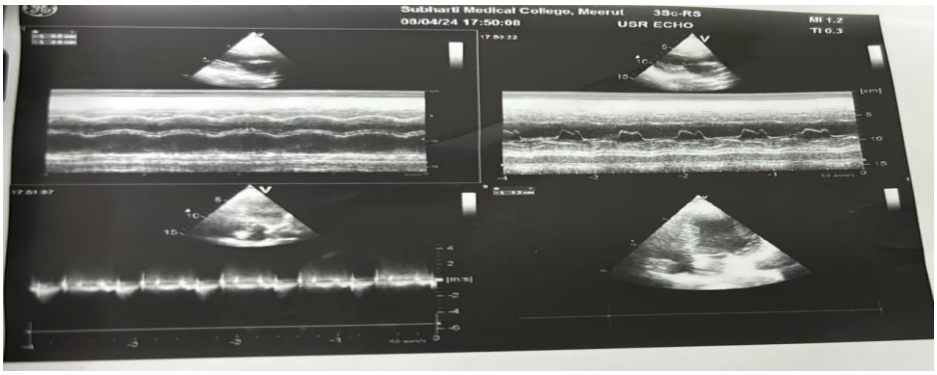

Figure 3

Treatment and Outcome

The patient was managed with Inotropic support and steroids and midodrine 10 mg twice daily, followed by 10 mg three times daily for postural hypotension and Tab eltroxin 50 mcg once a day and Inj h albumin 20%100nl intravenous for 3 days.

Patient was discharged on steroid support and midodrine, Eltroxin and adviced for high protien diet with Regular follow-up visits were scheduled for monitoring of renal and cardiac function.

DISCUSSION

This case underscores the diagnostic challenge and clinical manifestations of amyloidosis, especially in young patients presenting with nephrotic range proteinuria and Refractory hypotension. Early diagnosis and appropriate management are crucial for preventing further organ damage and improving patient outcomes.

CONCLUSION

Amyloidosis should be considered in the differential diagnosis of patients with nephrotic range proteinuria and Refractory hypotension. Timely diagnosis and management are essential for optimizing patient care and outcomes. Atypical type of amyloidosis can present in early part of life and to be diagnosed with panel of investigations

REFERENCES

1. Dember, L. M. Amyloidosis-associated kidney disease. Journal of the American Society of Nephrology, 2006; 17(12): 3458-3471.
2. Falk, R. H., Comenzo, R. L., & Skinner, M. The systemic amyloidoses. New England Journal of Medicine, 1997; 337(13): 898-909.
3. Gertz, M. A., Dispenzieri, A., & Sher, T. Pathogenesis, diagnosis, and treatment of immunoglobulin light-chain amyloidosis. Mayo Clinic Proceedings, 2004; 79(4): 593-602.
4. Isfort, R. W., Cody, R. J., & Tzamaloukas, A. H. Orthostatic hypotension: Pathophysiology, evaluation, and treatment. International Urology and Nephrology, 2016; 48(8): 1115-1125.
5. Kourelis, T. V., & Gertz, M. A. Immunoglobulin light chain amyloidosis. Hemato, 2017; 2(1): 1-13.
6. Merlini, G., Bellotti, V., & Obici, L. Amyloidosis and protein folding disorders: Aetiology and pathogenesis. British Journal of Haematology, 2013; 161(1): 3-17.
7. Sanchorawala, V. Light-chain (AL) amyloidosis: Diagnosis and treatment. Clinical Journal of the American Society of Nephrology, 2006; 1(6): 1331-1341.
8. Sethi, S., Vrana, J. A., & Theis, J. D. Laser microdissection and mass spectrometry-based amyloid proteomics: Approach to pathology diagnosis and amyloid typing. Journal of the American Society of Nephrology, 2012; 23(9): 1601-1605.