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The Role of Corticosteroid Therapy in Schnitzler Syndrome: A Case Report

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Abstract | **Case Report**

Background: Urticaria is a common inflammatory mucocutaneous syndrome with multiple etiologies. When recalcitrant to standard antihistamine therapy, a systematic diagnostic workup is essential to identify rare underlying systemic conditions such as Schnitzler Syndrome. ***Case Presentation:*** We report the case of a 41-year-old woman with a background of chronic asthma, fibromyalgia, degenerative retinopathy, and nasal polyposis, who presented with an 8-month history of recalcitrant urticarial eruption. Despite sequential trials of multiple antihistamine regimens including cetirizine, hydroxyzine, loratadine, levocetirizine, desloratadine, montelukast, and bilastine at standard and quadruple doses no clinical improvement was observed. Extensive etiological workup excluded common chronic urticaria, drug-induced, infectious, inducible, and autoimmune causes, as well as urticarial vasculitis, Adult-onset Still's disease, systemic lupus erythematosus, and Cryopyrin-Associated Periodic Syndromes (CAPS). Serum protein electrophoresis revealed a monoclonal peak in the gamma-globulin zone, confirmed by immunofixation as monoclonal IgG elevation, establishing the diagnosis of Schnitzler Syndrome. ***Treatment and Outcome:*** The patient was initiated on corticosteroid therapy with prednisolone 40 mg/day for 3 months, resulting in marked clinical improvement. ***Conclusion:*** This case highlights the importance of a thorough diagnostic approach in chronic recalcitrant urticaria. While antihistamines remain the first-line treatment, corticosteroid therapy may play a critical role in antihistamine-refractory forms, particularly in the context of Schnitzler Syndrome. Clinicians should consider monoclonal gammopathy screening in patients with persistent urticaria unresponsive to standard therapy.

Keywords: Schnitzler Syndrome, chronic urticaria, monoclonal gammopathy, corticosteroid therapy, antihistamines, case report.

INTRODUCTION

Urticaria is a common inflammatory mucocutaneous syndrome that manifests clinically as erythematous and edematous papules, often pruritic; the lesions are rather indurated and painful, transient, migratory, and may be accompanied by angioedema [1]. Histologically, superficial urticaria presents as edema localized to the dermis, while deep urticaria (angioedema) shows diffusion of edema toward the hypodermis. Positive diagnosis is based on history-taking and clinical examination, with multiple possible etiologies [1,2].

CASE REPORT

We report the case of a 41-year-old female patient with a medical history significant for chronic asthma under treatment (bronchodilators and corticosteroids during exacerbations), chronic inflammatory rheumatic disease (fibromyalgia) diagnosed and well-followed, treated with various therapeutic classes (level I and II analgesics, NSAIDs, antidepressants, currently discontinued) degenerative retinopathy followed in ophthalmology under treatment (anti-glaucoma agents), and a prior surgical history of nasal polyposis.

She was admitted for the management of a recalcitrant urticarial eruption evolving over 8 months. Clinical examination revealed urticarial lesions, white dermographism, and excoriation marks in the form of linear striae on the lower limbs, upper limbs, and sub-mammary region.

The patient was initially treated with several H1 antihistamines: cetirizine (Zadry 10 mg) 1 tablet/day and hydroxyzine (Atarax 25 mg) 1 tablet/day for 3 weeks with no improvement; then loratadine (Claritec 10 mg) 2

tablets/day for 2 weeks with no improvement; then quadruple-dose antihistamines levocetirizine (ICAM 5 mg) 2 tablets/day and desloratadine (Aerius 5 mg) 2 tablets/day for 2 weeks with no improvement; and finally montelukast 10 mg 1 tablet/day and bilastine (Bilaxten 20 mg) 1 tablet/day for 3 weeks with no improvement.

DISCUSSION

In view of the persistence of urticarial flares despite antihistamine treatment, a diagnostic and therapeutic workup was initiated. Common chronic urticaria was excluded, as no food trigger was identified, and total and specific IgE levels (trophic allergens and aeroallergens) were negative. The patient had a history of drug hypersensitivity to NSAIDs; however, the absence of concomitant NSAID use during urticarial episodes and the onset of urticaria after their discontinuation ruled out a drug-induced cause.

No infectious cause was identified: WBC, CRP, and ESR were normal; serologies for hepatitis B and C, syphilis, cytomegalovirus, and Epstein-Barr virus were negative; urine culture was sterile; stool parasitology and culture were normal. Inducible urticaria was also excluded: no mechanical urticaria (dermographism, delayed pressure, vibratory), thermal urticaria (cold, heat, cholinergic), aquagenic urticaria, or solar urticaria was noted [3].

Acquired histaminic and bradykinin-mediated angioedema was excluded: no respiratory symptoms were present, and functional and quantitative C1-esterase inhibitor levels were normal. Arguments in favor of systemic urticaria included the rheumatological and ocular involvement. Urticarial vasculitis was excluded, as the skin lesions were not fixed, and there was no purpura, fever, elevated ESR or CRP, nor hypocomplementemia.

Adult-onset Still's disease was excluded due to the absence of fever, associated arthritis, hyperleukocytosis, hepatic biological abnormalities, and antinuclear antibodies. Systemic lupus erythematosus was ruled out by a negative immunological workup.

Cryopyrin-Associated Periodic Syndromes (CAPS) characterized by recurrent urticarial eruptions, arthralgia or arthritis, ocular inflammation, fatigue, and headaches were considered, as our patient shared several features: urticaria with ocular and articular involvement. However, the absence of fever and biological inflammatory syndrome made this diagnosis unlikely [4].

Schnitzler Syndrome (recurrent urticarial eruption and monoclonal gammopathy) was strongly suspected: our patient presented with urticarial eruptions and fibromyalgia-type muscle pain. Serum protein electrophoresis (SPE) revealed a monoclonal-appearing peak migrating in the gamma-globulin zone, associated with a moderate decrease in alpha-2 globulins. This was confirmed by immunofixation, which demonstrated monoclonal IgG elevation. The patient was subsequently started on corticosteroid therapy (prednisolone 40 mg/day for 3 months) with marked clinical improvement [2,5].

CONCLUSION

Although antihistamines remain the first-line treatment for chronic urticaria, corticosteroid therapy appears to play a very important role in recalcitrant forms unresponsive to various antihistamine regimens.

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