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INTRAVENOUS IMMUNOGLOBULIN FOR CUTANEOUS FEATURES OF VEXAS SYNDROME: A CASE REPORT

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Premises. VEXAS syndrome is an autoinflammatory disorder caused by somatic mutations in the UBA1 gene and is characterized by severe inflammation and hematologic disorders with systemic manifestations. Cutaneous involvement is the most frequent feature and is often refractory to therapy.

Case Report. We describe an otherwise healthy 60-year-old man with VEXAS syndrome presenting with systemic inflammation (fever, arthralgia, elevated inflammatory markers) and myelodysplastic syndrome. Initial treatment with high dose glucocorticoids (2 mg/kg) was complicated by *Candida parapsilosis* sepsis. Following resolution of the infection after antifungal therapy, the patient developed severe cutaneous manifestations consisting of erythematous papules and plaques involving the chest, neck, face, and gluteal. Histopathological examination of a skin biopsy revealed leukocytoclastic vasculitis. Given the recent patient's immunocompromised status, intravenous immunoglobulin (IVIg) was administered at a dose of 2 g/kg over five days, while maintaining a stable glucocorticoid dosage. Treatment with IVIg resulted in marked improvement, with near- complete resolution of skin lesions after completion of the treatment course.

Conclusions. Management of VEXAS syndrome remains challenging, as no consistently effective therapies have been established. In our case, IVIg led to rapid and sustained improvement of cutaneous manifestations, suggesting that it may represent a safe and effective therapy. Further studies in larger cohorts are needed to define the role of IVIg in the treatment of VEXAS syndrome.