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When Prophylaxis Complicates Treatment: Dapsone-Induced Methemoglobinemia in a Patient with Adult-Onset Still's Disease and Macrophage Activation Syndrome

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BACKGROUND AND IMPORTANCE

Adult-onset Still's disease (AOSD) is a **rare autoinflammatory disorder** that may present with **life-threatening complications** such as **interstitial lung disease** and **macrophage activation syndrome (MAS)**, which occurs in only 8-14% of patients. Management is challenging due to disease relapses, multimorbidity, and drug-related toxicities. ***Pneumocystis jirovecii* pneumonia (PJP) prophylaxis is required in immunosuppressed patients**, although contraindications to first-line agents may force the use of alternatives with potential toxicity.

AIM AND OBJECTIVES

We report the case of a **72-year-old woman with AOSD** (involving skin, lungs and eyes) **on long-term anakinra treatment who developed severe dapsone-induced metahemoglobinemia**. In May 2025, glucocorticoid therapy was initiated for pulmonary disease progression. Due to history of cotrimoxazole-induced Stevens-Johnson syndrome, **cotrimoxazole was contraindicated for PJP prophylaxis and dapsone was prescribed** instead after confirming normal glucose-6-dehydrogenase (G6PD) activity. **After 5-6 weeks of treatment** the patient presented to **the emergency department** with fever, respiratory failure, cyanosis, anemia and livedo reticularis.

MATERIAL AND METHODS

Laboratory tests on admission showed **leukocytosis**, **thrombocytopenia**, and elevated inflammatory markers. **Methemoglobinemia reached 22%**. She was diagnosed of **dapsone-induced methemoglobinemia**. Therefore, dapsone was **discontinued and metahemoglobinemia treated with IV methylene blue** and ascorbic acid, showing rapid improvement. **During hospitalization**, she developed **MAS with hyperferritinemia** (117,200 ng/mL), **hypertriglyceridemia** (393 mg/dL), **hypofibrinogenemia** (1,64 g/L), **and cytopenias** (Hb 88 g/L, platelets 10×10^9/L) **and plasma's CD25 levels of 4165 UI/mL**. **For MAS treatment**, she required **intensive care treatment** with **high-dose methylprednisolone** (500 mg/day for 3 days, followed by oral corticosteroids in a tapering regimen at 1 mg/Kg/day), **tocilizumab** (8 mg/kg), **and anakinra** (200 mg/day). The **clinical pharmacist followed the patient throughout admission**, supporting immunosuppressive adjustments, and contributing to the therapeutic consensus that ensured an individualized treatment.

RESULTS

The patient achieved **clinical remission of MAS** after intensive immunosuppressive therapy. At discharge, **anakinra was switched to monthly tocilizumab due to persistent thrombocytopenia**. **For PJP prophylaxis**, patient is **currently treated with monthly inhaled pentamidine**. Treatment was tolerated without further adverse events.

CONCLUSIONS

This case highlights the **complexity of AOSD treatment in fragile elderly patients**, where overlapping diseases, prophylactic constraints and drug toxicities complicate management. **Pharmacist involvement** is **essential** to identify **safer prophylactic alternatives**, **optimize immunomodulation**, and **support multidisciplinary decision-making**.