



Case Report

An interesting case of SLE mimicking IgA Vasculitis

Authors

Ambroise MJ^{1*}, Varsha PR², Premkumar G³, Viswanathan KN⁴

¹Junior Resident, Department of General Medicine, Sri Manakula Vinayagar Medical College and Hospital, Puducherry, India

²Junior Resident, Department of General Medicine, Sri Manakula Vinayagar Medical College and Hospital, Puducherry, India

³Associate Professor, Department of General Medicine, Sri Manakula Vinayagar Medical College and Hospital, Puducherry, India

⁴Professor, Department of General Medicine, Sri Manakula Vinayagar Medical College and Hospital, Puducherry, India

*Corresponding Author

Dr Marie Jeffrey Ambroise

Junior Resident, Department of General Medicine, SMVMCH, Puducherry – 605107, India

Abstract

Leukocytoclastic vasculitis is a small vessel vasculitis which is often secondary to drugs, infections, connective tissue disorders or malignancy. It shares similarities with other vasculitis/ connective tissue disorders such as IgA vasculitis and systemic lupus erythematosus. Careful clinical analysis is warranted to differentiate this entity from other overlying disorders. Here, we discuss a case of a young adult female who presented to us with bilateral lower limb purpura, which proved to be a diagnostic quandary for the treating physicians.

Keywords: Systemic lupus erythematosus, Leukocytoclastic vasculitis, IgA Vasculitis.

Introduction

Leukocytoclastic vasculitis is an immune complex mediated small vessel vasculitis of the dermal capillaries and venules. The skin, gastrointestinal system and kidneys are the primary organs affected, presenting with a classic clinical presentation of arthritis, purpura (without thrombocytopenia), abdominal pain and renal failure. If leukocytoclastic vasculitis is suspected, a punch biopsy should be performed with direct immunofluorescence studies. If no systemic symptoms are present, laboratory testing including C-reactive protein, complete blood count (CBC),

basic metabolic panel, liver function tests, and urinalysis should be done as well. If there is a concern for systemic involvement, a more extensive workup needs to be performed.

Case

An 18 year old female presented with rash over both lower limbs of three weeks duration associated with fever, malaise and arthralgia. She denied history of abdominal pain, hematuria, recent upper respiratory infection, chest discomfort, breathing difficulty, early morning stiffness of joints or drug intake. Examination

revealed purpuric rash involving both lower limbs from the dorsum of foot to thigh, which were non-palpable and non-blanchable on pressure, with no truncal involvement (fig 1a). There was no mucosal involvement. She also had pallor and bilateral pitting pedal edema upto knee. A dermatology consult was requested and a punch biopsy of the lesion was advised. The tissue sample was obtained and histopathology revealed perivascular neutrophilic infiltration with fibrinoid necrosis suggestive of leukocytoclastic vasculitis (LCV). A provisional diagnosis of IgA vasculitis was suggested by the dermatologist based on the EULAR clinical criteria which was satisfied by bilateral lower limb purpura and histological evidence of LCV. However, some clinical and biochemical parameters correlated negatively with the above diagnosis. Firstly, the age of the patient did not correspond with the average age of presentation of IgA vasculitis. Secondly, on day 2 of admission patient developed an evanescent rash over the palmar aspect of both hands which

disappeared within one day (fig 2b). On day 3 of admission, patient complained of redness and irritation of her right eye, which on examination, was revealed to be episcleritis (fig 2a). Lastly, hemogram revealed pancytopenia with an elevated ESR and urine analysis revealed microscopic hematuria and albuminuria. A 24 hour urinary protein assessment showed a biochemically significant level of 585 mg/dL. This cluster of findings prompted us to consider an alternative diagnosis to the patient's presentation. Hence, a vasculitis workup was initiated. Surprisingly, the ANA and Anti-ds DNA turned out positive with low levels of serum C3 and C4. Hence, a diagnosis of leukocytoclastic vasculitis secondary to systemic lupus erythematosus was made. The patient was started on oral prednisolone and a prompt clinical response - disappearance of rash was seen within 72 hours of treatment (fig 1b). Furthermore, on follow-up, her hemogram showed improvement of her pancytopenia and decrease in urine RBCs and albumin.



Figure 1a and 1b



Figure 2a and 2b

Discussion

Leukocytoclastic vasculitis (LCV) is an immune complex mediated small vessel vasculitis of the dermal capillaries. The annual incidence of biopsy-proven leukocytoclastic vasculitis is approximately 45 per million individuals.¹ While LCV occurs in both genders and is common in all age groups, IgA vasculitis is more common in children of 5 – 15 years, with a slight male preponderance. LCV presents as erythematous macules with palpable purpura bilaterally on dependent areas of the body like the lower extremities and buttocks, with lesions appearing as crops, varying from 1 mm to 1 cm in diameter. Other possible cutaneous manifestations include hemorrhagic vesicles, bulla, pustules, nodules, crusted ulcers, and livedo reticularis. In our case,

we noticed development of purpura in both upper limbs also. Classical IgA vasculitis is symmetrical in distribution involving dependent areas such as the lower extremities and buttocks but it can also be seen in the upper extremities, while truncal and facial involvement are not uncommon. Our patient also had episcleritis of her right eye. Episcleritis is a common manifestation of systemic vasculitides, most frequently associated with rheumatoid arthritis. It can also be seen in other connective tissue disorders such as systemic lupus erythematosus, crohn's disease, ulcerative colitis, psoriatic arthritis, reactive arthritis, relapsing polychondritis, ankylosing spondylitis, polyarteritis nodosa, Behcet disease, Cogan syndrome, and infections like lyme disease, tuberculosis, syphilis, and herpes zoster.²

However, the association of episcleritis with IgA vasculitis is seldom reported in literature. Systemic symptoms noted with leukocytoclastic vasculitis may include low-grade fevers, malaise, weight loss, myalgia and arthralgia. These findings have been noted in approximately 30% of affected patients, with arthralgia being the most common manifestation. The findings of a greater than upper age limit at time of presentation, involvement of upper extremities and development of episcleritis lead to the initial diagnostic conundrum between the physician and the dermatologist. Furthermore, while IgA vasculitis is known to have associated renal involvement, its predilection to cause palpable purpura occurs almost always in the absence of thrombocytopenia.³ However, our patient's hemogram demonstrated pancytopenia, while urinalysis revealed significant proteinuria and hematuria. This prompted further workup in view of co-existing vasculitis/ connective tissue disorders which ended with identification of SLE as the culprit behind this presentation. Once the finding of LCV is confirmed histologically, the physician's work has only just begun. A thorough review of systems and physical exam should be followed by a targeted work-up to identify possible underlying causes and rule out systemic disease. In about half of patients, an underlying cause can be found: infection (15-20%), inflammatory disease (15-20%), drug (10-15%), and malignancy (<5%), with the remaining 45-55% are idiopathic.¹ Direct immunofluorescence is strongly recommended in cases of new-onset LCV. While a negative result carries a low yield, a positive result can sometimes be diagnostic of an underlying disease and can provide insight into the underlying disease pathophysiology. Strong IgA deposition without other antibody deposition is indicative of IgA vasculitis. LCV associated with underlying systemic lupus erythematosus can show diffusely positive immunofluorescence in addition to increased dermal mucin. Most cases of idiopathic LCV are mild and resolve with

supportive measures such as leg elevation, rest, compression stockings, and antihistamines. In more chronic or resistant cases, a 4-6 week tapering dose of corticosteroids can be used. Rarely, immunosuppressive steroid-sparing agents such as methotrexate, azathioprine, mycophenolate mofetil, dapsone, cyclophosphamide, and intravenous immunoglobulin may be needed. When leukocytoclastic vasculitis is associated with an underlying connective tissue disease or an autoimmune disease such as systemic lupus erythematosus or rheumatoid arthritis, better control of the underlying disease by an escalation of the immunosuppressive therapy may be needed to treat the leukocytoclastic vasculitis and prevent relapses. Mild cases of HSP usually do not require corticosteroids or immunosuppressive agents, and skin rash and arthralgia usually respond to NSAIDs. High-dose corticosteroids and/or immunosuppressive agents may be indicated in cases of severe renal involvement or systemic involvement.

Reference

1. Baigrie D, Bansal P, Goyal A, et al. Leukocytoclastic Vasculitis. [Updated 2020 Aug 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK482159/>.
2. Schonberg S, Stokkermans TJ. Episcleritis. [Updated 2020 Aug 10]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK534796/>
3. Sohagia AB, Gunturu SG, Tong TR, Herten HI. Henoch-schonlein purpura-a case report and review of the literature. *Gastroenterol Res Pract*. 2010;2010:597648. doi:10.1155/2010/597648