Case report

Leukocytoclastic Vasculitis

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Abstract:
Leukocytoclastic vasculitis (LCV), also known as necrotizing venulitis, hypersensitivity vasculitis or hypersensitivity angiitis is characterized by necrotizing inflammation around small dermal blood vessels, composed mainly of neutrophils and their debris. LCV is an uncommon condition, with a reported incidence rate of about 30 cases per million people per year. LCV usually presents as a rash on bilateral lower extremities which is usually a palpable, pruritic rash though any part of the body including internal organs may be affected.

In this article we present a case of vasculitis caused by taking over the counter medication containing Ibuprofen which improved after discontinuing the offending agent and treatment with steroids.

Keywords-Leukocytoclastic vasculitis, purpuric lesion, macular rash

Case Report
A 43-year-old male patient presented with painful palpable purpuric lesion on bilateral lower extremities and macular rash on his upper extremities bilaterally. Some lesions on his lower extremity were infected and had purulent discharge from it. Patient gave a history of upper respiratory tract infection a week prior, for which he took OTC medication containing Ibuprofen for a week, after which he developed these symptoms. Patient didn’t report any other significant past, family, personal or treatment history.

On physical examination he was afebrile with a pulse of 91bpm, Blood pressure of 150/80 mm Hg, respiratory rate of 12 breaths per minute, and SpO2 of 95% on room air. On respiratory examination chest was clear with normal vesicular sound bilaterally in all lung fields, all other system except for skin was unremarkable.

On local examination, we examined papular lesion present bilaterally on lower extremities and bilateral macular lesion on upper extremity. There was no peripheral oedema. A complete blood count revealed a white-cell count of 8,800/mm, Hemoglobin of 13.3 g/dl, and platelet count of 347,000 per mm³. A basal metabolic panel was unremarkable. On further investigation, he had a serum ANA titer of (1:80, Normal <1:20) and ESR- 56 mm/hr. (Normal range- < 20mm/hr.). Hepatitis B surface antigen and
Hepatitis C antibody were non-reactive. Urine analysis was normal.
The patient was diagnosed to have hypersensitivity vasculitis and was started on tab Prednisone 40mg for one week and then tapered over 2-3 weeks along with local treatment with Clindamycin. All OTC medications that the patient had been on were discontinued.

Discussion:
LCV is an inflammation of small blood vessels especially of the skin and it is characterized by collection of neutrophils in vascular wall leading to their damage. When IgA is the dominant immunoglobulin in immune complexes, systemic involvement is likely in both children and adults (Henoch-Schönlein purpura–HSP). LCV due to IgG- or IgM-containing immune complexes has less systemic involvement and a better prognosis than HSP. Other forms of LcV include cryoglobulinaemic, urticarial and ANCA-associated LcV (are these written as LcV not LCV??) as well as LcV associated with vasculopathy and coagulopathy in SCLE/SLE or in bacteremia/sepsis. Most cases of LCV are idiopathic with etiological triggers not being identified in as many as half of the cases.2

The potential other known causes are-

1. Medications- most commonly associated drugs are Antibiotics (Penicillin, Sulfonamides), NSAIDs and Diuretics. Drug-induced etiology (such as penicillin, sulfa, allopurinol, thiazides, Propylthiouracil, quinolones, retinoids) are implicated approximately in 10–24% of cases of LCV.2

2. Infections- Viral infection like Hepatitis C and HIV; bacterial infection like upper respiratory tract infections particularly with beta hemolytic streptococci

3. Collagen vascular diseases- SLE, rheumatoid arthritis, Sjogren syndrome.

4. LCV can present as a paraneoplastic syndrome particularly in older patients with constitutional symptoms and in those with anemia and immature cells on peripheral smears.4

LCV is usually idiopathic, but can also be secondary to etiological factors such as drugs, infections (like hepatitis B or C, HIV infection), chronic infection with bacteria, inflammatory bowel disease, collagen vascular disorder like rheumatoid arthritis, lupus erythematosus, Sjögren syndrome, Bechet disease, Wegner’s granulomatosis or it can rarely be associated with malignancy (like lymphomas, leukemia, squamous cell carcinoma of the lung). It may be confined to skin or it may be associated with systemic involvement mainly involving the joints, the gastrointestinal tract and the kidneys.

Usual presenting complaint of patients is cutaneous eruptions which may be asymptomatic, painful, itching or burning. On detailed history, the patient may get information of exposure to above mentioned or other potential medications, an episode of URTI or other infections or history of collagen vascular disorder and malignancy.

Skin biopsy reveals polymorphonuclear neutrophilic infiltration in and around blood vessel walls which leads to formation of leukocytoclasia. If it is drug-induced, then eosinophil infiltrate is also seen. The sample is taken from the most recent lesion, preferably within 24-48 hours of its formations, timing is important, since sample taken too early or
late may not reveal any diagnostic findings. However if the cause of LCV is relatively clear like a recent infection or new medication skin biopsy can be deferred if improvement in skin lesions is noted, as in our patient. Further investigation of underlying causes of LCV depends on the clinical presentation and can include serologic studies, such as antinuclear antibody, ANCA (cytoplasmic ANCA [cANCA], perinuclear ANCA [pANCA], atypical ANCA), and rheumatoid factor, serum protein electrophoresis with immunofixation to assess for paraproteinemia, cryoglobulins, and hepatitis C antibody, evaluation for underlying endocarditis or paraneoplastic syndromes among others.

In a presentation similar to our patient, it is difficult to determine whether it was the infection or the drug which caused LCV. However, many OTC cold medications containing ibuprofen can potentially lead to LCV highlighting the importance of judicious use of medications. The treatment depends on the etiology. If drug-induced withdrawal of offending drug is sufficient. But when internal organs are involved or when patient present with recurrent episodes steroids play a significant role in management. We decided to treat our patient with a short course of steroids due to extensive skin involvement and the patient was followed up in an outpatient setting till improvement of skin lesions.

References:

