INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

# FAMILIAL PYODERMA GANGRENOSUM ASSOCIATED WITH FAMILIAL MYELODYSPLASTIC SYNDROME

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Background: pyoderma gangrenosum (PG), an uncommon neutrophilic dermatosis, presents as an ulcerative disorder of the skin. More than half of cases are associated with underlying systemic disorder from which hematologic malignant neoplasms represent 6% of cases. Adults in their fourth to six decade of life are most commonly affected. Clinically, PG has 4 subtypes: ulcerative, bullous (associated with hematologic neoplasm), pustular and vegetative. We report a new family case of PG with the original association with a myelodysplatic syndrome (MDS).

Observation: we report a new case of pyoderma gangrenosum in two siblings, born of consanguineous healthy parents, with onset after puberty. The first patient is a 35-year-old man who presented for bilateral skin ulcers from knee to toes, treated with wet dressings without improvement. Laboratory workup ruled out coagulation disorders and vasculitis. Venous and arterial duplex ultrasonography were normal. A skin biopsy showed t he presence of non specific vascular changes not consistent with vasculitis. We made the diagnosis of PG after the exclusion of other conditions. Complete blood count test showed pancytopenia. We made the diagnosis of hypoplastic MDS upon findings in the peripheral blood and bone marrow as interpreted within the clinical context. He was treated with intravenous immunoglobulins with improvement in his clinical status. The second patient, a 30-year-old woman, reported the appearance of bilateral foot ulcers after pubertal period similar to her brother. Also, the diagnosis of PG and hypoplastic MDS was made in the same manner. She was treated with oral corticosteroids with mild improvement. Genetic studies were not performed for financial issues.

Key message:, To our knowledge, eight familial cases of PG were reported. This is the first case report of familial PG associated with familial MDS. The pathogenesis relation between the two diseases remains unclear.