

Urticaria: An Underrecognized Initial Manifestation to Systemic Complications

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Abstract

Urticaria is a common condition identified and treated in the primary care setting. It is a cutaneous vascular reaction characterized by locally increased vascular permeability. Clinically manifest as well circumscribed, intensely pruritic, raised wheals lesions that resolve within 8 to 24 hours without residual effects. Chronic urticaria, persist more than 24 hours and should raise suspicion for systemic disease, especially when urticarial vasculitis (UV) is present. UV is a small-vessel vasculitides, caused by leukocytoclastic vasculitis, where skin lesions last between 24 to 72 hours and heals leaving skin hyperpigmentation. Hypocomplementemic Urticarial vasculitis syndrome (HUVS) is a rare disorder characterized by urticaria that persist more than 24 hours or recur at short interval. It is associated with connective tissue diseases, such as systemic lupus erythematosus (SLE). HUSV is considered an independent immunological disease from SLE, or a subset of SLE with shared clinical, laboratory and immunologic features.

Keywords: Urticaria • Cutaneous vascular

Here, we present a case of 45 year-old Puerto Rican female with initial manifestation of urticarial-like flares in November 2017 after two consecutive episodes gallstone-induce acute pancreatitis. Urticarial rash developed initially on her arms and chest, slightly itching, which

lasted 48-72 hours. Characterized by resolution with residual skin hyperpigmentation, spontaneous recurrences, to eventually covered all of her body. Patient was evaluated by allergist and treated with steroids, high dose antihistamines and dapsone without improvement [1]. Patient was evaluated in February 2018 at rheumatology clinic. She was presenting with pruritic and painful urticaria, accompanied by constitutional symptoms and eye redness. Physical examination was significant for erythematous, palpable, non-blanching wheals; varying in size covering all her body; marked swelling of the hands, lower extremity edema, and decreased muscle strength through all extremities. Rheumatologic work up found hypocomplementemia; and negative antinuclear antibody [ANA], anti-double-stranded DNA, anti-Smith antibody, cytoplasmic anti-neutrophil cytoplasmic antibodies [C-ANCA], and perinuclear anti-neutrophil cytoplasmic antibodies [P-ANCA] [2]. Urinalysis with proteinuria and hematuria, and chemistry with normal renal function. Skin biopsy showed urticaria without vasculitis. Initial treatment consisted of prednisone, hydroxychloroquine, and azathioprine; with clinical resolution of urticaria, but urine examination with persistent hematuria and nephrotic range proteinuria. Renal biopsy findings showed Lupus Nephritis Class V membranous glomerulonephritis. Few months later patient developed anasarca which required inpatient care with intravenous diuretics and steroids; also clinical signs of systemic lupus erythematosus emerge including fatigue, oral ulcers, malar rash, arthralgias and photosensitivity. Cytotoxic agent infusion with cyclophosphamide was started due to persistent proteinuria and hematuria; with clinical improvement, decreasing of proteinuria and normalization of complement levels [3].

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