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Rapid Improvement of Scleromyxedema Associated With IgG Kappa Monoclonal Gammopathy After Intravenous Immunoglobulin (IVIG)

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Program: Dermatology
Type: Case Report

Background: A 76-year-old male with a history of asthma and chronic inflammatory demyelinating polyneuropathy status post IVIG presented to clinic with 4 months of worsening skin changes and Raynaud’s phenomenon. On examination he had many firm, waxy skin-colored papules on the face, ears, neck, trunk, arms, hands, and knees often in a linear array. Deep furrows were present on the brow (leonine facies), chest, and back (Shar-Pei sign). Induration of the skin surrounding central depressions over the proximal interphalangeal joints (doughnut sign) was noted.

Case: A punch biopsy of the upper back revealed a superficial and deep perivascular lymphocytic infiltrate with abundant interstitial mucin and focal CD68-positive histiocytes between collagen bundles. Given clinical concern for scleromyxedema, serum and urine protein electrophoresis followed by immunofixation was performed which demonstrated IgG kappa monoclonal gammopathy. Subsequent bone marrow biopsy showed a kappa plasma cell change of 10%. Extensive workup for his Raynaud’s phenomenon yielded a positive ANA, 1:320 with a speckled pattern, negative anti-Scl-70, and anti-centromere antibodies. Consent was obtained to use this case for educational purposes.

Conclusion: Scleromyxedema is a chronic disease of cutaneous mucin deposition with high morbidity and mortality. This patient uniquely demonstrated IgG kappa, rather than lambda, monoclonal gammopathy. There are no established treatment guidelines, however IVIG is used most often as first-line therapy. The patient was started on IVIG 2 g/kg given over 2 consecutive days, every 4 weeks. At 3-month follow-up, he demonstrated significant improvement of his skin findings after 4 cycles and reported noticeable improvement within weeks of his first infusion.

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Herpes Simplex Infection Masquerading as Cutaneous Vasculitis: A Clinicopathologic Case Series

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Background: Herpes simplex virus (HSV) is known to have classic histopathologic features of ballooning degeneration and acantholysis with characteristic viral cytopathic changes, including enlarged pale keratinocytes with steel-gray nuclei, marginated chromatin, multinucleated cells, nuclear molding, and eosinophilic intranuclear inclusions. It has been established that leukocytoclastic vasculitis (LCV) is a common concurrent finding. We present a case series of two patients with HSV who had histopathologic findings of LCV without viral cytopathic changes. Consent was obtained for educational use of these cases.

Case: Patient One is a 52-year-old male, with CNS lymphoma who was hospitalized for pneumonia and presented with oral ulcerations, generalized umbilicated papules, crusted erosions, and punched-out vesicles. Biopsy revealed LCV with erythrocyte extravasation, fibrinoid necrosis, perivascular neutrophil-rich infiltrate, and karyorrhexis, without cytopathic changes. Polymerase chain reaction (PCR) was diagnostic for HSV. He was successfully treated with acyclovir. Patient Two is a 56-year-old male, with diabetes mellitus and end-stage renal disease who was hospitalized for bacteremia secondary to septic arthritis, and then developed a widespread vesicular eruption, concerning for HSV. Biopsy revealed superficial neutrophilic inflammation, focal fibrinoid necrosis, and subepidermal vesicles, without viral inclusions. However, HSV PCR was positive. He initially improved with acyclovir, but developed retiform purpura with re-biopsy showing LCV, thought to be due to post-infectious vasculitis.

Conclusion: This case series highlights that clinicians should be aware that cutaneous HSV can present as fulminant LCV on pathology. Despite lack of viral cytopathic changes and negative immunohistochemistry, HSV infection should not be ruled out in the setting of strong clinical suspicion. Therefore, it is essential to obtain PCR when clinical suspicion is strong, given the high sensitivity of this diagnostic technique.

Histopathologic images from patient two showing superficial neutrophilic inflammation, focal fibrinoid necrosis, and subepidermal vesicles, without viral inclusions.

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