Medical Journal of Bakırköy

Med J Bakirkoy 2025;21(3):339-342
DOI: 10.4274/BMJ.galenos.2025.2025.1-2

Case Report

Hypereosinophilic Syndrome Masquerading as a Dialyzer Reaction: A Great Mimicker

Diyalizör Reaksiyonu Gibi Görünen Hipereozinofilik Sendrom: Harika Bir Taklitçi

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ABSTRACT

Hypereosinophilic syndrome is a disorder characterized by the persistent marked elevation of eosinophils, which eventually leads to the inflammatory mediator release resulting in eosinophilic infiltrations in target organs. The occurrence of hypereosinophilic syndrome among end stage renal disease (ESRD) patients is rarely documented. As hypereosinophilic syndrome most commonly presents as intradialytic hypotension, it may mimic dialyzer reactions among ESRD patients with regular haemodialysis; hence, making the diagnosis and initiation of treatment challenging for attending physicians. We report a case of hypereosinophilic syndrome that presented with recurrent episodes of intradialytic hypotension mimicking a dialyzer reaction.

Keywords: Dialyzer reaction, ESRD, haemodialysis, hypereosinophilic syndrome, mimicker

ÖZ

Hipereozinofilik sendrom, eozinofillerin kalıcı belirgin yükselmesiyle karakterize bir hastalıktır ve bu da sonunda hedef organlarda eozinofilik enfiltrasyonlarla sonuçlanan enflamatuvar mediatör salınımına yol açar. Son dönem böbrek hastalığı (SDBH) hastalarında hipereozinofilik sendromun ortaya çıkması nadiren belgelenir ve raporlanır. Hipereozinofilik sendrom en sık intradiyalitik hipotansiyon olarak ortaya çıktığından, düzenli hemodiyaliz uygulanan SDBH hastalarında diyalizör reaksiyonlarını taklit edebilir ve bu nedenle tanıyı ve tedavinin başlatılmasını ilgili hekimler için zorlaştırır. Diyalizör reaksiyonunu taklit eden tekrarlayan intradiyalitik hipotansiyon ataklarıyla ortaya çıkan bir hipereozinofilik sendrom olgusunu bildiriyoruz.

Anahtar Kelimeler: Diyalizör reaksiyonu, SDBH, hemodiyaliz, hipereozinofilik sendrom, taklitçi

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Cite as: Hashim Y, Hashim SNH, Lim KY, Kazmin NEM, Shamsi MYA, Kamaruzaman L, et al. Hypereosinophilic syndrome masquerading as a dialyzer reaction: a great mimicker. Med J Bakirkoy. 2025;21(3):339-342

Received: 03.01.2025
Accepted: 28.04.2025
Publication Date: 03.09.2025

INTRODUCTION

Hypereosinophilic syndrome is a rare disorder characterized by hypereosinophilia with absolute eosinophil count of more than 1.5x10^9/L (1). It is an umbrella term used to describe diseases leading to peripheral blood eosinophilia, the causes of which include eosinophilic neoplasm, drug hypersensitivity reactions, helminth infections, and idiopathic causes (2). Hypereosinophilic syndrome has a wide range of manifestations, from non-specific subtle pruritus, rashes, and flushing, to life-threatening angioedema and shortness of breath, and may lead to multiorgan failure due to eosinophilic infiltrations (3). This can mimic intradialytic hypotension due to dialyzer reactions during haemodialysis in end stage renal disease (ESRD) patients (4).

CASE REPORT

A 51-year-old woman presented with hypotension ten minutes into haemodialysis, preceded by a week-long duration of generalized pruritus and skin rash. She was a known case of ESRD secondary to diabetic kidney disease, who had been on long-term regular haemodialysis for a month, using a polyethersulfone dialyzer. She had no known food or drug allergies and there was no previous episode of anaphylaxis. Her physical examination revealed a generalized pruritic macular erythematous skin rash. Her blood pressure was 138/80 mmHg, and her pulse rate was 98 beats per minute. Other systemic examinations were unremarkable. Her initial blood investigations revealed a significantly elevated eosinophil count of 18.4x10^9/L and a deranged renal profile that is consistent with ESRD. Other blood investigations, including liver function tests, were unremarkable.

She was admitted and treated for acute endogenous eczema with topical steroids, resulting in a significant resolution of her skin rashes. However, she had recurrent episodes of intradialytic hypotension with severe nausea and hot flushes after five-to-ten minutes into each haemodialysis session, with a persistently elevated eosinophil count ranging from 14.1 to 18.4x10^9/L (Figure 1). Further investigations to look for secondary causes of hypereosinophilia, including stool analysis for parasites and peripheral blood film, were unremarkable.

Despite using gentler modalities of haemodialysis including sustained low efficacy dialysis and continuous veno-venous haemodialysis, and various types of dialyzers including a polysulfone membrane (FX 1.4), polyethersulfone and polyvinylpyrrolidone blend (Theranova 400) and copolymer of acrylonitrile and sodium methallyl sulfonate (AN69), she persistently experienced hypotension, severe nausea, and hot flushes five-to-ten minutes into each haemodialysis session. Eventually, she was treated with oral prednisolone at a dose of 1 mg/kg for idiopathic hypereosinophilic syndrome, which drastically reduced her eosinophil count to a normal range. A trial of haemodialysis with a polyethersulfone dialyzer, conducted two days later, was uneventful. She was discharged with a tapering dose of prednisolone, and was able to tolerate the subsequent haemodialysis sessions without hypotension and rebound eosinophilia.

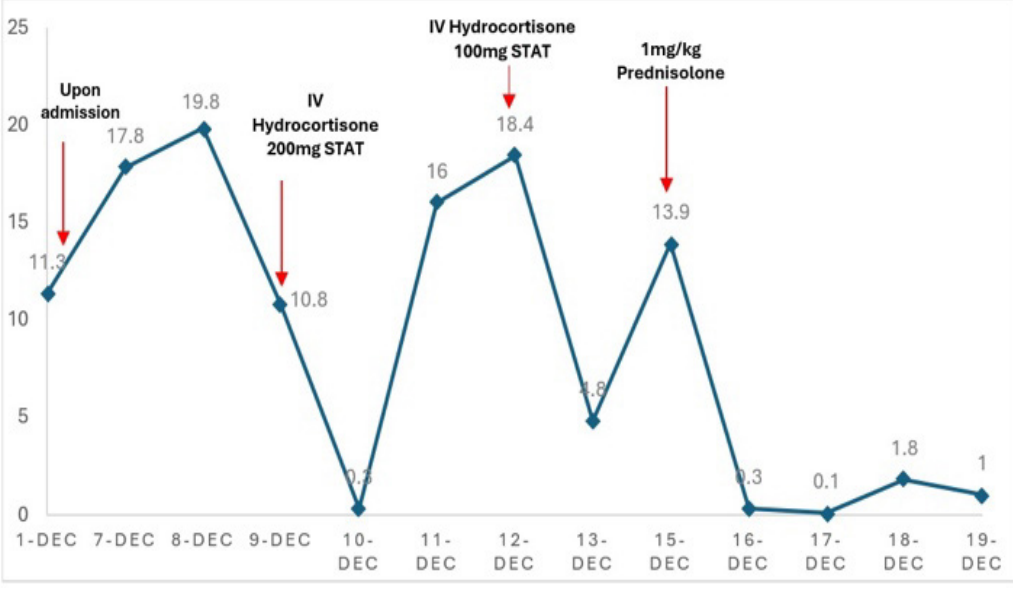

Figure 1. Eosinophil count trend throughout admission

She has given consent for this case to be reported and published.

DISCUSSION

Hypereosinophilia is a relatively common finding in daily clinical practice, and it is frequently associated with constellations of signs and symptoms that provide valuable clues for establishing the diagnosis. Hypereosinophilia is not a rare occurrence among ESRD patients receiving haemodialysis due to the reactions towards the haemodialysis circuit. It is often associated with the type of dialyzer used. With the emergence of modern membranes, hypereosinophilia among ESRD patients has become less common (5).

The cause of recurrent hypotension and gastrointestinal symptoms immediately after the initiation of dialysis in our case had been initially postulated to be due to hypersensitivity reactions to the dialyzer. Dialyzer reactions refer to every abnormal sequela following the initiation of haemodialysis and are a result of the interactions between blood and its constituents with the dialyzer membrane. Dialyzer reaction can be divided into two types: type A reaction usually occurs immediately after the commencement of dialysis and attributed to preformed antibodies against an allergen leading to immunoglobulin E (IgE)-mediated reactions, whereas type B reaction is delayed and non-IgE-mediated, resulting in milder and self-limiting symptoms (6,7).

The most common dialysis membrane reactions occur with ethylene oxide membranes and less commonly with biocompatible membranes. The relative risk of hypersensitivity reaction with synthetic membranes is 10-20% higher than that of cellulose membranes (8). Our case had been using a polyethylsulfone membrane for haemodialysis and she developed immediate hypersensitivity reactions five to ten minutes into haemodialysis. However, despite changes in dialyzer, her symptoms did not improve. Due to the persistent elevation of eosinophils, the diagnosis of hypereosinophilic syndrome was made in the absence of secondary causes.

Among ESRD patients, the common manifestations of hypereosinophilic syndrome include intradialytic hypotension with myriad other target organ damage such as chest pain, abdominal cramps, and generalized pruritus, which can be treated with systemic corticosteroids (9). Systemic corticosteroids are the gold standard treatment for hypereosinophilic syndrome with resolution of symptoms and normalization of blood parameters in 85% of cases, but the exact dosing and duration of corticosteroids remain controversial (2).

Although corticosteroids remain the first-line treatment for the management of idiopathic hypereosinophilic syndrome for induction and maintenance during acute settings, there are certain groups of patients who cannot tolerate high-dose corticosteroids, and will require initiation of a second-line agent. The use of hydroxyurea, methotrexate, cyclosporine, and pegylated interferon alpha has been reported in the successful treatment of idiopathic hypereosinophilic syndrome, even though the clinical evidence is scarce (10).

CONCLUSION

Persistent hypereosinophilia in the absence of any secondary causes should prompt the suspicion of idiopathic hypereosinophilic syndrome. Hypereosinophilic syndrome in ESRD patients, with regular haemodialysis, may complicate diagnosis by mimicking haemodialysis intolerance due to dialyzer reactions.

ETHICS

Informed Consent: She has given consent for this case to be reported and published.

FOOTNOTES

Authorship Contributions

Surgical and Medical Practices: Y.H., S.N.H.H., K.Y.L., N.E.M.K., M.Y.A.S., L.K., R.M., Consept: Y.H., S.N.H.H., K.Y.L., M.Y.A.S., Design: Y.H., S.N.H.H., K.Y.L., N.E.M.K., L.K., R.M., Data Collection or Processing: Y.H., K.Y.L., N.E.M.K., Analysis or Interpretation: Y.H., K.Y.L., N.E.M.K., M.Y.A.S., L.K., R.M., Literature Search: Y.H., S.N.H.H., K.Y.L., Writing: Y.H., S.N.H.H., K.Y.L.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declare that this study received no financial support.

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