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Successful therapy with apremilast in immunodeficient patient with pyoderma gangrenosum

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Introduction & Objectives:

Pyoderma gangrenosum is a relatively rare disease whose etiopathogenesis is not fully understood. Immune system dysfunction plays a role. It is characterized by the occurrence of small red papules and nodules that rapidly disintegrate into very painful irregularly shaped ulcerations with necrotic base., infiltrates can also be observed in internal organs (lungs, liver, bones).

The histological picture is not entirely typical. The aim of this case report is to present the course of the disease and the effect of apremilast therapy in a polymorbid immunosuppressed patient.

Materials & Methods:

Female, 61 years old, is treated with type 2 diabetes mellitus, severe pulmonary hypertension, corticoid osteoporosis, has proven hypogamaglobulinemia type IgA . Since 2021 abscess deposits with spontaneous perforation and secretion of hemorrhagic-serous fluid have formed on the extremities.

Systemic corticotherapy with prednisone at an initial dose of 60 mg/day in combination with antiulcerative and antibiotic treatment was indicated due to suspicion of pyoderma gangrenosum. In four weeks from the setting of treatment, there was a significant regression of the lesions. When the dose of prednisone was reduced below 30 mg/day, rapid recurrence and formation of disintegrating defects occurred. Due to high doses of corticosteroids, steroid type 2 diabetes mellitus with the need for PAD compensation, the development of Cushing's syndrome in the face, pathological fractures of the Th11, L1, L3-4 vertebrae gradually occurred.

Considering the existing complications and the little effect of the current high-dose corticosteroid treatment, continuation of corticosteroid monotherapy was contraindicated. On 07/2023, apremilast was administered in gradual titration up to 30 mg twice daily in combination with prednisolone 10 mg/day and intensive local treatment using the moist healing method. From 01/2024 monotherapy with apremilast was indicated.

Results:

Complete healing of ulcerations and remission occurred on apremilast therapy 04/2024

Conclusion:

Based on this case report, the conclusions can be generalized in the levels we describe below.

Pyoderma gangrenosum is associatid with idiopathic intestinal inflammation, haematological diseases (myelodysplastic syndrome, myelofibrosis, paraproteinaemia), rheumatoid arthritis or tumours can be observed in up to 50%. Liver diseases (hepatitis, primary biliary cirrhosis), autoimmune diseases (vasculitis, lupus erythematosus and Sjogren's syndrome) and surgical interventions (around the stoma, surgical scars. Associated diseases and paraneoplasia should always be excluded. Oral or intravenous corticosteroids are most commonly used in therapy. Other immunosuppressive therapies include apremilast, azathioprine, cyclophosphamide,

mycophenolate mofetil, methotrexate and dapsone. In recent years, biological therapy, especially TNF alpha inhibitors (adalimumab, infliximab), has also been used.