Escherichia coli: an uncommon cause of severe urticarial vasculitis

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Sir,

Urticarial vasculitis is an uncommon clinicopathological entity with a reported prevalence of 5% wherein episodes of urticaria are accompanied by leukocytoclastic vasculitis that may be normo-complementemenetic or hypo-complementemenetic (low C1q and C4 levels, and variably decreased C3 levels) [1,2]. Bacteremia may present as leukocytoclastic vasculitis via septic emboli, immune complex injury or via bacterial seeding of vessels that causes necrosis through direct bacterial action. Escherichia coli, a rare cause of leukocytoclastic vasculitis, is not a well described cause of urticarial vasculitis among reported cases.

A 46-year-old male was hospitalized with widespread, intensely pruritic deep dusky red lesions, edema of hands/feet, painful left knee for 4 days and no fever. Historically, a week earlier he had developed an increasing painful scrotal swelling, fever and chills. He had coalescing, dusky red and ecchymotic lesions, few having dusky erythematous wheals and central pallor, widely distributed over trunk and extremities while sparing the palms, soles and scalp (Fig. 1). His left knee had mildly painful movements without swelling. Scrotal swelling was firm, tender, and non-transilluminant. He was put on intravenous ceftriaxone (1gm twice daily), oral doxycycline (100mg twice/d), and cetirizine 10mg/d with the provisional diagnosis of epididymo-orchitis and urticarial vasculitis. Except for leukocytosis (total leukocyte count 12,900/cmm), lymphopenia (18%) and monocytes (16%), his laboratory investigations including serum biochemistry, VDRL, HIV serology, smears from urethra and vesiculopustular lesions, chest and knees x-rays were essentially normal. Abdominal ultrasonography (USG) revealed mild hepatomegaly with prominent portal vein. Scrotal USG showed mild left hydrocele and swelling of skin overlying left testis. He became febrile (temperature 39°C) and developed fresh wheals with dusky erythema. Histology showed unremarkable epidermis, mild dermal edema and inflammatory cell infiltrate in papillary and upper dermis, and urticarial vasculitis. Except for leukocytosis (total leukocyte count 12,900/cmm), lymphopenia (18%) and monocytes (16%), his laboratory investigations including serum biochemistry, VDRL, HIV serology, smears from urethra and vesiculopustular lesions, chest and knees x-rays were essentially normal. Abdominal

Figure 1: (a) Deep red to brown colored, coalescing, ecchymotic lesions with characteristic dusky erythema and central pallor are involving the trunk. (b) Dusky erythematous, ecchymotic, coalescing lesions with central pallor are seen over forearms. Arrow indicates typical urticarial wheal while other lesions are in various stages of resolution.

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added after the urine culture showed *E. coli* sensitive to it. Pus culture sensitivity from surgical incision/drainage of scrotal swelling after it became fluctuant showed growth of *E. coli* sensitive to cefoperazone/sulbactum (administered intravenously 1gm twice/d for 7 days). Prednisolone was tapered off over next 2 weeks as new lesions stopped. The scrotal wound was closed by secondary suturing after another 2 weeks. No recurrences reported on follow up visit at 3 weeks.

Urticarial vasculitis mostly remains idiopathic or may occur secondary to connective tissue diseases (systemic lupus erythematosus, rheumatoid arthritis, Sjogren’s syndrome), serum sickness, neoplasia (leukemias, or breast, pituitary, thyroid, colon and pancreatic tumors), drugs (diltiazem, cimetidine, ACE inhibitors, antibiotics, interferon, NSAIDs, potassium iodide), and infections (hepatitis B, hepatitis C, HIV, syphilis, infectious mononucleosis, upper respiratory infections) [1,3]. Clinically, the wheals last for 48-72 hours, associated with burning, pain or tenderness, and foci of purpura and induration. Angioedema-like swelling of lips, tongue, eyelids, and hands is seen in 40% cases [2]. Resolution occurs usually with purpura or hyperpigmentation [1]. All these features distinguish urticarial vasculitis from true urticaria. Fever, malaise, myalgia, fatigue, and specific organ involvement (arthralgia, arthritis, serositis, glomerulonephritis, interstitial nephritis, Raynaud’s phenomenon) are common accompaniments especially in hypo-complementemic variety [2]. Conjunctivitis and episcleritis may also occur [4]. Although response to therapy is unpredictable, antihistamines or nonsteroidal drugs (ibuprofen, naproxen) may suffice in normo-complementemic or idiopathic cases. Patients with organopathy or more severe cases may need systemic corticosteroids, hydroxychloroquine, colchicine, dapsone, azathioprine or cyclophosphamide [4]. Complications such as skin ulcers or multiorgan damage (lungs, eyes, kidneys) occur often in secondary or hypo-complementemic variety [3]. Prognosis is generally good in normo-complementemic or idiopathic cases. Spontaneous recovery may occur but some cases require intermittent treatment lasting for several years. However, the overall prognosis is often dictated by the prognosis of the underlying disease. This patient was normo-complementemic and recovered completely without recurrence after *E. coli* infection was eradicated.

**CONSENT**

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure. The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article.

**REFERENCES**