Acta Derm Venereol 2014; 94: 244–245

SHORT COMMUNICATION

Urticarial Vasculitis Associated with Essential Thrombocythaemia

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Accepted Apr 3, 2013; Epub ahead of print Aug 27, 2013

Urticarial vasculitis is a form of leucocytoclastic vasculitis whereby the skin lesions resemble urticaria. It is associated with systemic lupus erythematosus, Sjögren's syndrome, hepatitis B and C viruses (1). Rarely it is associated with an underlying haematological disorders and, to the best of our knowledge, has never been reported in association with essential thrombocythaemia. We present a case report of urticarial vasculitis associated with essential thrombocythaemia.

CASE REPORT

A 32-year-old woman presented with a several month history of recurrent urticaria without angioedema, 4 months post-partum. Most of the urticarial lesions persisted for > 24 h. She was otherwise fit and well with no systemic upset, and was not taking any prescribed or over the counter medication. On examination, she had widespread urticarial wheals and residual ecchymoses at the sites of old lesions (Fig. 1). A urine dipstick and blood pressure were normal. She was initially treated with loratadine 10 mg once daily without success. She was subsequently switched to fexofenadine 180 mg once daily, hydroxyzine 25 mg once daily and oral prednisolone 30 mg once daily, which controlled her symptoms.

A biopsy was taken from an uticarial lesion on the arm. Histology showed numerous perivascular neutrophils and eosinophils with margination of neutrophils in the lumen of vessels and some leucocytoclasis with red cell extravasation in keeping with an urticarial vasculitis (Fig. 2).

Blood tests revealed an erythrocyte sedimentation rate of 47 mm/h (normal range 1–12), a platelet count of $1{,}098 \times 10^9$/l (normal range 135–400) and an eosinophilia of 1.0×10^9/l (normal range 0–0.2). Complement levels, thyroid function, serum iron, haemoglobin, C-reactive protein, anti-nuclear antibody, anti-nuclear cytoplasmic antibody, antiphospholipid antibody and rheumatoid factor were normal or negative. There was no paraproteinaemia and syphilis and hepatitis serologies were negative.

The thrombocytosis was initially thought to be reactive but on further review the patient's platelet count was noted to be raised at 668×10^9/l, eight months prior to her presentation. A haematology opinion was sought.

The patient was negative for a JAK-2 mutation and subsequently had a bone marrow biopsy. This showed an increased number of megakaryocytes with focal

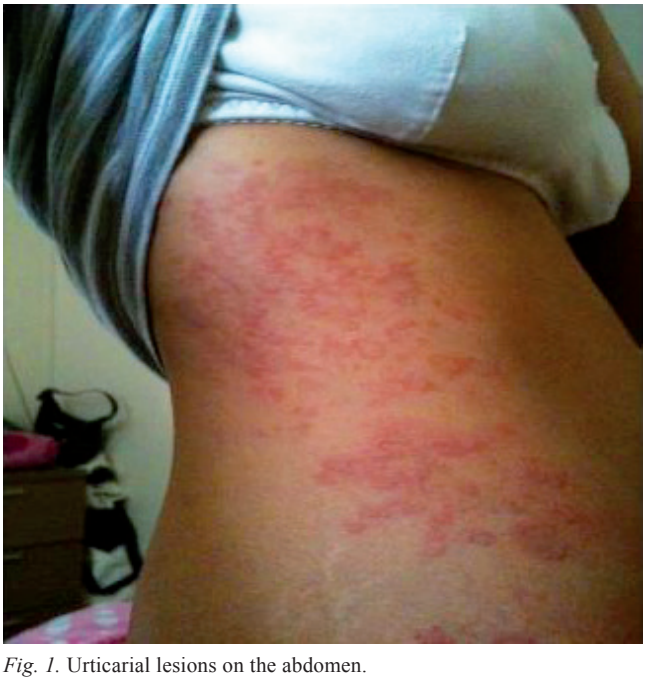

Fig. 1. Urticarial lesions on the abdomen.

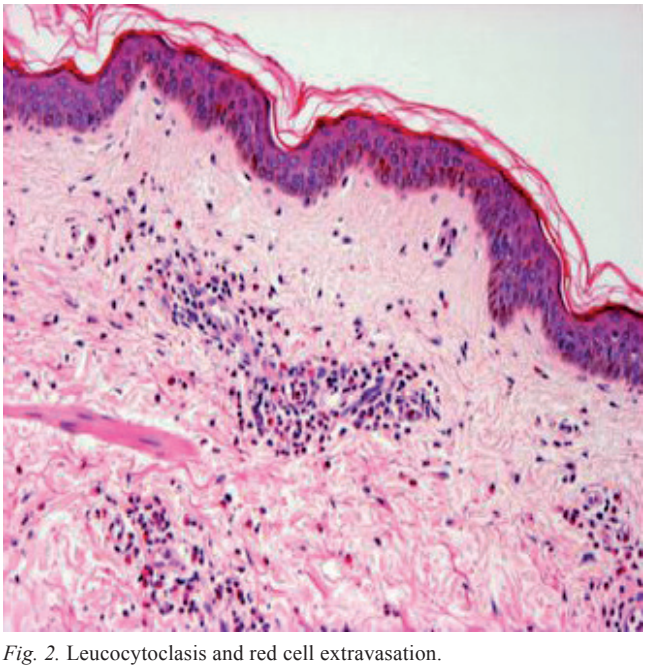

Fig. 2. Leucocytoclasis and red cell extravasation.

 doi: 10.2340/00015555-1653
 ISSN 0001-5555

clustering and large hyperlobulated and a few small hypolobulated forms. A diagnosis of essential thrombocythaemia was made given this finding in conjunction with a persistent thrombocytosis, normal serum iron and haemoglobin level. Low dose aspirin (75 mg once daily) was commenced to prevent thromboembolic complications. The patient was subsequently weaned off steroids and her symptoms remain well controlled for the past 6 months on antihistamines and aspirin.

DISCUSSION

Urticarial vasculitis is a clinicopathological entity where skin lesions resemble urticaria, but last longer than 24 h, and histology shows a leucocytoclastic vasculitis (1). Urticarial vasculitis comprises hypocomplementemic and normocomplementemic subtypes. Normocomplementemic urticarial vasculitis is associated with many conditions including hepatitis B, hepatitis C, lymphoma, infectious mononucleosis, Lyme disease, polycythaemia rubra vera, idiopathic thrombocytopenia purpura, Schnitzler's syndrome, Muckle-Wells syndrome and Cogan's syndrome. Urticarial vasculitis is thought to result from the formation of immune complexes in the blood that then deposit in vessel walls (2).

Hypocomplementemic urticarial vasculitis is strongly associated with female gender, systemic lupus erythematosus and the presence of anti-nuclear antibodies. Renal and pulmonary involvement is more commonly seen. Normocomplementemic urticarial vasculitis runs a more benign course, with less frequent systemic involvement and although associated with the presence of anti-nuclear antibodies, patients rarely fit the criteria for systemic lupus erythematosus (3).

Leucocytoclastic vasculitis is associated with lymphoproliferative disorders, but scarcely with essential thrombocythaemia. Suarez Conde et al. (2) reported a man who presented with leucocytoclastic vasculitic ulcers, which subsequently led to the diagnosis of essential thromobocythaemia. The vasculitis improved following treatment with hydroxyurea for essential thrombocythaemia.

Essential thrombocythaemia is a rare, chronic, myeloproliferative disorder characterised by an increase in the peripheral platelet count ($>450{,}000/mm^3$) in the absence of an underlying disorder (5). Complications include arterial and venous thromboembolism, and abnormal bleeding. Cutaneous manifestations associated with essential thrombocythaemia include ecchymosis, petechiae, purpura, erythromelalgia, Raynaud's phenomenon and livedo reticularis (4). Skin signs can precede and lead to the diagnosis of essential thrombocythaemia (4).

The treatment of essential thrombocythaemia is aimed at reducing symptoms and reducing the risk of thromboembolic complications. The most common symptoms are vasomotor and these are usually controlled with low dose aspirin. In those patients with a high risk of thromboembolic complications such as age >60 or a prior history of thromboembolic disease then cytoreductive therapy maybe indicated. Cytoreductive agents that are used include hydroxyurea, alpha interferon, pegylated interferon and anegrelide (6).

In one review, 58 of 268 (22%) patients with essential thrombocythaemia had an associated cutaneous manifestation (4). Two patients were found to have urticaria. The type of urticaria or whether it preceded the essential thrombocythaemia is not mentioned. Ecchymoses, haematomas, petechiae and purpura were the most common cutaneous manifestations seen; occurring in 24 (41%) patients (4).

To our knowledge, this is the first reported case of urticarial vasculitis associated with essential thrombocythaemia. In our patient the thrombocythaemia preceded the development of urticarial vasculitis, but it was the urticarial vasculitis that subsequently led to the diagnosis of essential thrombocythaemia. Essential thrombocythaemia should thus be excluded in patients who present with urticarial vasculitis.

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