CASE IN POINT

Leukocytoclastic Vasculitis Masquerading as Chronic Idiopathic Thrombocytopenic Purpura

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Background: Idiopathic thrombocytopenic purpura and leukocytoclastic vasculitis can present in a similar fashion and can be very hard to differentiate clinically without a biopsy. This can cause diagnostic dilemma and delay in management. A thorough evaluation is recommended to determine etiology, although about half are idiopathic.

Case Presentation: A patient aged 79 years with long-standing thrombocytopenia secondary to chronic idiopathic thrombocytopenic purpura presented with a rash. Although it was thought to be secondary to idiopathic thrombocytopenic purpura, a biopsy revealed presence of leukocytoclastic vasculitis.

Conclusions: Although most leukocytoclastic vasculitis cases are mild and resolve without intervention, many go undiagnosed due to biopsy delays. Health care professionals should determine and treat the underlying cause.

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Idiopathic thrombocytopenic purpura is an immune-mediated acquired condition affecting both adults and children.[1] Acute idiopathic thrombocytopenic purpura is the most common form, which happens in the presence of a precipitant, leading to a drop in platelet counts. However, chronic idiopathic thrombocytopenic purpura can occur when all the causes that might precipitate thrombocytopenia have been ruled out, and it is persistent for ≥ 12 months.[2] Its presence can mask other diseases that exhibit somewhat similar signs and symptoms. We present a case of a patient presenting with chronic idiopathic thrombocytopenic purpura with diffuse rash and was later diagnosed with idiopathic leukocytoclastic vasculitis.

CASE PRESENTATION

A 79-year-old woman presented to the hospital with 2-day history of a rash. The rash was purpureal and petechial and located on the trunk and bilateral upper and lower extremities. The rash was associated with itchiness and pain in the wrists, ankles, and small joints of the hands. The patient reported no changes in medication or diet, no recent upper respiratory tract or gastrointestinal infections, fever or chills, night sweats, or weight loss. The patient's medical history consisted of thrombocytopenia about 5 years before and since then had been following up with a hematologist and underwent an extensive workup, including bone marrow biopsy without a definite diagnosis.

The patient mentioned that at the time of diagnosis the platelet count was about 90,000 but had been fluctuating between 50 and 60,000 recently. The patient also reported no history of gum bleeding, nosebleeds, hemoptysis, hematemesis, or any miscarriages. She also had difficulty voiding for 2 to 3 days but no dysuria, frequency, urgency, or incontinence.

The patient was diagnosed with a urinary tract infection 1 day before presentation and was started on ciprofloxacin 500 mg daily for 5 days. Her home medications included diphenhydramine as needed, metoprolol, and levothyroxine 125 µg. Her medical history was significant for hypertension, bradycardia with pacemaker placement, and obstructive sleep apnea. There were no noteworthy elements in her family and social history.

Laboratory results were significant for 57,000/µL platelet count (reference range, 150,000-450,000), elevated d-dimer (6.07), and < 6 mg/dL C4 (reference range, 88-201). Hemoglobin level, coagulation panel, hemolytic panel, and fibrinogen level results were unremarkable. The hepatitis panel, Lyme disease, and HIV test were negative. The peripheral blood smear showed moderate thrombocytopenia, mild monocytosis, and borderline normochromic normocytic anemia without schistocytes. The autoimmune panel to evaluate

thrombocytopenia showed platelet antibody against glycoprotein IIb/IIIa, glycoprotein Ib/Ix, and glycoprotein Ia/IIa, suggestive toward a diagnosis of chronic idiopathic thrombocytopenic purpura. However, the skin biopsy of the rash was indicative of leukocytoclastic vasculitis.

An autoimmune panel for vasculitis, including antinuclear antibody and antidouble-stranded DNA, was negative. While in the hospital, the patient completed the course of ciprofloxacin for the urinary tract infection, the rash started to fade without any intervention, and the platelet count improved to 69,000/µL. The patient was discharged after 3 days with the recommendation to follow up with her hematologist.

DISCUSSION

Leukocytoclastic vasculitis is a small vessel vasculitis of the dermal capillaries and venules. Histologically, leukocytoclastic vasculitis is characterized by fibrinoid necrosis of the vessel wall with frequent neutrophils, nuclear dust, and extravasated erythrocytes.[3]

Although a thorough evaluation is recommended to determine etiology, about 50% of cases are idiopathic. The most common precipitants are acute infection or a new medication. Postinfectious leukocytoclastic vasculitis is most commonly seen after streptococcal upper respiratory tract infection. Among other infectious triggers, *Mycobacterium*, *Staphylococcus aureus*, chlamydia, Neisseria, HIV, hepatitis B, hepatitis C, and syphilis are noteworthy. Foods, autoimmune disease, collagen vascular disease, and malignancy are also associated with leukocytoclastic vasculitis.[4]

In our patient we could not find any specific identifiable triggers. However, the presence of a urinary tract infection as a precipitating factor cannot be ruled out.[5] Moreover, the patient received ciprofloxacin and there have been several case reports of leukocytoclastic vasculitis associated with use of a fluroquinolone.[6] Nevertheless, in the presence of chronic idiopathic thrombocytopenic purpura, which also is an auto-immune condition, an idiopathic cause seemed a reasonable explanation for the patient's etiopathogenesis.

The cutaneous manifestations of leukocytoclastic vasculitis may appear about 1 to 3 weeks after the triggering event if present. The major clinical findings include palpable purpura and/or petechiae that are nonblanching. These findings can easily be confused with other diagnoses especially in the presence of a similar preexisting diagnosis. For example, our patient already had chronic idiopathic thrombocytopenic purpura, and in such circumstances, a diagnosis of superimposed leukocytoclastic vasculitis can be easily missed without a thorough investigation. Extracutaneous manifestations with leukocytoclastic vasculitis are less common. Systemic symptoms may include low-grade fevers, malaise, weight loss, myalgia, and arthralgia. These findings have been noted in about 30% of affected patients, with arthralgia the most common manifestation.[7] Our patient also presented with pain involving multiple joints.

The mainstay of diagnosis for leukocytoclastic vasculitis is a skin biopsy with direct immunofluorescence. However, a workup for an underlying condition should be considered based on clinical suspicion. If a secondary cause is found, management should target treating the underlying cause, including withdrawal of the offending drug, treatment or control of the underlying infection, malignancy, or connective tissue disease. Most cases of idiopathic cutaneous leukocytoclastic vasculitis resolve with supportive measures, including leg elevation, rest, compression stockings, and antihistamines. In resistant cases, a 4- to 6-week tapering dose of corticosteroids and immunosuppressive steroid-sparing agents may be needed.[8]

CONCLUSIONS

Although most cases of leukocytoclastic vasculitis are mild and resolve without intervention, many cases go undiagnosed due to a delay in performing a biopsy. However, we should always look for the root cause of a patient's condition to rule out underlying contributing conditions. Differentiating leukocytoclastic vasculitis from any other preexisting condition presenting similarly is important.

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Ethics and consent

No informed consent was obtained from the patient; patient identifiers were removed to protect the patient's identity.

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