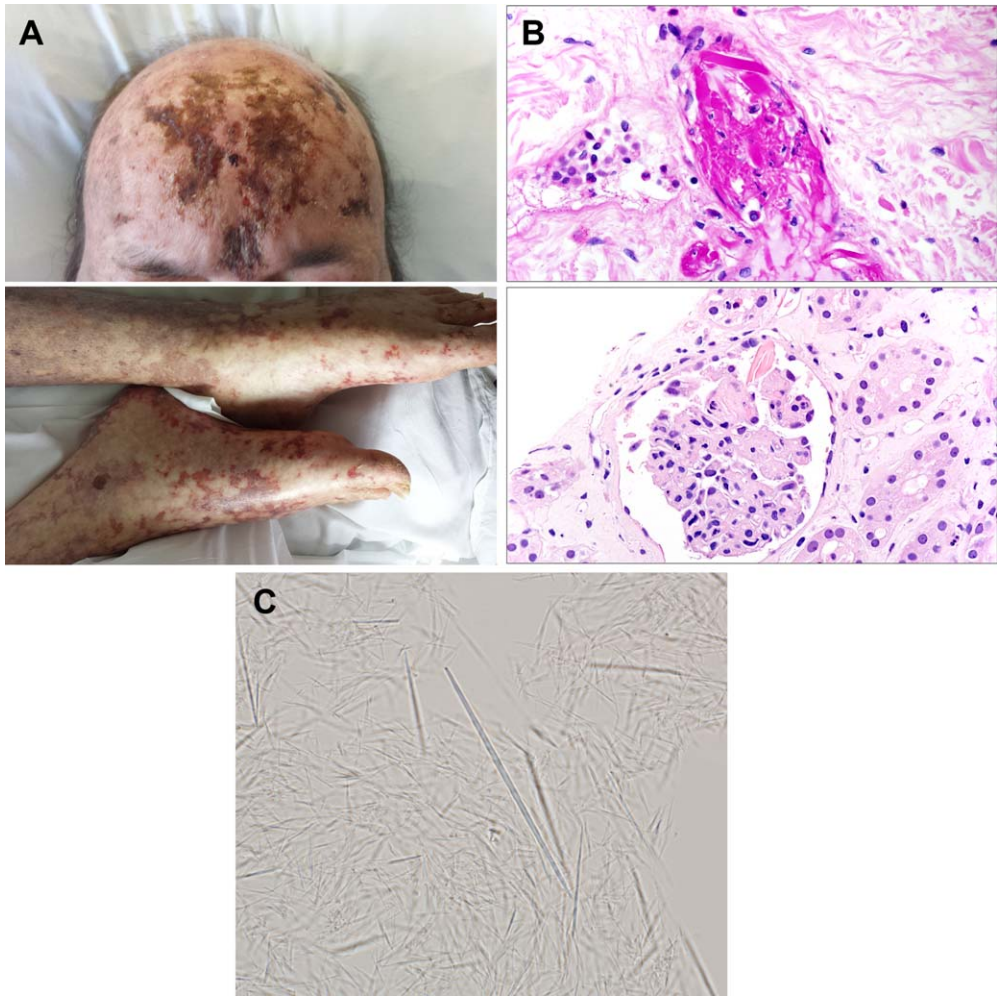


A case of cryocrystalglobulinemia

Sarah E. Avedschmidt,[1] Evan A. Farkash,[2] and Chisa Yamada[1]

A 52-year-old male with chronic obstructive pulmonary disease was repeatedly hospitalized with respiratory and renal failure at an outside institution. He developed deep venous thrombosis, basal ganglia hemorrhage, and sepsis; he was found to have an IgGκ monoclonal gammopathy (serum protein electrophoresis, 0.1 g/dL M-protein).

From the [1]Transfusion Medicine Division and [2]Anatomical Pathology, Department of Pathology, University of Michigan, Ann Arbor, Michigan.

Address correspondence to: Chisa Yamada, MD, Blood Bank/Transfusion Medicine, Department of Pathology, University of Michigan, 1500 E. Medical Center Drive, SPC 5054, 2F228 UH, Ann Arbor, MI 48109-5054; e-mail: yamadac@med.umich.edu.

Received for publication September 11, 2015; revision received December 22, 2015; and accepted December 23, 2015.
doi:10.1111/trf.13528

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TRANSFUSION 2016;56;1678–1679

Seven months later, he presented at our institution with a diffuse purpuric rash covering his entire body (see figure, A, purpuric rash on forehead and feet), renal failure, and mild thrombocytopenia. Antibody screen or direct antiglobulin test and hemolysis labs were negative, ADAMTS13 activity was 53%, and bone marrow biopsy was unremarkable. A skin biopsy showed neutrophilic thrombotic vasculopathy with crystals; a kidney biopsy showed cortical necrosis, fibrocellular crescents, arterial thrombi, and extensive tubular injury with occasional tubular and glomerular crystals (see figure B, intravascular crystals in skin biopsy [top, PAS staining] and glomerular crystals in kidney biopsy [bottom, H&E staining]). Cryocrit was 12% and he was diagnosed with Type I cryocrystalglobulinemia. Cryocrystalglobulinemia is a rare type of cryoglobulinemia that results in microvascular lesions due to the crystallization of monoclonal immunoglobulins often causing more severe manifestations than other cryoglobulinemias.[1] Therapeutic plasma exchange (TPE) treatment is a Category I application for cryoglobulinemia by the apheresis recommendations from the American Society for Apheresis.

After steroid, cyclophosphamide, and bortezomib therapy and eight TPE procedures (one plasma volume exchange replacing with albumin) over 1 month, the cryoglobulin became undetectable and renal function improved. However, he developed gastrointestinal bleeding from the cryoglobulinemic vasculopathy and relapsed with a cryocrit of 42%, renal and respiratory dysfunction, and mental status change. The cryoprecipitate extracted from his blood sample for cryocrit showed spindled crystals (see figure C, crystals in cryoprecipitate extracted from the patient blood). After medication therapy, dialysis, and 11 additional TPE procedures over 1 month, his cryocrit decreased to 5% with improvement of mental status but not organ function. He eventually withdrew all treatment and expired 12 months after his initial diagnosis. In our case, TPE improved symptoms and lowered cryocrit by removing cryocrystalglobulins temporarily; however, it did not change the disease process.

CONFLICT OF INTEREST

The authors have disclosed no conflicts of interest.

REFERENCE

1. Gupta V, El Ters M, Kashani K, et al. Crystalglobulin-induced nephropathy. J Am Soc Nephrol 2015;26:525-9.