Sterile Neutrophilic Folliculitis with Vasculopathy in a Young Male Patient with Infective Endocarditis

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Sterile Neutrophilic Folliculitis with Vasculopathy in a Young Male Patient with Infective Endocarditis

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HISTORY OF PRESENT ILLNESS: The patient is a 34 year-old male. He was admitted to the hospital with methicillin sensitive Staphylococcus aureus (MSSA) infective endocarditis complicated by septic pulmonary emboli, cervical osteomyelitis with soft tissue extension, bacteremia and acute kidney injury. On hospital day four, the patient abruptly developed a follicular rash on bilateral upper extremities. He denied any associated symptoms including itching, burning or pain. He had a fever of 38.6 C.

MEDICAL/SURGICAL HISTORY: Hepatitis C, anemia, osteomyelitis, intravenous drug abuse, opioid addiction

MEDICATIONS: Vancomycin, cefazolin, linezolid, triamcinolone 0.1% cream

CURRENT TREATMENT: Triamcinolone 0.1% cream twice daily, treatment of MSSA endocarditis with vancomycin, cefazolin, linezolid, treatment of AKI with dialysis

PHYSICAL EXAMINATION: On initial exam, pink to purple, some eroded, papules on the bilateral distal upper extremities. Scattered, non-blanching, bright pink macules on the distal lower extremities. Three days later, the lesions evolved into larger, firm, pink, centrally umbilicated, semi-translucent papules with few similar-appearing lesions on the bilateral lower extremities. Seven days later, the lesions began to spontaneously involute, leaving pink, crusted papules and macules without scars.

LABORATORY DATA: Blood culture positive for MSSA, C-reactive protein 119mg/L (<7.0), hemoglobin 9.9 g/dL (12.5-17), creatinine 2.91mg/dL (0.53-1.3), hepatitis C virus RNA 8,830,000 IU/mL. HIV Ag/Ab, C3, C4, TSH, tissue culture, wound culture, remaining CBC, CMP are negative or WNL.

REFERENCES

1. Magro C, Crowson A. Sterile neutrophilic folliculitis with perifollicular vasculopathy: a distinctive cutaneous reaction pattern reflecting systemic disease. *J Cutan Pathol.* 1998;25(4):215-221.
2. Patterson JW. *Practical Skin Pathology: A Diagnostic Approach*. Philadelphia, PA: Elsevier/Saunders; 2013.

BIOPSY: *Health Network Laboratories* (S18-27413, 07/03/2018) Right Arm: "Upper and mid dermal perivascular chronic inflammation and dense perivascular and interstitial acute inflammation with background leukocytoclastic debris, perivascular fibrin deposition with fibrinoid degeneration of the vessel walls, subepidermal and intraepidermal vesicle formation with associated neutrophils and some amount of epidermal necrosis concentrated around a central hair follicle."

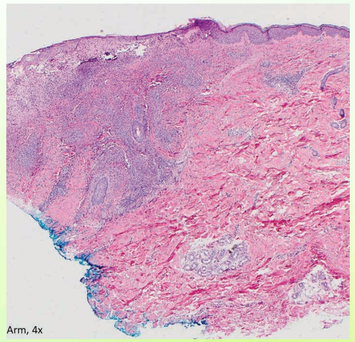

FIGURE 1: Punch biopsy of right arm: Hematoxylin and Eosin stain at 4x magnification. Mid dermal and perivascular acute and chronic inflammation concentrated around central hair follicle.

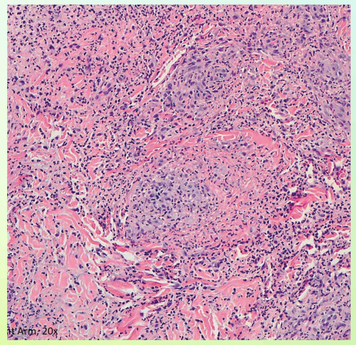

FIGURE 2: Punch biopsy of right arm: Hematoxylin and Eosin stain at 20x magnification. Leukocytoclastic debris, perivascular fibrin deposition with fibrinoid degeneration of vessel walls.

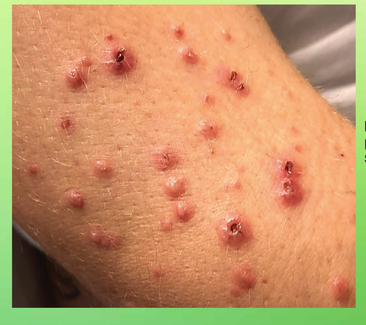

FIGURE 3: Right arm with firm, pink, centrally umbilicated, semi-translucent papules

DISCUSSION: Sterile neutrophilic folliculitis with vasculopathy is a distinctive cutaneous pattern first described by Magro and Crowsen in 1998. This rare entity is characterized by a constellation of histopathological findings, which consist of neutrophilic or suppurative and granulomatous folliculitis in addition to a Sweet's-like vascular reaction or leukocytoclastic vasculitis. It is believed to be a reaction pattern to underlying systemic diseases such as Behcet's disease, reactive arthritis, inflammatory bowel disease, hepatitis B, various connective tissue diseases, and distant extracutaneous infections.

The association between sterile neutrophilic folliculitis with vasculopathy and an underlying systemic disease is based on speculation. The exact etiology remains unclear. The leading theory describes an aberrant humoral or cell-mediated immune response to various endogenous or exogenous triggers in a predisposed host.

Lesions vary in clinical presentation and may appear as folliculitis, vasculitis, or vesiculopustular or acneiform eruptions preferentially involving the trunk, lower extremities, and upper extremities. Constitutional symptoms such as fever, arthralgias, or malaise may accompany the lesions.

Histopathology reveals neutrophilic or suppurative and granulomatous folliculitis in conjunction with either 1) perivascular and intramural neutrophilic infiltrate with leukocytoclasia and erythrocyte extravasation, demonstrating a Sweet's-like vascular reaction or 2) fibrinoid necrosis of vessel wall erythrocyte extravasation, demonstrating a leukocytoclastic vasculitis. Tissue cultures and special stains reveal the absence of infectious pathogens.

The finding of sterile neutrophilic folliculitis with vasculopathy does not necessarily indicate the need for an exhaustive systemic disease workup. However, an infectious trigger or underlying systemic illness such as connective tissue disease, inflammatory bowel disease, or Behcet's disease should be considered. Treatment options have not yet been elucidated, however management of the underlying condition is the mainstay of treatment at this time.