Case Report

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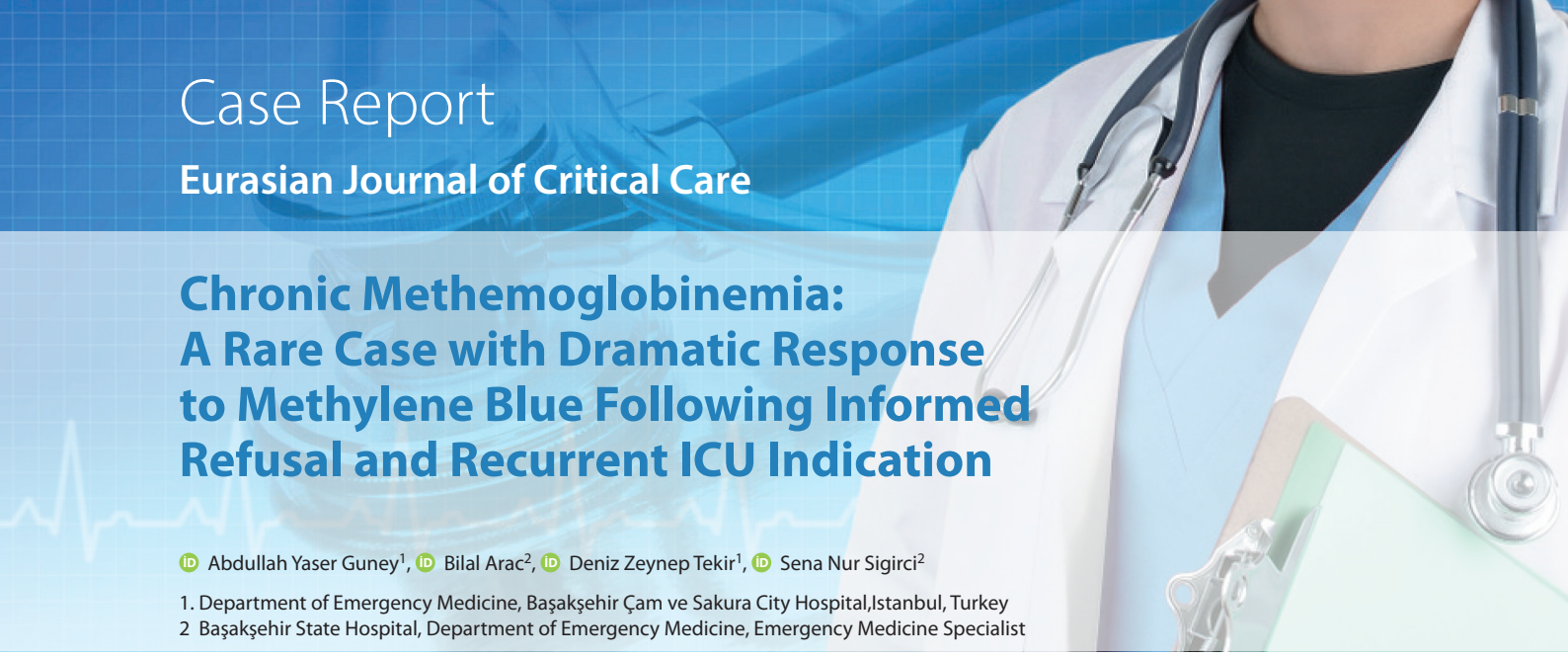

Chronic Methemoglobinemia: A Rare Case with Dramatic Response to Methylene Blue Following Informed Refusal and Recurrent ICU Indication

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Abstract

A 46-year-old female patient with a lifelong history of cyanosis affecting the lips and fingertips since childhood presented to the emergency department. On admission, she exhibited mild dyspnea and low oxygen saturation (84%). Arterial blood gas analysis revealed a pH of 7.4, pCO_2 of 40 mmHg, HCO_3 of 24 mmol/L, hemoglobin level of 17 g/dL, and a methemoglobin (MetHb) level of 38%. The patient initially refused referral and left the hospital, but returned two days later with recurrent symptoms. Intravenous methylene blue was administered at a dose of 1–2 mg/kg. Following treatment, the MetHb level decreased to 3%, and the patient's symptoms resolved. This case highlights that acute episodes in patients with chronic methemoglobinemia can be effectively managed with early diagnosis and timely intervention.

Introduction

Methemoglobinemia is a rare hematologic disorder characterized by the oxidation of hemoglobin's iron from the ferrous (Fe^{2+}) to the ferric (Fe^{3+}) state due to oxidative agents, impairing its oxygen-carrying capacity. Under normal physiological conditions, methemoglobin levels in the blood remain between 1–2% (1). Clinically, methemoglobinemia presents with decreased oxygen saturation and central cyanosis that does not respond to oxygen therapy. The diagnosis is confirmed by arterial blood gas analysis, and treatment involves the use of reducing agents such as methylene blue (2).

Case Report

A 46-year-old female patient presented to the emergency department with complaints of cyanosis of the lips and fingertips. The patient reported a history of intermittent cyanosis of the lips and fingers since childhood. Physical examination revealed no pathological findings in either lung, with equal bilateral respiratory movements. Respiratory rate was 15 breaths/min, blood pressure 135/65 mmHg, heart rate 90 bpm, and peripheral oxygen saturation (SpO_2) was 84%. The patient was neither tachypneic nor dyspneic. She denied any recent medication use, but reported frequent consumption of beetroot in Konya, raising the possibility of a dietary etiology. Referral to a tertiary care center for further evaluation and treatment was requested via the national referral system. However, the patient declined the referral, stating that her symptoms were not new and had occurred intermittently since childhood. After signing an informed refusal form, she left the hospital voluntarily.

Two days later, the patient presented to a tertiary emergency department as an outpatient with similar complaints. Blood gas analysis revealed results nearly identical to the previous visit. Due to the indication for intensive care, intravenous methylene blue therapy was initiated in the emergency department.Venous blood gas analysis revealed the following values: pH 7.394, pCO_2 40.7 mmHg, pO_2 37.6 mmHg, HCO_3^- 23.9 mmol/L, lactate 1.8 mmol/L, and methemoglobin (MetHb) level of 38%, confirming the diagnosis of methemoglobinemia.Follow-up

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blood gas analysis performed two hours after treatment showed stable values: pH 7.394, pCO_2 40.7 mmHg, pO_2 37.6 mmHg, HCO_3^- 23.9 mmol/L, lactate 1.8 mmol/L, and a significantly decreased MetHb level of 3%.

Discussion

In this case, the patient was found to have chronic methemoglobinemia resulting from a clinical condition to which she had grown accustomed. Chronic methemoglobinemia is typically congenital in origin and may be triggered by specific stressors or environmental factors. The condition often resolves spontaneously. We believe that the patient's refusal to be transferred from the initial facility was primarily due to her previous experiences with similar episodes that had resolved without treatment. In this case, no clear trigger for the methemoglobinemia episode was identified; however, it was noted that the patient consumed large quantities of beetroot in her hometown, which may have played a contributing role. No medication-related cause was found.In a review published by Hord et al. in 2009, the biological transformations of dietary nitrates and their potential health effects were comprehensively discussed. It was emphasized that excessive consumption of high-nitrate foods (e.g., beets) could increase the risk of methemoglobinemia under certain conditions [3].The treatment of methemoglobinemia varies depending on the patient's clinical status and the level of methemoglobin. In mild cases, withdrawal of oxidative agents and supportive care may be sufficient. However, in cases where methemoglobin levels exceed 20% or severe symptoms are present, specific treatment becomes necessary [4].

Conclusion: This case illustrates that the diagnosis of chronic methemoglobinemia may be delayed due to stable vital signs and the presence of long standing symptoms. Prompt recognition and administration of methylene blue led to rapid symptom resolution in this patient, emphasizing the importance of a multidisciplinary approach in such rare presentations.

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