Granulomatosis with polyangiitis (GPA), formerly known as Wegener’s Granulomatosis, is a rare immune-mediated disorder, affecting fewer than 200,000 US patients per year. It is a poorly understood disease that is genetically associated with the HLA-DRB1 gene. A larger combination of genetic factors as well as environmental elements are involved in its pathogenesis. GPA affects the body systemically by damaging small- and medium-sized blood vessels throughout the body, with a predilection for the airways, sinuses, and also the kidneys. Typical presentations range from mild forms of sinusitis to severe forms of ulcerations of the mucous membranes leading to eventual saddle nose deformity and development of glomerular nephropathies. GPA can also affect the skin creating palpable purpura, ulcers, nodules, and pustules and more rarely can affect the CNS.

We report a rare case of GPA presenting with pyoderma gangrenosum-like lesions, spinal abscess, and more rarely can affect the CNS. A 63-year-old married, retired saleswoman with a past medical history of recurrent sinusitis presented with 10 days of progressively worsening back pain. An MRI demonstrated a phlegmonous prevertebral collection from T10 through L1, concerning for osteomyelitis.

The patient had concurrently presented with a large non-healing ulcer on her right calf, a smaller non-healing ulcer on her lateral right thigh, as well as a painless subcutaneous nodule on her inner right arm and lower left back. The largest ulcer had started as a nodule about 6 months prior where it sustained repetitive trauma, eventually opening and progressing to become a deep erythematous 4x4 ulcer with a violaceous border and necrotic granulation tissue in the center situated over the right tibia.

Given concern for osteomyelitis or bacterial seeding, a CT-guided core biopsy was performed of the spinal collection and showed chronic inflammation, including aggregates of small lymphocytes, plasma cells, and foamy histiocytes, associated with fat necrosis. An X-ray of the largest ulcer on her right calf showed no extension to the bone. A deep wound punch biopsy revealed microabscesses and focally diffuse neutrophilic infiltrates with histiocytic inflammatory response, as well as scattered multi-nucleated giant cells. An infectious disease workup of both areas, including mycobacterium, fungal organisms, bacterial and viral organisms, was negative.

Pathology records from prior sinus surgery demonstrated extensive chronic inflammation with focal necrosis. Immunologic studies included an ANA titer of 1:640 of a homogenous pattern, a C4 of 50 as well as a positive PR3-ANCA. Chest X-ray, renal function and urinalysis were normal. Given her presentation of sinusitis and positive PR3-ANCA, a diagnosis of Granulomatosis with Polyangiitis with Pyoderma Gangrenosum-like lesions was made. She was treated with prednisone and methotrexate with initial good response but about 2 months later, had developed nasal ulceration with the start of progression to saddle nose deformity. She was then switched over to rituximab treatment due to the progressive disease.

**Learning Points**

- Pyoderma gangrenosum-like lesions, spinal abscess, and more rarely can affect the CNS.
- GPA affects the body systemically by damaging small- and medium-sized blood vessels throughout the body.
- Typical presentations range from mild forms of sinusitis to severe forms of ulcerations of the mucous membranes leading to eventual saddle nose deformity and development of glomerular nephropathies.
- GPA can also affect the skin, creating palpable purpura, ulcers, nodules, and pustules.
- More rarely can affect the CNS.

**Resources**