Pyoderma Gangrenous Associated with a Chronic Myeloproliferative Disorder

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Pyoderma Gangrenosum (PG) is a rare neutrophilic dermatosis which exhibits an intense dermal inflammatory infiltrate composed mostly of neutrophils with little or no evidence of primary vasculitis. The condition is diagnosed clinically as there are no specific serologic or histologic markers. Typically, it presents as a superficial ulceration of the pretibial region of the legs. We present the case of a patient where a chronic leg ulcer preceded the diagnosis of a chronic myeloproliferative disorder. The patient, an 82 years-old female with severe osteoarthritis, splenomegaly, and recurrent leg abscesses was referred to hematology for further investigation. At the time of presentation, her WBC count was 12.9 x 10⁹/L, hemoglobin 7.5 g/dL and platelet count 742×10⁹/L. Bone marrow biopsy sections showed mixed megakaryocytic-granulocytic proliferation (Figure 1) with diffuse and large clusters of pleomorphic megakaryocytes and marrow fibrosis. The bone marrow aspirate revealed no cytogenetic abnormalities. She developed subcutaneous nodules over the left tibial region. These nodules soon developed into bullous lesions and within two weeks they coalesced and produce extensive ulceration of the leg (Figure 2), which almost threatened the viability of the limb. The histologic changes of the cutaneous lesions were characterized by a dense neutrophilic dermal infiltrate resembling acute neutrophilic dermatosis with no evidence of primary vasculitis. An aspirate from one of the bullous lesions showed an extensive neutrophilic infiltrate (Figure 3).

The patient was started on high dose prednisolone, which resulted in immediate arrest of the progression of the disease and a gradual healing of the ulcer.

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Figure 1: Photomicrograph of the bone marrow biopsy section demonstrating highly cellular marrow with mixed granulocytic (G) and megakaryocytic (M) cellular proliferation. H & E stain.

Figure 2: Photograph showing extensive ulceration of the left leg.

Figure 3: An aspirate from one of the bullous lesions showing extensive neutrophilic infiltrate.