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Plasmapheresis in a Patient with “Refractory” Urticarial Vasculitis
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Background: Immune complexes have been found in the circulation approximately 30 to 75% of patients with urticarial vasculitis and much evidence supports the role of these immune complexes in the pathogenesis of urticarial vasculitis. Plasmapheresis is effective in removal of these immune complexes. However, few cases have been reported regarding the use of plasmapheresis in the treatment of urticarial vasculitis.

Methods: A 35-year-old woman presented with history of recurrent episodes of generalized painful urticarial plaques often lasting 9 years associated with swelling of her parts of body. Examination revealed multiple urticarial plaques distributed all over the body (particularly in the extremities, palms and soles). The initial laboratory studies, including a complete blood count, thyroid function tests - thyroid autoantibodies, erythrocyte sedimentation rate, hepatitis markers, liver and renal function tests, urinary analysis, stool analysis for parasite ova, total IgE, C3, C4, C1q, CH50, C1 inhibitor levels and antinuclear antibodies were found to be within normal range. Skin prick tests were performed with commonly consumed foods in Turkey found to be negative. A biopsy from an affected area of skin revealed an urticarial vasculitis. Based on the biopsy results, the patient was diagnosed with UV. Treatment with H1/H2-antihistamines and oral corticosteroids (1 mg/kg/day) had been unsuccessful; therefore hydroxychloroquine 400 mg/day was added. Unfortunately hydroxychloroquine was stopped in the second month due to the emergence of an adverse event (keratopathy). The patient underwent plasma exchange 2 times with an interval of 6 months. Five percent albumin solution as replacement fluid was used. One plasma volume was processed in each session. Apheresis procedure was performed with the “Cell Separator” device. The plasmapheresis procedures were completed without any adverse events. At 13 months after the plasmapheresis, the urticarial plaques were reappeared, but the severity and duration of symptoms were lower than before the plasmapheresis. The newly lesions were re-treated with short-term oral antihistamine regimen.

Conclusions: In conclusion, the presented report supports the usability of plasmapheresis in patients with “refractory” UV. Further clinical studies are needed to confirm our experience.

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Delayed Diagnosis of Hereditary Angioedema. A Case Report
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Background: Hereditary angioedema (HAE) was first described by Quincke in 1882 and appointed by Osler in 1888, is a rare disease caused by deficiency of gene esterase inhibitor C1 (C1 INH). Prevalence varies from 1:10,000 to 1:150,000. The attacks are usually sporadic and often associated with traumatic or stressful events. Treatment included management of acute attacks and prophylactic therapy in specific situations where attacks may occur.

Methods: A 40-year-old male with a family history of father facial angioedema. He had experienced 15 episodes of angioedema during the previous 5 years. During these events than lasted 3 to 5 days edema affected his eyelids, lips, hands, feet and testicles. And sometimes was associated to abdominal pain and shortness of breath. He went several times to medical office and emergency room, where he received treatment with antihistamines without improvement.

Results: The laboratory evaluation of complement components showed C4 2±0.8 (NV 20–50), CH50 10.1 (NV 20–50), C1 inhibitor quantitative <1.2 ng Ep/mL (NV > 10.7), and C1 esterase inhibitor functional 104% (NV > 67%), once the diagnosis of type I hereditary angioedema was done, we started danazol therapy that has prevented recurrence of symptoms.

Conclusions: It is important to do a detailed history for the diagnosis and treatment in cases of angioedema. Most patients improve when receiving the right treatment. Recurrent angioedema events even with treatment, the physician must search for malignity and/or autoimmunity disease.

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Urticaria Pigmentosa. Case Report
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Background: Mastocytosis is a disorder characterized by an abnormal proliferation of mast cells and release of cell mediators. The incidence is 1 per 1000 skin diseases attending in dermatology services. Mastocytosis can be divided into 3 different clinical variants: cutaneous, systemic and malign mastocytosis. Urticaria pigmentosa is the most common variant (70–90%) of mastocytosis. Of all cases 55% occur during the first 2 years of life. When the bone marrow, lymph nodes, liver and spleen are affected the disorder is called systemic mastocytosis.

Methods: Case 1: A 20 month old male with history of penicillin and erythromycin allergy, as well atopic family history. Began at 4 months with itchy brown-marrow papules in the back, then generalized except palms and soles. The lesions were exacerbated by heat and rubbing. There was no fever, weight loss, or any other systemic symptoms in the history. Blood count and biochemical laboratories were normal. Skin biopsy reported the presence of mast cells, confirming urticaria pigmentosa diagnosis. The management included antihistamines, restricted diet and emollients with improved of symptoms. Case 2: A 9 month old male with no history of atopy. At the first visit he had 4 months with skin lesions characterized by hyperpigmented maculopapular eruption, scattered on head, over trunk and extremities. Darier’s sign was positive. Skin biopsy is performed with confirming the diagnosis of mastocytosis.

Conclusions: The urticaria pigmentosa diagnosis is mainly clinical, with emphasis on the Darier’s sign, which is pathognomonic and positive in 90% of cases. In some cases a skin biopsy is required to confirm the diagnosis.