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Case Study
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A Rare Form of Granulomatosis with Polyangiitis

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Clinical Image

Granulomatosis with Polyangiitis (GPA), formerly known as Wegener's Granulomatosis, is a rare immune-mediated disorder, affecting small to medium sized vessels. It is a poorly understood genetically transmitted disease, likely associated with the HLA-DPB1 gene, although it is more probable that a larger combination of genetic factors as well as environmental elements affect its diagnosis [1].

GPA affects the body systemically by damaging small- and medium-sized blood vessels throughout the body, with a predilection for the airways, specifically the nose and lungs. Typical presentations range from mild forms of sinusitis to severe forms of ulcerations of the mucous membranes leading to eventual saddle nose deformity. Other typical organs involved are the kidneys in glomerular-nephropathies. GPA can also affect the skin *via* palpable purpura, ulcers, nodules, and pustules [2] and more rarely can affect the CNS system [3]. We report a rare case of GPA presenting with pyoderma gangrenosum-like ulcers, spinal abscess, sinusitis, and positive ANCA.

A 63-year-old married, retired saleswoman with a past medical history of recurrent sinusitis presents with a concern of relentless and worsening back pain. A CT scan was significant for small lytic erosions along T11 and T12 suspicious for osteomyelitis. An MRI had demonstrated a phlegmonous prevertebral collection extending from T10 through L1 (Figure 1, 2). 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

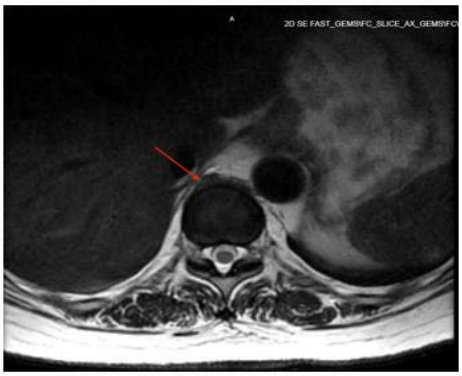

Figure 1: MRI of spine showing phlegmonous prevertebral collection extending from T10-L1.

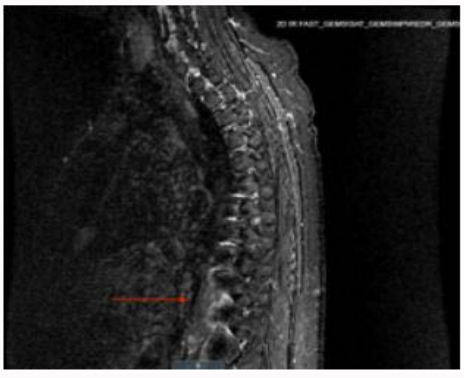

Figure 2: MRI of spine showing phlegmonous prevertebral collection extending from T10-L1.

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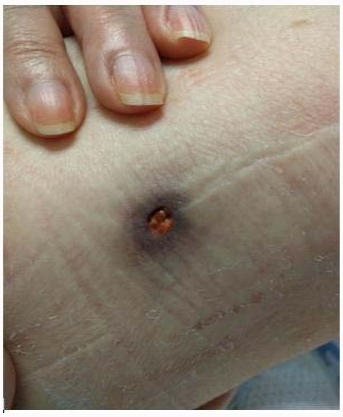

Figure 3: Lateral right thigh (0.5 cm × 0.5 cm).

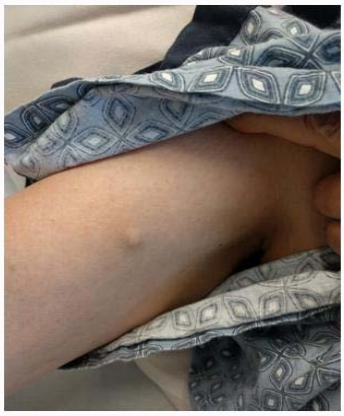

Figure 4: Inner right arm.

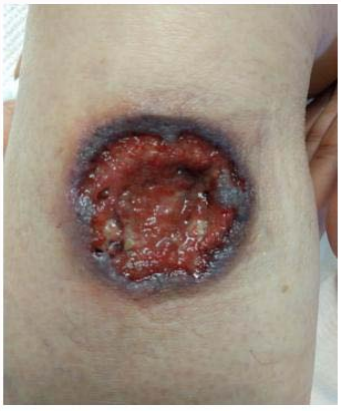

Figure 5: Medial right shin (4 cm × 4 cm).

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 deeply erythematous 4 × 4 ulcer with a violaceous border and necrotic, granulation tissue in the center (Figures 3-6).

With 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

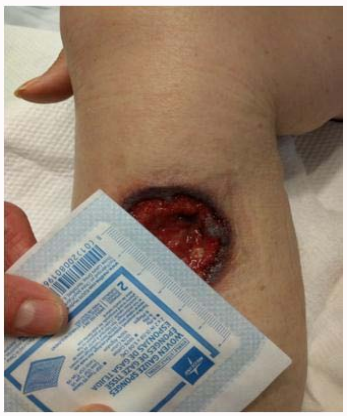

Figure 6: Medial right shin (4 cm × 4 cm).

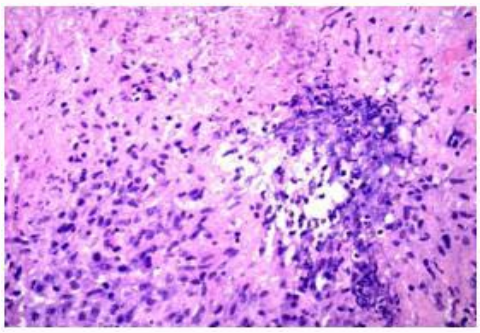

Figure 7: Histological examination of spinal abscess revealing loose granulomatous infiltrates with micro-abscesses and focal necrosis.

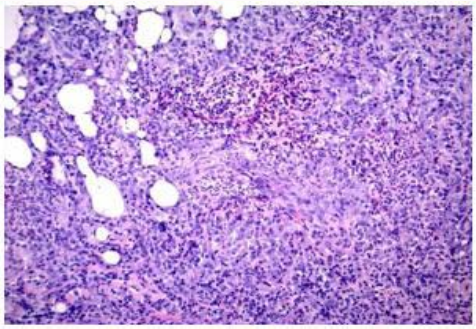

Figure 8: Histological examination of spinal abscess revealing loose granulomatous infiltrates with micro-abscesses and focal necrosis.

bone. A deep wound punch biopsy was performed which had shown micro-abscesses and focally diffuse neutrophilic infiltrates, 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 (Figure 7,8).

A biopsy of her sinuses done prior to her admission 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 managed with prednisone and methotrexate with initial good

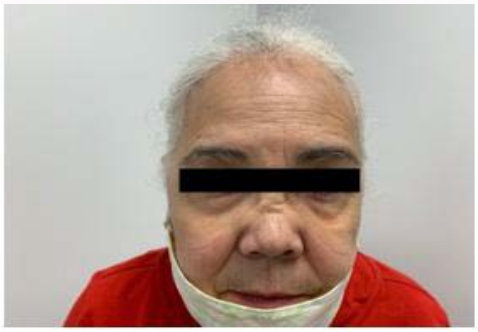

Figure 9: Saddle-nose deformity.

Figure 10: Saddle-nose deformity.

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 (Figure 9,10).

Granulomatosis with polyangiitis (GPA), previously known as Wegener's Granulomatosis is a disease most commonly affecting small-sized blood vessels throughout the body, most commonly in the sinuses, lungs and kidneys [4]. The method of insult is immune-mediated by autoantibodies directed against the neutrophilic granules (PR3-ANCA) thereby causing tissue damage [5].

One of two diagnostic criteria, per the American College of Rheumatology (ACR), must include 2 out of 4 of the following: (1) nasal/oral inflammation, (2) nodules, fixed infiltrates, or cavities shown in chest X-rays, (3) urinary sediment shown in the urinalysis, and/or (4) granulomatous inflammation shown on biopsy of arteries or perivascular areas. Having 2 out of 4 criteria represent an 88.2% sensitivity and a 92% specificity to the disease [6]. Although a PR3-ANCA is not required within the diagnostic criteria, it is present 80% to 90% of the time in patients with GPA [7].

Although all parts of the body can be affected, the most typical presentation includes the upper respiratory tract. It can include recurrent sinusitis with inflammation that can resemble a severe common cold with persistent rhinorrhea, nasal crusting, nasal congestion or bleeding, as well as ulcerations of mucous membranes [4]. Another typical organ system involved are the kidneys, in which patients can develop hypertension, peripheral edema, and can eventually lead to a more serious condition, glomerulonephritis.

Less commonly seen is involvement of the skin, including papules, subcutaneous nodules, ulcers, petechiae or purpura seen in about 15% of patients [8]. Although skin lesions can be seen, the intense ulcerations, like that of Pyoderma gangrenosum, are not typical [9].

Rarer presentations include neurological and cardiac abnormalities, ranging from peripheral neuropathy to hemiplegia and pericarditis, cardiomyopathy and myocardial infarction. Other generalized systemic symptoms include fevers, malaise, polyarthralgia, loss of appetite and general fatigue [4].

Spinal involvement occurs in about 2% of patients, and although the biopsy in our case seemed inconclusive of vasculitis, over 50% of biopsies related to GPA can be considered nonspecific. The most typical pattern of GPA, however, includes granuloma formation with necrosis [3]. Our case illustrates the rare association of both PG-like lesions as well as spinal involvement presenting in GPA [10].

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