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Normocomplementemic Urticarial Vasculitis in a Boy and his Response to Treatment.

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Abstract

Urticarial vasculitis is an uncommon from of cutaneous vasculitis and is extremely rare to present during childhood, especially in boys. It typically presents with multiple systemic inflammatory manifestations. Skin biopsy is needed to confirm the diagnosis. In this report a 10 years old Saudi boy with prolonged urticarial rash, fever, and arthritis is presented. His skin biopsy was consistent with leukocytoclastic vasculitis. His treatment course with several anti-inflammatory agents is described.

Keywords: Urticarial Vasculitis, Children, Dapsone, Leukocytoclasis

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Introduction

Urticarial vasculitis (UV) is a type of small blood vessel inflammation that presents with urticarial or urticarial like skin rash and is often associated with systemic manifestations. The diagnosis of UV is based on a group of clinicopathological features. Typically, patients with UV present with cutaneous lesions that last for > 24 hours in the same place and heal with residual faint hyperpigmentation [1]. The rash is usually itchy and is frequently associated with pain and burning sensation. The most common associated systemic feature is arthritis or arthralgia. Patients may also have fever, malaise, and myalgia. In addition, few patients were reported to have abdominal, renal, pulmonary, or other autoimmune manifestations [2].

UV is an uncommon disease. Only about 5% of those with chronic urticaria are diagnosed with UV [1].It typically presents during adulthood, and is extremely rare in children. Here, a child from Saudi Arabia who presented with UV at 10 years of age is reported and his treatment course is described.

Patient Report

A 10 years old Saudi boy presented with 6 weeks history of classical urticarial rash that started in his upper and lower limbs then spread all over his body. The urticarial lesions would last for 15 to 20 hours. They were itchy with burning and painful sensation. This was associated with fever up to 40°C, myalgia, and bilateral ankle pain and swelling that made him unable to walk for few days. He later developed polyarticular disease, presenting with pain in bilateral knees, hands, and feet. No other systemic manifestations. He was fully vaccinated. No upper respiratory tract infection preceded his illness. His family history was negative for similar problems. His parents were not consanguineous. He was in 4th grade at school and doing well. No recent travel.

Ophthalmologic and cardiovascular examination including echocardiography, were normal. His blood tests, while symptomatic, showed WBC count of 11.7 X 10^9 cells/L with neutrophilia of 8.7 X 10^9 cells/L, otherwise normal. ESR was 120 mm/hour. C3 and C4 were normal. Also, ASO titer, ANA, SSA, SSB, dsDNA, and rheumatoid factor were all negative. His Hepatitis A, B, and C screen was negative as well. His thyroid function test was normal. Bone marrow examination was normal. Skin biopsy from a lesion was consistent with, but not completely typical of UV (Figure 1).

He was treated initially with antihistamines up to twice a day of 10 mg loratidine, but only had partial response in terms of his urticaria. His arthritis responded to treatment with non-steroidal anti-inflammatory medications for few weeks only. He was then shifted to hydroxychloroquine 200 mg QD. However, after a week on this medication his skin rash as well as his arthritis, unexpectedly, got worse. His treatment was then changed to dapsone 50 mg QD. His symptoms faded within a week and he continued to be in complete remission for 2 months. He then stopped the medication, but unfortunately all his symptoms relapsed. He was restarted on dapsone with good response, but not as good as the first time. The dose was then. Dou-

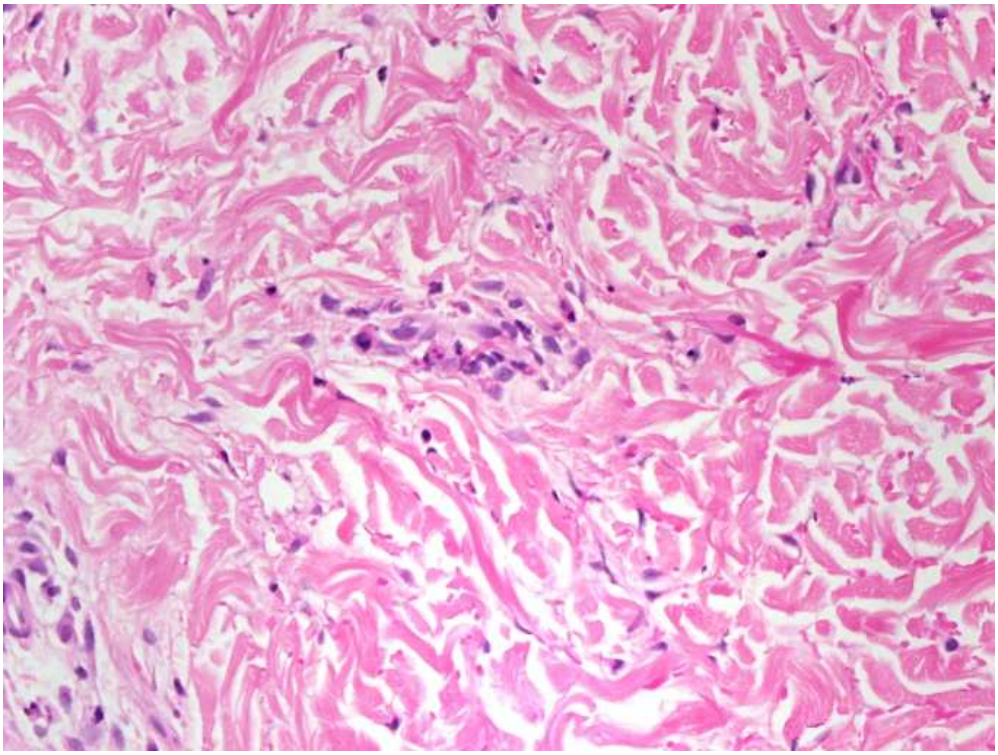

Figure 1. *Skin biopsy showing small dermal blood vessel (center) infiltrated by neutrophils with endothelial cell swelling, which indicates cell injury suggestive of UV. H&E stain. Magnification X 400*

bled and his condition improved to almost complete remission. He continued, however, to have breakthrough episodes of mild rashes but no arthralgia or fever. No ad verse effects were noted from dapsone treatment. His ESR normalized during periods of remission, but will increase again during breakthrough exacerbations

Discussion

The clinical and laboratory features as well as the skin biopsy of the boy in this report are consistent with UV. However, there are few atypical clinical features of this patient's presentation. UV is mainly an adulthood disease with an average age at presentation of around 40 years. This has been evident through several case series in the literature [3-9]. It is extremely rare for UV to present in childhood (<18 years of age). Only 7 children with UV were individually reported in the literature [10-16]. It is also more common in females. The cutaneous urticarial lesions typically last for >24 hours, but our patient's rash did not last that long. It also did not leave faint hyperpigmentation marks unlike many patients with UV. Nevertheless, all these variations in the clinical picture were reported in the literature [2, 9].

Skin biopsy is important to make the diagnosis, although not pathognomonic. The histological features of UV typically include neutrophilic perivascular infiltrate (leukocytoclasis) with some lymphocytic infiltrate, which may predominate in some cases [6], and occasional eosinophilic infiltrate. Also, endothelial cell swelling, red blood cell extravasation, and perivascular fibrinoid deposition is seen secondary to vessel wall injury [1, 2]. The biopsy from the reported patient showed features consistent with leukocytoclastic vasculitis commonly seen in patients with UV. However, it did not show some of the typical features of vasculitis like red blood cells extravasation or perivascular fibrinoid deposition. It has to be acknowledged here that detecting typical vasculitic changes in skin biopsy is not always easy [2]. This is because pathological changes are usually focal in nature and the picture also depends on the age of the lesions. Early lesions are more helpful in making the diagnosis and tend to show neutrophilic perivascular infiltrates, but mild vessel wall disruption. Older lesions, on the other hand, tend to show more lymphocytic infiltrates [4, 9]. Sometimes multiple biopsies are required to make the diagnosis.

Management of UV can be challenging. The subtype of UV can influence response to treatment. UV has been classified as either Normocomplementemic (NUV), which makes about 70-90% of all patients, or hypocomplementemic (HUV) with low C3, C4, or CH50. Patients with hypochomplementemic UV are much more likely to have systemic manifestations and some of them are eventually diagnosed with systemic lupus erythematosus [2]. The use of anti-histamines has lead to partial response in

most reports, mainly in relation to the control of itching and in ameliorating the rash. Steroids have been used with better success [1, 6]. Anti-neutrophilic medications particularly dapsone or colchicine have been used with even greater success, either alone or in combination with low dose steroids in both HUV and NUV [1, 2]. The response of our patient to dapsone supports a diagnosis of autoimmune vasculitis, especially UV. Resistant cases have been managed with different immune modulators, as in other autoimmune vasculitis conditions, with variable success. These included hydroxychloroquine, cyclosporine, mycophenolate mofetill, cyclophosphamide, methotrexate, or azathioprine. In more resistant cases, biologics like intravenous immunoglobulins, rituximab, or anakinra have been tried [1, 2, 9].

In conclusion, this report adds to the pediatrics literature our experience of such an extremely rare condition and its management.

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