Adult onset Henoch-Schönlein Purpura, treated with Colchicine

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INTRODUCTION

Henoch-Schönlein Purpura (HSP) is a systemic, IgA mediated autoimmune small vessel vasculitis (IgAV) (1,2,3). Clinical manifestations include nonthrombocytopenic palpable purpura, arthritis, abdominal pain and nephritis (2). The etiology remains unclear but is associated with bacterial infections, viral infections, medications (quinolones, clarithromycin, acetaminophen, codeine and TNF alpha inhibitors), tumors, alpha-1-antitrypsin deficiency and Familial Mediterranean Fever (3). Incidence is highest in patients 4-6 years old, however, up to 15% of cases occur in the adult population (1, 4). In adults, HSP tends to have more severe onset with a higher incidence of long term sequelae, in contrast to the more benign course that often occurs in the pediatric population (4). Regardless of age of onset there is a slight male predominance (M:F ≈ 2:1; 1.0) and Caucasian populations have the highest incidence (1).

While adult onset HSP has been reported previously, it is diagnosed infrequently. Our case brings attention to the possible development of HSP in individuals utilizing TNF-alpha inhibitors and to the benefit of using colchicine as a treatment modality during the acute presentation of HSP.

CASE REPORT

50-year-old Caucasian male developed palpable purpura on his bilateral lower extremities twenty-four hours after arthroplasty assault in Hawaii. The patient was treated for presumed infection, with Minocycline then Doxycycline, without clinical improvement. Six days after the onset of the lesions, the patient returned from vacation and was evaluated by his dermatologist who obtained 2 punch biopsies (H&E and DIF) and a bacterial culture. At dermatologic follow-up three days later he had developed hemorrhagic bullae and plaques with central necrosis. Bacterial culture had grown group B strep. The patient was prescribed Clindamycin and advised to go to ED. One day later the patient developed arthralgia and edema in his bilateral wrists and hands but denied hematuria or abdominal pain. The following day the patient noted worsening joint pain as well as new onset abdominal pain. Labs included: +ASO, +Hematuria (UA), elevated serum IgA, CBC WNL, compliment WNL. At this point patient met EULAR criteria for Adult HSP and Colchicine 0.6mg daily was initiated. The following day the patient reported significant improvement in joint pain and moderate improvement in abdominal pain. Cutaneous lesions improved significantly over the following month. Biopsy results returned: H&E LCV, DIF, IgA vasculitis.

PMHI: Psoriasis (Adalimumab), HTN (Lisinopril)

CLINICAL & HISTOLOGIC FINDINGS

The clinical appearance and disease progression of HSP are not the same in adults and children. Virtually all patients with HSP have palpable purpura, however adults are more likely to present with hemorrhagic lesions, more severe arthritis and have more frequent renal involvement (4). The majority of patients will develop urinary abnormalities within 4 weeks of diagnosis but, it can take months for HNP nephritis to develop (4). Patients with a diagnosis of HSP should be followed with urinalysis for one year (1).

Recent research suggests that adult onset HSP is likely underdiagnosed, in part, due to ACR criteria for IgAV including age at onset of under 20 years. Previously unstudied in the adult population, the EULAR/PRINTO/PRES IgAV classification criteria has been shown by one study to have a higher sensitivity and specificity than the ACR criteria when evaluating adult patients for HSP (5). While etiology is often unclear, infections including streptococcal pharyngitis, medications, malignancy, alpha-1-antitrypsin deficiency and Familial Mediterranean Fever have been implicated in the development of HSP (1). Recently, several cases of HSP developing in patients on TNF alpha inhibitors have been reported (6). In our case, with elevated ASO titers and use of Adalimumab, it is unclear what exactly initiated the HSP cascade.

Corticosteroids are often the chosen treatment for HSP. Given our patients history of psoriasis we avoided steroids due to the risk of psoriatic flare upon taper. We elected to use low does Colchicine 0.6mg daily due to its reported success in treating cutaneous vasculitis (7). The patient’s joint pain and cutaneous findings rapidly improved further supporting the use of this treatment modality in acute episodes of HSP.

CONCLUSION

Adult onset HSP is an uncommon diagnosis that differs in clinical appearance and long-term management from HSP in the pediatric population. This case demonstrates the importance of considering HSP as a diagnosis for adult patients presenting with small vessel vasculitis, and further supports the use of Colchicine as a treatment for patients with HSP.

REFERENCES