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Clinical Image Open Access

# Neutrophilic Dermatosis Masquerading as Rheumatoid Vasculitis in a Patient with Occult Myelodysplastic Syndrome

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## Clinical Image

A 74 year old female with 15 year history of seropositive deforming rheumatoid arthritis (RA) treated with methotrexate for 9 years in the past (discontinued due to cytopenias), presented for evaluation of unusual skin lesions (Figure 1).

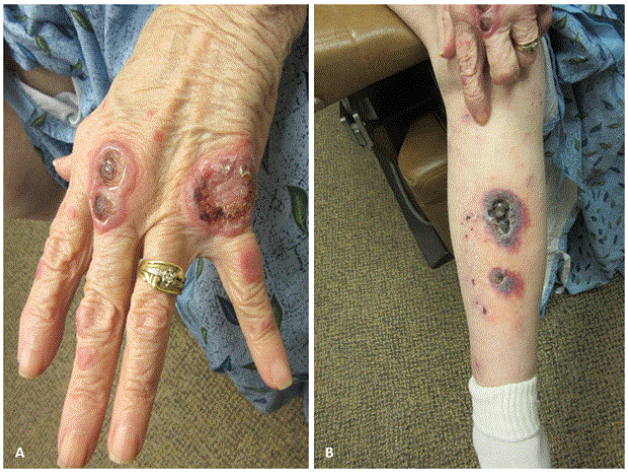


**Figure 1:** Skin lesions developing into ulcerated plaques with necrosis.

These started as a painful, purulent, erythematous vesicle on the dorsal 3rd metacarpophalangeal joint (MCP), progressing rapidly to multiple similar lesions developing into ulcerated plaques with necrosis over other MCPs, legs, trunk, buttocks and face over a 1 month period. Drug reaction, infection and cutaneous vasculitis secondary to RA were all considered in the differential. Dermatology evaluation raised concern for neutrophilic dermatoses. Skin biopsy confirmed widespread dermal neutrophilic infiltration without vasculitis. A diagnosis of bullous sweets syndrome (1A) and pyoderma gangrenosum (1B) (PG) was made. Laboratory tests were concerning for severe anemia (Hb 6.9 g), macrocytosis (MCV 100.5), leucopenia (WBC 2.6) and neutropenia (absolute neutrophil count 980).

These in conjunction with her recent diagnosis of a neutrophilic dermatosis, raised suspicion for an underlying hematologic malignancy. A subsequent bone marrow biopsy (Figure 2) showed hypercellular marrow with panhyperplasia (2A, Hematoxylin-Eosin stain, 200x), dysplastic erythroid precursors with budding nuclei (2B, Wright Giemsa stain 600x), numerous ring sideroblasts (2C, Iron stain 600 x), dysplastic megakaryocytes with small separated nuclei (2D, Wright-Giemsa stain 600x), increased monocytic and myelomonocytic precursors (2E, Butyrate and Chloracetate Esterase stain, 400x) compatible with myelodysplastic syndrome (MDS). Peripheral blood smear also showed dysplastic hypogranular neutrophils (left), large platelets (center) and atypical-looking monocytes (right) (2F, Wright-Giemsa 600x). The patient's skin lesions responded well to oral glucocorticoid therapy initially, but PG recurred as the dose was tapered and Infliximab was initiated. MDS was managed with supportive care.

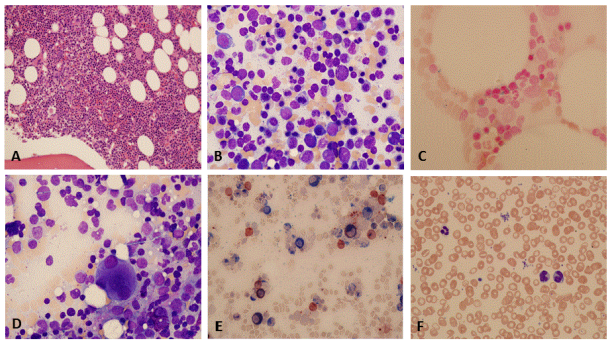


**Figure 2:** Bone marrow biopsy.

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