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

Elliot Blue

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

Jacob Kingsley¹, **Robert Borucki**², Dillon Clarey², Megan Arthur²

¹College of Medicine, University of Nebraska Medical Center

²Department of Dermatology, College of Medicine, University of Nebraska Medical Center

Mentor: Megan Arthur

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

Elliot Blue¹, **Ritu Swali**¹, Elizabeth Mata¹, Corey Georgesen¹

¹Department of Dermatology, College of Medicine, University of Nebraska Medical Center

Mentor: Corey Georgesen

Program: Dermatology

Type: Case Report

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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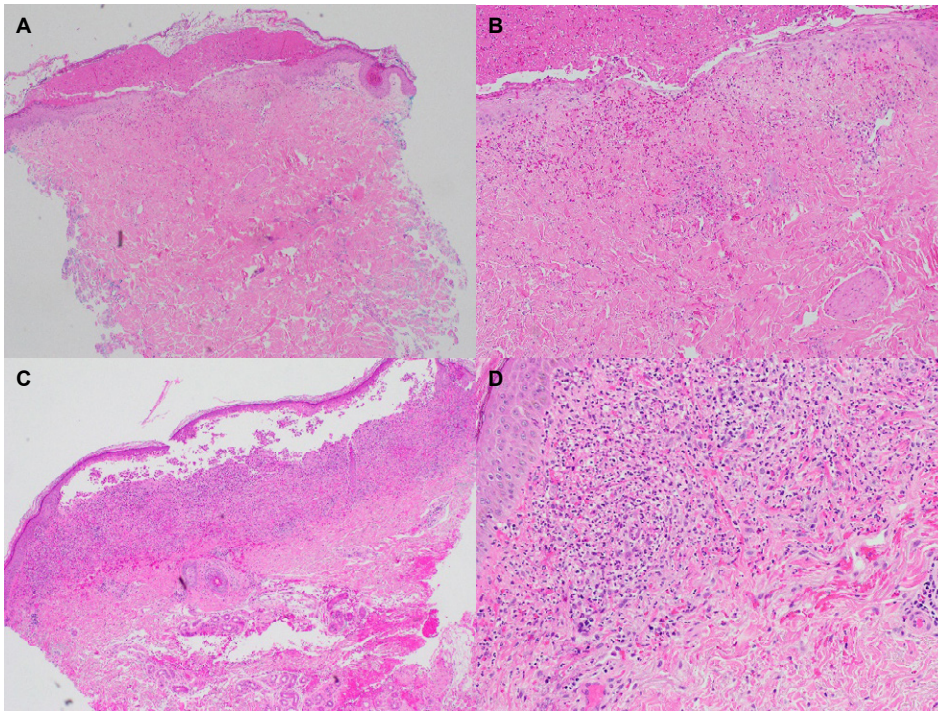


Figure 1. Low power (A) and high power (B) histopathologic images from patient one showing leukocytoclastic vasculitis (LCV) with erythrocyte extravasation, fibrinoid necrosis, perivascular neutrophil-rich infiltrate, and karyorrhexis, without cytopathic changes. Low power (C) and high power (D)

Atypical Knee Hyperextension Injury

Ryan Wendt¹, Jason Meredith¹

¹Department of Family Medicine, College of Medicine, University of Nebraska Medical Center

Mentor: Jason Meredith

Program: Family Medicine

Type: Case Report

Background: Subchondral knee fractures are typically non-traumatic, over-use injuries that occur in the middle aged or elderly. Adolescents rarely suffer subchondral fractures of the knee. These fractures are usually managed conservatively, and typically require several months of recovery before returning to baseline.

Case: A 14-year-old female volleyball player presented for evaluation of left knee pain. She injured her knee during agility drills

the day prior, feeling a pop in her knee and subsequent instability. She initially tolerated weight bearing but was unable the following day. On exam, she had a moderate effusion, limited range of motion (ROM) secondary to pain, and tenderness to palpation over lateral proximal tibia and lateral femoral condyle. Plain films were obtained and unremarkable; however, MRI demonstrated subchondral impaction fractures of the anterolateral tibia and lateral femoral condyle.

She was made non-weightbearing, placed in a compression sleeve, and advised to transition slowly to weight bearing as tolerated. At 6-week follow up, she remained NWB secondary to pain. Her ROM remained significantly restricted. She was

made toe-touch weight bearing with goal of transitioning to weight bearing as tolerated in 7-10 days. She was referred to physical therapy and made slow improvements in strength and ROM over 2.5 months. She returned to volleyball 5 months post injury. Consent was obtained to use this case for educational purposes.

Conclusion: This case represents a rare presentation of a non-contact, knee hyperextension injury causing subchondral compression fracture without other associated knee injury in an adolescent. While rare in the young, management and expectant recovery was like that seen in older adults. ■

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Chronic Lymphocytic Infiltration With Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS): A Case Report in a Teenager

Mohamed Darwish¹, Daniel Zhou¹, Kathleen Healey¹, Lakshman Arcot Jayagopal¹, Elizabeth Hartman¹, Mac McLaughlin¹, Rana Zabad¹

¹Department of Neurology, College of Medicine, University of Nebraska Medical Center

Mentor: Rana K. Zabad

Program: Multiple Sclerosis/
Neuroimmunology

Type: Case Report

Background: Chronic lymphocytic infiltration with pontine perivascular

enhancement responsive to steroids (CLIPPERS) is a rare central nervous system inflammatory disorder first described in 2010. The mean age of onset is 50-years. Very few cases were reported in youth. Its cardinal features include punctate gadolinium enhancement 'peppering' the pons, adjacent hindbrain, and spine on magnetic resonance imaging (MRI). Brain biopsy demonstrates a

T-cell predominant infiltrate. CLIPPERS is a steroid-dependent condition requiring long-term immunosuppression with methotrexate, hydroxychloroquine, or cyclophosphamide. Recently, treatment with rituximab resulted in disease remission. We describe a pediatric case of CLIPPERS with planned rituximab treatment.