

DAPSONE INDUCED METHEMOGLOBINEMIA IN A PATIENT WITH JUVENILE DERMATOMYOSITIS

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BACKGROUND

Methemoglobin is a hemoglobin that has been oxidized by changing its configuration of heme iron from the ferrous (Fe^{2+}) to the ferric (Fe^{3+}) state, leading to hypoxia in two ways. First, the oxygen does not bind to methemoglobin ferric hemes. Second, the presence of methemoglobin promotes a shift to the left side of the oxygen dissociation curve and as a consequence it reduces the delivery of oxygen to the tissues. Dapsone is a drug that is used in the treatment of skin manifestations of dermatomyositis. Long-term administration of dapsone at standard doses (100 mg/day) results in the methemoglobinemia in about 15% of patients.

CASE REPORT

A 23-year-old female patient, diagnosed with juvenile dermatomyositis, evolved with heliotrope, Gottron papules, proximal muscle weakness, increased muscle enzymes and, electroneuromyography with myopathic pattern. At the time of the diagnosis, she received pulse therapy with methylprednisolone and methotrexate, with partial improvement in muscle disease activity. However, the patient showed persistent skin activity, difficult to reduce corticosteroids, and in a multidisciplinary follow-up with Dermatology, it was chosen to associate dapsone 2 mg/kg/day. About 3 months after, the patient presented symptoms of tachypnea with the need for oxygen through a nasal catheter and mild central cyanosis. She also presented a discrepancy between pulse oximetry and partial pressure of oxygen in arterial blood gases. Chest X-ray was unaltered, transthoracic echocardiogram with no signs of pulmonary hypertension or other relevant alterations, and a negative polymerase chain reaction test for SARS-CoV-2. Arterial blood gases indicated pO_2 80 mmHg, pCO_2 35 mmHg, HCO_3 28 mmol/L, O_2 saturation of 95.7% and methemoglobin dosage of 10.9%. Based on all the exams and investigation, the diagnosis of methemoglobinemia has been confirmed. Considering the mild clinical symptoms and associated with chronic muscle weakness, mild anemia, and given methemoglobinemia of less than 20%, we have decided to discontinue only dapsone and observe the progress of the respiratory condition. No additional treatment was indicated at that time. After few days, the patient evolved with improvement in tachypnea, normalization of pulse oximetry and methemoglobin levels.

CONCLUSION

We presented this case to emphasize that medications frequently used in patients with rheumatic diseases, including dapsone and antimalarial drugs, can lead this potentially fatal complication. Early recognition of methemoglobinemia is important to avoid negative outcomes.

KEYWORDS

Methemoglobinemia, Dermatomyositis, Dapsone.