587 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 part 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.

588 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 Eq/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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589 Urticaria Pigmentosa. Case Report

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Antihistamines are the first line of treatment. Symptoms relieve spontaneously before adolescence in 50% of pediatric patients. In some cases, a malignant transformation of mastocytosis could occur, condition that is called “mast cell leukemia”.

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590 A Case of Chronic Urticaria Complicated by Raoultella Ornithinolytica Urinary Tract Infection, Bronchospasm and Angioedema

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Background: Raoultella ornithinolytica (formerly Klebsiella ornithinolytica) has been recovered from water, soil, plants and occasionally mammalian mucosae. It is one of the organisms that cause histamine fish poisoning by converting histidine to histamine as it possesses the enzyme histidine decarboxylase.
Methods: We report a case of a 51 year old woman with a history of rheumatic heart disease, allergy to aspirin and penicillin, reflux oesophagitis, atopic dermatitis, stress incontinence and a cystocele attributed to chronic obstructive pulmonary disease. She presented to the emergency unit with a 5 years history of urticaria of unknown cause, complicated by symptoms of a urinary tract infection and 3 episodes of angioedema in the preceding 10 days. She was not on regular medication but on intermittent prednisone, promethazine and was recently treated with omeprazole and sucralfate for reflux oesophagitis. She denied previous use of angiotensin converting enzyme inhibitors. Urine culture grew Raoultella ornithinolytica (>100 000 CFU/mL) after 3 days.
Results: The angioedema resolved on treatment of the UTI with oral ciprofloxacin. She was lost to follow-up and when seen 3 years later she still had intermittent flares of urticaria, but had had no episodes of angioedema.
Conclusions: R ornithinolytica, most commonly described as a causative agent of histamine fish poisoning in humans, possesses histidine decarboxylase, an enzyme that converts histidine into histamine. The resulting elevated levels of histamine usually do not cause severe disease but complications can arise in people with chronic histamine mediated diseases.

591 A Case of Idiopathic Recurrent Isolated Orbital Angioedema with Exophthalmos

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Background: Idiopathic angioedema is a term applied to recurrent episodes of angioedema of unknown etiology. The following is a case report of idiopathic recurrent isolated orbital angioedema with exophthalmos which responds to prolonged courses of oral corticosteroids.
Methods: A 67 year old Caucasian female with aspirin exacerbated respiratory disease (AERD) sought treatment for an acute, progressive painless left eye swelling with exophthalmos without visual deficits or urticaria. High dose corticosteroids were initiated followed by a low maintenance dose. The swelling subsided after one year of corticosteroid therapy. Ten years later, orbital swelling with exophthalmos returned in the same eye. No medications, such as aspirin1 or non steroidal anti-inflammatory drugs,2 were associated with the swelling. A CT of the orbits revealed an isolated proptosis with swelling of the medial and inferior rectus muscles and mild hypertrophy and swelling of the left lacrimal gland. A complete history and physical examination were negative. The family history likewise was negative.
Results: High-dose systemic glucocorticoid therapy was initiated. Symptoms resolved after 1 month of tapered corticosteroid therapy, however, swelling recurred in the orbit within one week. Low dose maintenance corticosteroids were reinitiated with resolution of the orbital swelling. Work-up for acquired C1 esterase deficiency was negative.
Conclusions: An atypical case of recurrent idiopathic isolated orbital angioedema with exophthalmos in a patient with AERD and no triggering factor, systemic findings and a negative evaluation is presented.

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592 A Case of Multiple Simultaneous Urticarial Syndromes Refractory to Treatment

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Background: We report the case of a patient with 3 forms of physical urticaria and his response to treatment.
Methods: An atopic asthmatic 11 year old male was evaluated for a history of recurrent pruritus with a variable, erythematous rash unresponsive to therapy. Since the age of 5 years, he has experienced small red, raised, pinpoint, puritic “bumps” over his entire body except the palms of his hands and soles of his feet. The duration of the lesions was generally 5 minutes to about 1 hour. They occurred with exercise, stress, cold air, and cold water. At the time of the evaluation, the patient was treated with oral levocetirizine 5 mg daily and hydroxyzine 50 mg at bedtime without resolution of symptoms.
Results: In clinic, the patient had a positive ice cube test, a positive dermatographia test and a negative warm test tube test. Methacholine and autologous sweat testing were declined. Otherwise he had a normal physical examination with a negative Darier sign. Laboratory studies did not reveal a disease process responsible for the urticaria. Based upon his historical symptoms and clinical findings, he was diagnosed with 3 distinct types of physical urticaria; cholinergic urticaria, cold urticaria and dermatographia. The dose of anti-histamine therapy was doubled and the patient returned to clinic in 4 weeks to report that his symptoms were slightly improved but had not resolved.
Conclusions: Physical urticarias are usually controlled by antihistamine therapy but refractory cases are not uncommon. This patient also has poorly controlled asthma for which he is scheduled to start omalizumab therapy upon turning 12 in 1 month. We will continue to follow this case to observe if omalizumab has an effect upon his urticarial symptoms.

593 Chronic Urticaria as First Sign of Sarcoidosis

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