A Transplant Patient with Blue Lips, Tongue, and Fingertips
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Case Description
A 55-year-old woman, who received a kidney transplant 7 years prior, was admitted with cough, shortness of breath, and low-grade fever. Imaging showed bilateral perihilar infiltrates and bronchoscopic culture confirmed the diagnosis of Pneumocystis jiroveci pneumonia. Given her history of sulfa allergy (skin rash and hives), she was treated with trimethoprim and dapsone. Pretreatment, glucose-6-phosphate dehydrogenase levels were normal. On the third day of treatment, she developed cyanotic discoloration of her tongue (Figure 1A), lips (Figure 1B), and fingertips. Peripheral oxygen saturation was initially 90%, but improved to 94% with supplemental oxygen. Methemoglobin levels in the blood were 8.9% (normal ≤1.5%). On this basis, she was suspected to have dapsone-induced methemoglobinemia. Dapsone was discontinued and low-dose cotrimoxazole was started. Over the next 5 days, her methemoglobin levels decreased to 1.1% and she no longer required supplemental oxygen.

Methemoglobinemia is an infrequent complication of medications including dapsone, metoclopramide, nitroglycerine, primaquine, sulfonamides, quinones, chloroquine, and anesthetic agents (e.g., benzocaine, lidocaine, and prilocaine) (1,2). Congenital methemoglobinemia is relatively rare. Acquired methemoglobinemia results when increased oxidative stress within red blood cells (e.g., due to medications) leads to the conversion of iron, within hemoglobin, from its ferrous to its ferric form, which cannot bind oxygen. It should be suspected in any patient with cyanotic discoloration and/or evidence of hypoxia, but not necessarily hypoxemia, because the partial pressure of oxygen in the blood can remain normal. During phlebotomy, chocolate, brown, or dark-red blood may be observed. Unlike our patient’s mild case, hypoxia is often refractory to supplemental oxygen. The best first test to detect the condition is a venous or arterial blood gas analysis with co-oximetry (3). The Evelyn–Malloy assay, which quantifies the methemoglobin, remains the most accurate test for diagnosis. Management includes removing the offending agent and, if hypoxia is severe or not improved with supplemental oxygen, administering methylene blue or ascorbic acid (4).

Teaching Points
• Methemoglobinemia should be suspected in a patient with new hypoxia, while on therapy with Dapsone.
• Diagnosis involves sending an arterial or venous blood gas analysis and quantitative estimation of methemoglobin level, if available.
• Definite therapy is discontinuing the offending agent and, if the condition is severe, treatment with methylene blue or ascorbic acid.

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E.G. Clark reviewed and edited the manuscript; C. Hesketh conceptualized the study; S. Sriperumbuduri wrote the original draft; and all authors contributed to data collection and critical evaluation of the collected data.

References


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