



MORNING REPORT

CLINICAL REASONING FOR A WORSENING RASH: WHEN YOU'RE LEFT WITH THE DIAGNOSIS OF EXCLUSION

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A 74-year-old man with a history of myelodysplastic syndrome was transferred from an outside hospital with subacute, progressive cutaneous and oral lesions and fever which had worsened despite broad spectrum antibiotics. A few weeks prior to admission he was started on 60 mg of prednisone for a new diagnosis of temporal arteritis. The dose of prednisone had been decreased to 30 mg on admission. The patient reported a recent history of inadvertently injuring his left fourth finger, a tender, “marble-like” mass in his left axilla, and erythema and swelling of his left eyelid.

The differential diagnosis includes three main categories: infectious diseases, rheumatologic disorders, and paraneoplastic conditions. A typical bacterial infection seems less likely given the patient’s worsening on broad spectrum antibiotics. Atypical infections such as Nocardia, mycobacteria, or fungal infections should be considered. The patient was recently diagnosed with a rheumatologic disease, temporal arteritis, though this does not explain the lesions on his skin or mucosa. Other considerations include neutrophilic dermatoses, inflammatory disorders that can present with fever and rash. Given patient’s age and history of myelodysplastic syndrome, one should consider paraneoplastic syndromes and age-appropriate cancer screening.

The patient’s exam revealed left conjunctival irritation and a left upper palpebral conjunctival erosion, multiple 2-6 cm eschars with heaped up brightly erythematous to slightly violaceous borders on the left upper chest, left axilla, and right neck. Additionally, he had an exophytic, hyperplastic, erythematous plaque on the upper frontal gingiva, and a necrotic appearing finger. Dermatology was consulted and performed a punch biopsy of the left upper chest that revealed a diffuse dermal neutrophilic infiltrate with ulceration and necrosis. Blood and tissue cultures were negative, as was a broad-spec-

trum polymerase chain reaction for common types of bacteria, making infection unlikely. An evaluation for occult malignancy, including a computed tomography scan of the chest, abdomen, pelvis, and positron emission tomography scan, were unremarkable. The patient’s skin and oral lesions continued to worsen.

In conjunction with dermatology, it was determined that the most fitting diagnosis was “pyodermatitis/pyostomatitis vegetans” (PD/PSV) a type of neutrophilic dermatosis that involves skin and mucosa. Typical first-line treatment is steroids so the patient’s prednisone dose was restarted at 1mg/kg/day (a dose of 70 mg daily) and he quickly began to improve. His oral lesions sloughed off and his skin lesions began to recover.

Neutrophilic dermatoses are a large, uncommon heterogeneous group of auto-inflammatory skin disorders that occur in patients with underlying hematologic malignancies, inflammatory bowel disease, and rheumatologic conditions. The most familiar examples are Sweet syndrome and pyoderma gangrenosum. Other conditions do exist on the spectrum and share features of both disorders.

Neutrophilic dermatoses had been considered early in the patient’s course. PD-PSV would be the best fit but was thought less likely given the rarity in patients without inflammatory bowel disease. This diagnosis was later reconsidered after work-up for other conditions returned negative. Ultimately, PD-PSV was determined to be the best fit given the distribution of skin and mucosal involvement, lack of more viable alternative diagnosis, and swift response to high-dose steroids. Additionally, the history of pathergy following injury of the patient’s finger fits with a diagnosis of neutrophilic dermatosis.^{1,2}

The name of the disorder is descriptive: pyo comes from the Greek word for pus, the breakdown product of neutrophilic inflammation, dermatitis means inflamma-

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tion of the skin, stomatitis means inflammation of the mucosa, and vegetans describes the “heaped up” morphology of the lesions. PD-PSV is a rare subtype seen almost exclusively in patients with underlying inflammatory bowel disease. It is the only type of neutrophilic dermatosis known to consistently affect the mucosa (both oral and genital).³ Eyelid involvement has been documented previously, most notably in a report by Leibovitch et al which describes a case of bilateral eyelid margin ulceration and pustules unresponsive to broad-spectrum antibiotics.⁴

The patient subsequently underwent an outpatient colonoscopy and had three colonic biopsies which were negative for colitis. At a follow-up visit three months after discharge the patient was noted to have complete resolution of his skin and mucosal lesions. A gradual steroid taper was initiated without recurrence at two years of follow up.

First-line treatment for PD-PSV consists of treating the underlying condition. When not feasible, high-dose prednisone, 0.5mg/kg to 1.0mg/kg/day, is warranted in the absence of contraindications. Steroid-sparing agents include dapsone and colchicine, which both have anti-neutrophilic properties.⁵

The patient’s lack of improvement on the initial dose of 60 mg of prednisone and rapid improvement on 70 mg is interesting and indicates that sufficient immunosuppression had not been reached with 60 mg.

The initial temporal artery biopsy slides from the outside hospital were reviewed by members of our ocular pathology department and the diagnosis of temporal arteritis was not substantiated.

The inflammation seen on the temporal artery biopsy was likely neutrophilic inflammation from PD/PSV.

It is unclear whether the patient’s history of MDS was contributory in this case, but we suspect that it may have been given the strong association between leukemia and other marrow abnormalities with Sweet syndrome. There is a single case report of pyoderma vegetans in a patient with MDS of type Refractory Anemia with Excess Blasts.⁶

Summary Points:

- Neutrophilic dermatoses are an unusual group of disorders which should be considered in patients with neutrophilic inflammation underlying rash in the absence of infection.
- Pathergy is a unique sign that is suggestive of neutrophilic dermatoses.
- History of neoplastic conditions particularly lymphoma and leukemia as well as inflammatory bowel disease should be identified as particular risk factors for neutrophilic dermatoses.

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