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Idiopathic mixed small and medium vessel cutaneous vasculitis: A case report

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Vasculitis is a rare, inflammatory condition of the blood vessels whereby excess leukocytes within the vessel leads to a loss of structural integrity, and possible destruction. This family of disorders can lead to varying degrees of organ and skin damage. Vasculitis may be due to primary disease or secondary due to an underlying disorder, drug reaction, or infection. In a large number of cases cutaneous vasculitis may present as an idiopathic condition and affect both small and medium sized vessels. We present a case of small and medium-vessel vasculitis on the lower extremity with cutaneous manifestations, without an identifiable cause.

Key Words: Vasculitis, leukocytoclastic cutaneous vasculitis, polyarteritis nodosa

Vasculitis is an inflammatory condition of the blood vessels whereby excess leukocytes within the vessel leads to a loss of structural integrity, and possible destruction. Cutaneous vasculitis may present with varying clinical manifestations and may be caused by systemic disease or secondarily due to an underlying disorder, drug reaction, or infection [1-2]. While numerous etiologies have been reported, a number of cases are idiopathic. Clinical, histopathologic, and laboratory evaluation are imperative to appropriately diagnose cutaneous vasculitis. Once confirmed though biopsy, treatment should begin with symptomatic management and treatment of any underlying disorder or causative agent. In more severe cases immunomodulatory medications or steroids.

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Case Report

A 64-year old male with a medical history significant for hypertension and hypercholesterolemia presented with a three-week history of multiple painful lesions to his bilateral lower extremities. Physical examination revealed several non-blanchable, erythematous lesions with hemorrhagic vesicles, areas of necrosis, and palpable purpura. Initial treatment by his primary care physician consisted of mupirocin and methylprednisolone with continued worsening of lesions. The patient reported that he had recently begun taking HCTZ, simvastatin, and metoprolol, but the lesions had started before the onset of these medications.

An extensive serologic evaluation was completed which included rheumatoid factor, antinuclear antibody, anti-DS DNA, cryoglobulin, complement levels, hepatitis panel, HIV and various other studies. All laboratory studies were unremarkable. A chest x-ray was performed which was negative. The patient also underwent evaluation by numerous other specialists including rheumatology, gastroenterology, and vascular surgery.

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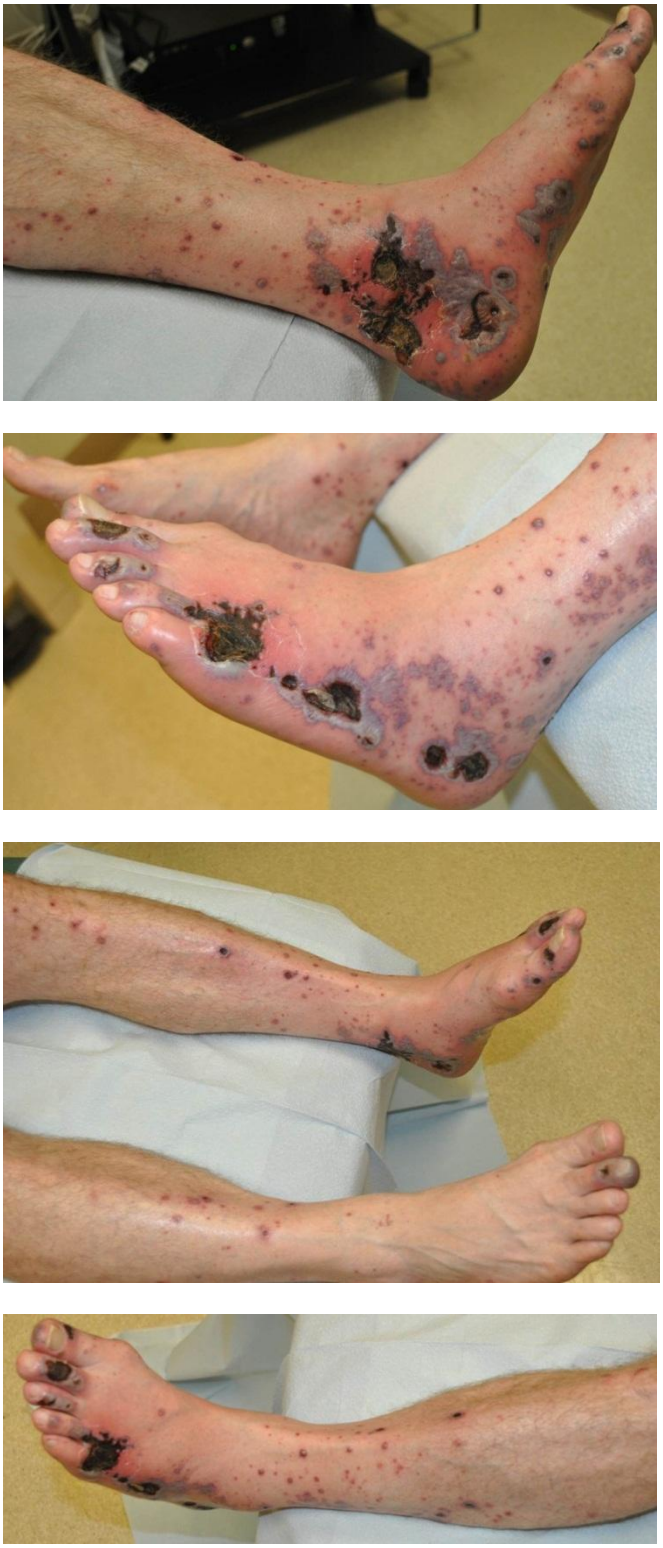


Figure 1-4 Clinical appearance of lower extremities at initial presentation.

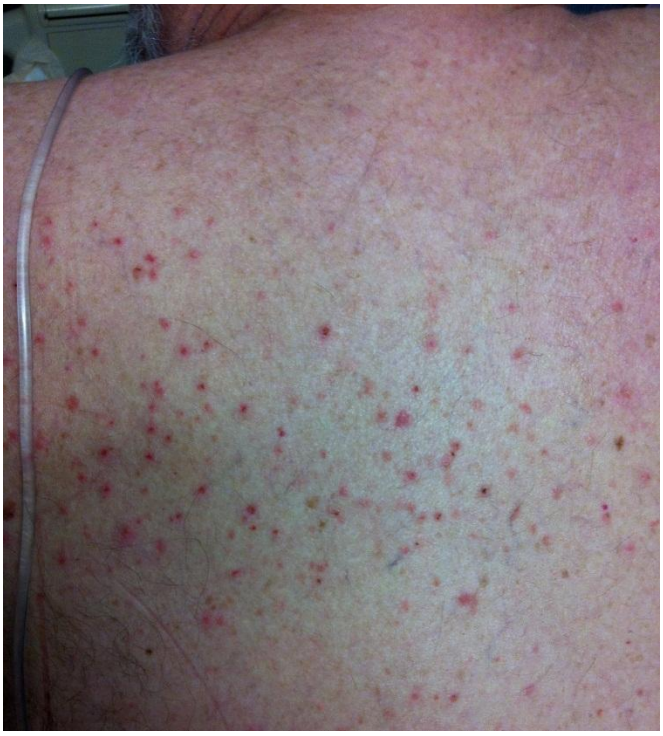


Figure 5 Patients back at initial presentation.

Biopsies were obtained and sent for histopathologic examination and immunofluorescence. Histopathology confirmed cutaneous leukocytoclastic vasculitis. Direct immunofluorescence revealed medium vessel disease.

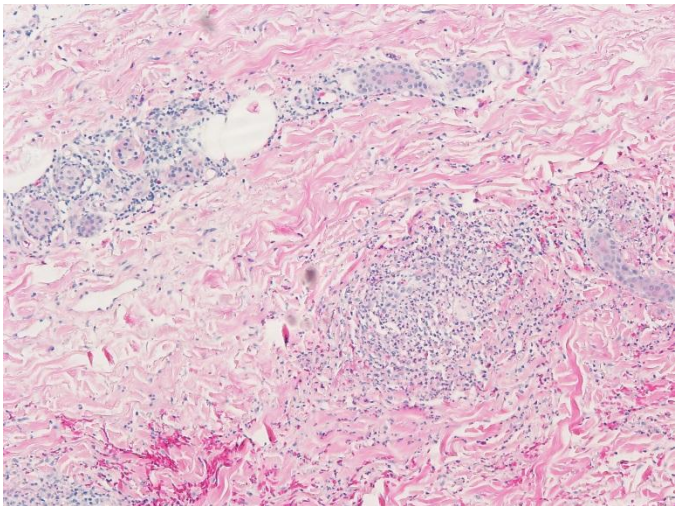


Figure 6 Histopathology Slides revealing leukocytoclastic vasculitis.

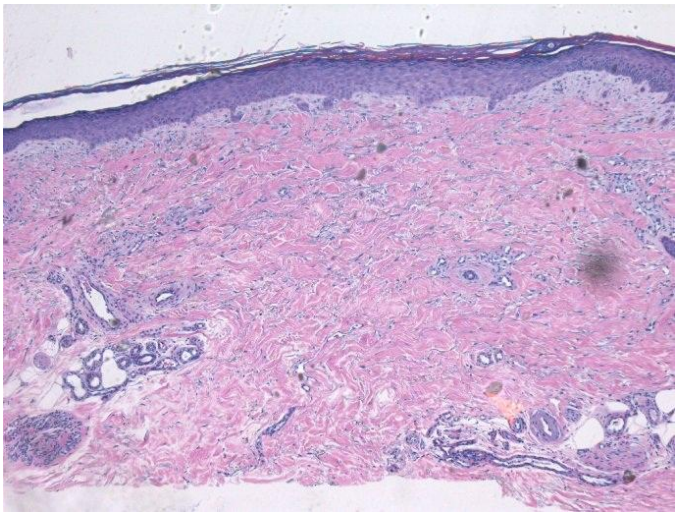


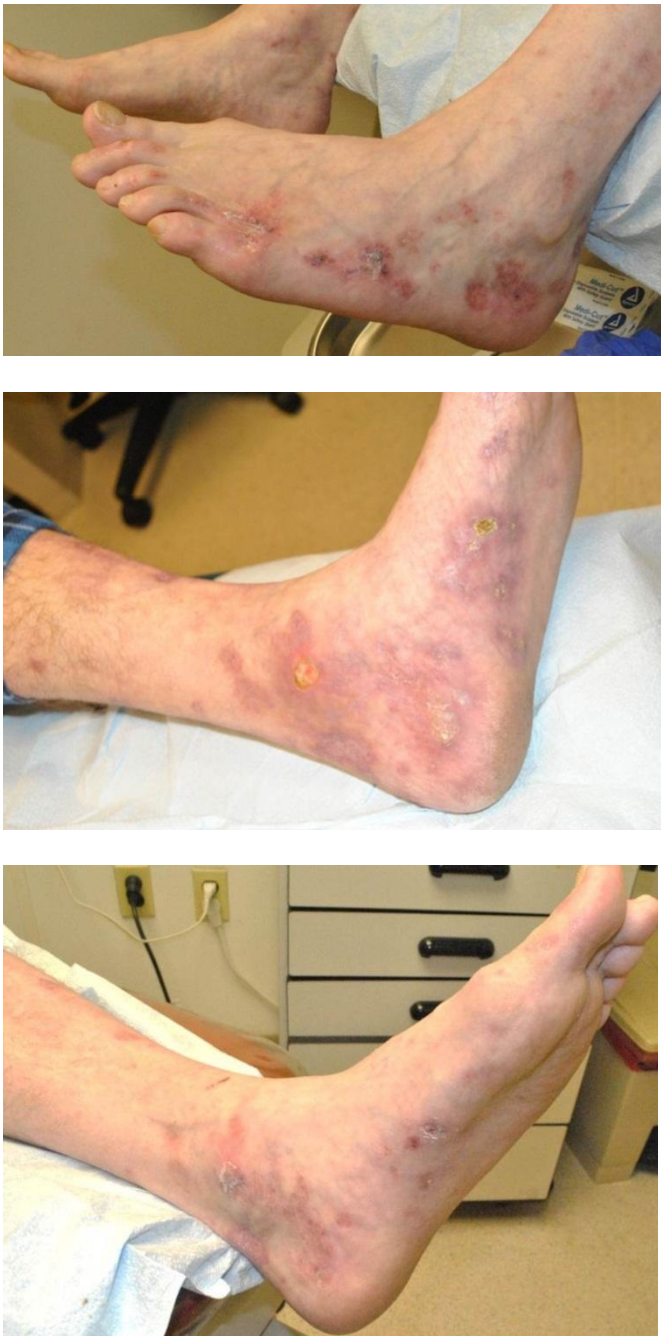
Figure 7 Direct immunofluorescence revealing medium-sized vessel disease.

At our institution, the patient was started on prednisone 50 mg BID, and tapered accordingly. Local wound care was also initiated and included intermittent sharp debridement. Over a six-month period the wounds diminished in size, and went on to complete resolution. The patient continued to follow up once the taper was finished and to this date, has not had a recurrence.

Discussion

Primary systemic vasculitis is uncommon, accounting for only four percent of cases in patients with cutaneous vasculitis. Infections lead the secondary causes accounting for 23% of cases followed by medications (20%), connective tissue disease (12%) and malignancy (4%). It has been reported that no identifiable cause is present in 3-72% of patients [1].

Vasculitis varies and may be classified based on the size of the blood vessel involved. Small-sized blood vessels are less than less than 50 μm and include capillaries, postcapillary venules, and nonmuscular arterioles. They are found primarily in the superficial papillary dermis. Medium-sized blood vessels are between 50-150 μm in diameter and contain muscular walls. They are often found in the deep reticular dermis at the junction of the dermis and subcutaneous tissue [2].



Figures 8-10 Follow up at eight months.

Systemic vasculitis may be caused by a variety of disorders, and vary depending on the vessel size. Large vessel vasculitis includes Giant Cell Arteritis and Takayasu arteritis. Medium vessel vasculitis includes Polyarteritis Nodosa and Kawasaki disease while small vessel vasculitis includes Wegener granulomatosis, Churg-Strauss syndrome, Microscopic polyangiitis, and Henoch-Schonlein purpura [3]. Secondary cases of vasculitis include

autoimmune disease, infection, malignancy, or drug reaction commonly from penicillins, sulfonamides, allopurinol, thiazides, quinolones, propylthiouracil and hydantoins [4].

Many clinical features are present in a patient with cutaneous vasculitis. Palpable purpura may be the first clinical sign of vasculitis, which results from extravasation of erythrocytes into the dermis [4]. Purpura does not blanch with pressure and may be found symmetrically along dependent regions. Petechiae may be found and present as non-blanchable non-palpable pinpoint macules. Digital necrosis and ulceration can result due to a decrease in vascular perfusion in the skin. Superficial ulceration is common in small vessel disease while deeper ulcerations are present in medium vessel disease [4]. Livedo reticularis, hemorrhagic bullae, and urticaria are other clinical features of cutaneous vasculitis. According to Ekenstam and Callen in a study of 82 patients diagnosed with cutaneous leukocytoclastic vasculitis, palpable purpura was present in 51 patients followed by urticarial-like lesions (17 patients), ulcerations (8 patients), vesiculobullous lesions (5 patients), erythematous plaques (5 patients), nodules (5 patients), livedo reticularis (3 patients), erythematous papules (1 patent), and necrosis (1 patient) [5]. The majority of patients only manifested one type of skin lesion. In 12 patients two or more types of skin lesions were present. The most common location of skin lesions was the lower legs.

If cutaneous vasculitis is suspected it is imperative to perform a biopsy to confirm the diagnosis. Two biopsies should be obtained and sent for both histopathologic and direct immunofluorescence. Biopsies should be performed within 24-48 hours after the appearance of a vasculitic lesion to prevent the pathological characteristics of vasculitis from being lost [2]. Next, it is imperative that biopsies be of the appropriate depth. The depth of the biopsy determines which vessels are being examined. If medium vessel vasculitis is suspected, the biopsy must include subcutaneous fat. An incisional biopsy should be employed if larger vessels are suspected. Lastly, the biopsy should be obtained from non-ulcerated sites or from the edges of the ulceration if possible.

Once cutaneous vasculitis has been confirmed through biopsy it is essential to determine the etiology, evaluate for systemic disease, identify any treatable cause, and remove any offending agent. A thorough history can establish if any medications, infections or systemic disease can account for vasculitis. A thorough physical exam should be performed followed by laboratory and radiological evaluation, and further diagnostic testing as indicated. If chronic or systemic vasculitis is suspected the following laboratory studies should be performed: complete blood count with differential, BUN/Creatinine, liver function panel, urinalysis, stool guaiac, hepatitis B and C virus serologies, cryoglobulins, precipitins, complement levels (CH50, C3, C4), ANCA, antinuclear antibody, and rheumatoid factor. Other tests to consider may include blood cultures and echocardiography if fever or heart murmur is present, and anti-streptolysin O titers in children [6].

Treatment of cutaneous vasculitis should start with the removal or treatment of the underlying cause. Any offending medications or causative agent should be removed, and any underlying systemic disorder should be treated. According to Carlson et al, treatment should be guided and based on the severity of disease [6]. Treatment should begin with management of symptoms and may include antihistamines, non-steroidal anti-inflammatory drugs, rest, and elevation, and avoiding cold exposure. For mild limited skin disease with persistent, recurrent, or symptomatic disease, dapsone and colchicine have been effective. For moderate to severe skin disease, with extensive or recurrent disease treatment may consist of methotrexate, azathioprine and or prednisone. Systemic vasculitis is treated with prednisone and may be combined with cyclophosphamide, azathioprine, cyclosporine, or mycophenolate mofetil. Other treatments may be based on the specific underlying disorder and may include TNF inhibitors, intravenous immunoglobulin, and plasmapheresis.

Conclusion

Cutaneous vasculitis is a rare condition that can be caused by a variety of different conditions and may present with varying degrees of clinical manifestations. If cutaneous vasculitis is suspected it is imperative to perform a biopsy to be sent for both histopathology and direct immunofluorescence. Once cutaneous vasculitis is suspected, treatment should be guided and based on the severity of the condition as well as any systemic disease.

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