Vasculitis Mimics

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Abstract:

Vasculitis is a heterogeneous group of rare diseases associated with significant morbidity and mortality. Although many criteria are suggested for classification of vasculitis nothing is ideal, as many other medical conditions resemble vasculitis clinically as well as pathologically. So only clinical experience is needed for diagnosing vasculitis after exclusion of various vasculitis mimics. Most common conditions mimicking vasculitis are infections followed by drugs, malignancy, Hypercoagulable states, inherited disorders and certain miscellaneous conditions like cardiac myxoma, calciphylaxis etc. This review is an attempt to increase awareness of these commoner conditions before diagnosing rare primary vasculitis syndrome.

Keywords: vasculitis, vasculitis mimics, infection.

Introduction:

Vasculitis is pathologic term that means inflammation and necrosis of blood vessels (1). It involves immunologically mediated responses to usually unknown antigens, which results in vessel wall damage. Weakening of the vessel wall can lead to aneurysms, dissections or bleeding and narrowing of the lumen resulting in ischemic damage and necrosis of the affected end organs and tissues (2).

Clinically vasculitis term is used to describe heterogeneous specific syndromes and carries significant morbidity and mortality. American College Of Rheumatology (ACR 1990) has suggested classification criteria for classifying these disorders into various types of large, medium and small vessel sized vasculitis (3). Further, vasculitis can be classified as generalized (systemic) or localized (isolated). It may be restricted disease causing inflammation of the blood vessels without any known cause where it is called as primary vasculitis or it may occur in association with a known underlying disease like infections, malignancy, medicines and various connective tissue diseases which is called as secondary vasculitis.

A variety of common and uncommon conditions may resemble primary vasculitis clinically and radiologically, or histologically which sometimes can satisfy ACR 1990 classification criteria also. These conditions are known as vasculitis mimics (4). Other names for these disorders are vasculitis look-alikes, vasculitis simulators and pseudo-vasculitis syndromes.

Hence, it is important to recognize these vasculitis mimickers from true vasculitis so that unnecessary treatment with steroid, immunosuppressive and cytotoxic agents, all with harmful side-effects can be avoided (5).

Endothelial dysfunction has important role in vascular injury which was understood by doing in vitro human umbilical vein endothelial cell cultures. Application of this knowledge to diseases in human being and to vasculitis and its mimickers is still under investigation (6).

There are certain clues for considering mimickers of vasculitis. If clinically patient has splinter hemorrhages, predominant liver abnormalities, presence of a heart murmur, history of drug abuse, high-risk sexual activity, presence of

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malignancy, then screening for vasculitic mimics should be done.

Certain disorders like infections or medicines can cause true vasculitis e.g., Hepatitis C viruses causing cryoglobulinemic vasculitis (CV), Hepatitis B virus causing polyarteritis nodosa (PAN), HIV associated vasculitis which is secondary to underlying infection. But some authors classify similar disorders as vasculitis mimickers. But discussion of this dilemma is beyond scope of this article.

Vasculitis mimickers can be classified based on etiology and depend on the vessel size involved.

➢ **Etiological classification (Table 1):**

Infections	Bacterial endocarditis, Tuberculosis, Disseminated gonococcal Infection, Pulmonary histoplasmosis, Coccidioidomycosis, Syphilis, Lyme disease, Whipple' s disease
Drugs	Cocaine, Amphetamines, Ergot alkaloids, Methysergide, Arsenic, Fluroquinolones, Hydralazin, Propilthiouracil, Anti TNF inhibitors
Inherited disorders	Marfan's syndrome, Neurofibromatosis, Ehler–Danlos syndrome (types IV and VI), Loeys–Dietz syndrome, Pseudoxanthoma elasticum
Hypercoagulable States	Antiphospholipid antibody syndrome, Thrombotic thrombocytopenic purpura
Malignancy	Atrial myxoma, Lymphoma, leukemia, Carcinomatosis
Miscellaneous	Sarcoidosis, Atheroembolic disease, Goodpasture's syndrome, Amyloidosis, Migraine, Cryofibrinogenemia

➢ **Size of vessel and vasculitis mimickers** [7]

Large arteries	Medium arteries	Small arteries
Infection: • Acute (e.g. mycotic aneurysms associated with septicemia or endocarditis), • Chronic (e.g. syphilis, tuberculosis, HIV, leprosy) **Atherosclerosis Congenital causes:** Aortic coarctation, Middle aortic syndrome **Hereditary disorders:** Marfan's syndrome, Neurofibromatosis, Ehler–Danlos syndrome (types IV and VI), Loeys–Dietz syndrome, Pseudoxanthoma elasticum **Fibromuscular dysplasia** **Iatrogenic:** Post-radiation therapy **Chronic periaortitis / inflammatory aortic aneurysm**	**Viral infection:** HBV, HCV, HIV, Herpes viruses **Other infections:** Infective endocarditis, Mycotic aneurysms **Atherosclerosis** **Malignancy:** Lymphoma, Leukemia Fibromuscular dysplasia (includes segmental arterial mediolysis) **Hereditary disorders:** Ehler–Danlos syndrome, Neurofibromatosis, Grange syndrome **Iatrogenic (postprocedural)** **Hypercoagulable states:** Thrombotic thrombocytopenic Purpura, Antiphospholipid syndrome	**Infection:** Infective endocarditis, Mycotic aneurysm with emboli, Sepsis, HIV **Cholesterol microemboli syndrome** **APLA Syndrome** **Ecthyma gangrenosum** **Thrombocytopenia with hemorrhage** **Drugs:** Cocaine, Amphetamine

Infections:

Infections are responsible for a number of different types of vasculitis. Conversely, patients with vasculitis have heightened risk to get infections, which sometimes mimic relapse [8].

Infections can mimic all spectrum of vasculitis. Many acute and chronic infections are vasculitis mimickers. Infective endocarditis (IE) has the ability to mimic the entire spectrum of Vasculitides. IE can cause mycotic aneurysms, septic vasculitis or immune complex-mediated purpura, glomerulonephritis (GN) or septic embolism. Glomerulonephritis associated with IE has granular immune complex deposits in glomerular basement membrane (GBM) whereas vasculitic GN as seen in Wegener's granulomatosis or microscopic polyangiitis are pauci-immune (4). Jain S and colleagues (9) has reported a case of vasculitis with blood cultures positive for Neisseria gonorrhoeae. Mastrolonardo M, et al described a case with extensive, vesicobullous, hemorrhagic, and necrotic cutaneous vasculitis was the sole manifestation of disseminated gonococcal infection (10).

Syphilis although is rarely seen infection now a days in India, its aortic and cardiac involvement closely resemble large vessel vasculitis (11).

The association of Tuberculosis with Takayasu's arteritis (TA) has been discussed several times in literature. In past, one study has suggested that TA have heightened immune response to Mycobacterium tuberculosis antigens, in particular to its 65 kDa HSP, suggesting that this organism may have a role in the immunopathogenesis of this disease (12). Other recent study does not support a direct role of MT in the pathogenesis of arterial lesions in either recent or longstanding TA, but does not exclude the possibility of a cross-reaction between mycobacterial and arterial antigens [13]. Other case reports have been described in literature with various types of vasculitis in association with pulmonary and extra pulmonary tuberculosis (14,15,16).

Among infections, viral infections have been found to be associated with true vasculitis like Hepatitis B virus (HBV) with polyarteritis nodosa and hepatitis C virus (HCV) with cryoglobulinemic vasculitis (17). Ilan Y described a case of cutaneous vasculitis and cryoglobulinemia with persistent hepatitis A infection (18). Inman RD et al described a case of cryoglobulinemia with relapsing hepatitis A infection as its extra hepatic manifestation (19).

Vasculitides are a rare but major manifestation of HIV infection, with an incidence of less than 1%, excluding adverse drug reactions (20). A wide range of vasculitic manifestations were reported in HIV-infected individuals. Vasculitis was described early in the disease with CD4 counts >500/µl, and in severe immune-compromised patients with CD4 counts <200/µl (21) All sizes of vessels can get involved in HIV associated vasculitis. Small vessel vasculitis in the form of cryoglobulinemia (22), vasculitic neuropathy, medium vessel vasculitis PAN (23,24), and large vessel vasculitis like coronaritis (25) cerebral vasculitis (26).

Some of them can be directly attributed to HIV infection and their treatment might require new therapeutic approaches, such as plasma exchanges, specific antiviral agents and/or vasodilators (27).

Other viruses causing vasculitis are erythrovirus B19, cytomegalovirus, varicella-zoster virus and human T-cell lymphotropic virus (HTLV)-1 have also been reported to be associated with or implicated in the development of vasculitides (28,29).

Some bacteria, fungi or parasites can also cause vasculitis, mainly by direct invasion of blood vessels or septic embolization, leading, e.g., to the well-known feature of 'mycotic aneurysm' (30).

General clues to infectious vasculitis are associated fever, abnormality of the peripheral leukocyte count, and recent or current extraneural infection (31). A Chauhan and colleagues have reported a case of proven Churg-Strauss vasculitis associated with ascaris infection (32)

Drug induced vasculitis (DIV):

DIV usually attacks the skin and sometimes the

subcutaneous part of the skin but sometimes also the kidneys and the lungs (33,34, 35, 36).

Multiple agents have been described causing various types of vasculitis but the incidence is low. The drugs that are known to cause vasculitis are hydralazine and PTU (37), quinolones, interferon, biologics (38) and drugs of abuse (cocaine, metamphet), leukotriene inhibitors causing Churg Strauss (39). Ergotamine and cocaine abuse causes vasculitis like manifestations due to vasospasm.

Features that distinguish drug-induced vasculitis from idiopathic autoimmune syndromes are presence of specific Antineutrophil cytoplasm (ANCA) antibodies. ANCA antibodies with specificity to more than one lysosomal antigen, combined with presence of antibodies to histones and beta-2 glycoprotein 1 constitute a unique serological profile for drug-induced vasculitis/drug-induced lupus-like disease (40).

DIV patients typically harbor ANCA directed to one or more neutrophil cytoplasm antigens, the most common antigens being the granule proteins MPO, HLE, cathepsin G, and lactoferrin (41).

Usually DIV are self limited and has cutaneous manifestations in the form of palpable purpura or maculopapular rash in legs along with arthralgia, myalgia but rarely there can be systemic disease.

Malignancy:

Commonest malignancies commonly associated with vasculitis are hematologic (e.g., lymphoma, leukemia), myelodysplastic syndrome (MDS), lymphoid, and solid tumor (42). The commonest solid tumors associated with vasculitis are carcinomas of urinary organs, lung, and gastrointestinal tract (43). Common vasculitis pattern associated with solid tumors are cutaneous leukocytoclastic vasculitis, Henoch-Shönlein purpura, WG, CSS, polyarteritis nodosa, and giant cell arteritis. Whereas hematologic malignancies are associated with predominant cutaneous vasculitis (44). Chronic or persistent vasculitis with poor response to usually effective therapy, especially in elderly patients, should raise questions about underlying malignancy.

Inherited disorders:

Hereditary causes of large-artery aneurysms such as Marfan's syndrome, Loeys-Dietz syndrome and Ehler-Danlos syndrome type IV (45,46) mimic large vessel vasculitis . Under-recognized mimics of medium-vessel vasculitis include segmental arterial mediolysis and Grange syndrome. In literature, a patient with Livedoid vasculitis who had doubly inherited thrombophilia, with heterozygous mutations in the Factor V Leiden (G1691A) and prothrombin (G20210A) has been described (47). In one study by Casato M et al investigations were done to see the contribution of inherited and acquired thrombophilic defects to the clinical manifestations of mixed cryoglobulinemia vasculitis. Clinical manifestations of mixed cryoglobulinemia were analysed as dependent covariates: severity of purpura, presence of necrotic skin ulcers, presence of peripheral neuropathy and presence of kidney disease. Hyper-homocysteinaemia was found as a risk factor for severe cutaneous manifestations in mixed cryoglobulinemia (48).

Atherosclerosis:

Although an inflammatory condition of vessels, atherosclerosis is not a vasculitis, it resembles vasculitis clinically as it may present as absent or weak pulses, bruit over vessel and rarely with digital gangrene in severe cases. Even angiographically it mimics vasculitis showing arterial stenosis of variable degree and aneurysm. But histologically these two disorders differ. Strong clinical suspicion of atherosclerosis is warranted particularly in elderly patients having risk factors for atherosclerosis.

Endothelial dysfunction (ECD), widely regarded as initial lesion in atherogenesis, has been shown to occur commonly in primary vasculitis (49). Both focal vascular inflammation and systemic inflammatory mediators like C reactive protein and tumor necrosis factor (TNF) alpha cause endothelial dysfunction (50). Understanding the role of endothelial function in the clinical outcome of systemic vasculitis deserves more attention for which research are necessary which can aid in inventing new therapy for vasculitis (51). Hisashi Masugata (52) et al

suggested detection of atherosclerosis in early stages of large vessel vasculitis before hypertension sets in by measuring cardio-ankle vascular index (CAVI).

Hypercoagulable states:

Antiphospholipid antibody syndrome (APLA): APLA can result in thrombotic microangiopathy and/or true vascular inflammation. The ultimate manifestation can mimic systemic vasculitic syndrome. There are reports of association of vasculitides with APLA (53). Boltin D et al described an ALA patient with resistant hypertension later found to be due to thrombosis and complete occlusion of the left renal artery (54). Egan AC described combination of thrombotic and inflammatory processes resulted in endocarditis, aneurysm formation and thrombosis (55).

Thrombotic Thrombocytopenic Purpura (TTP):

There are case reports in literature of co association of TTP with vasculitis particularly PAN (56,57).

Miscellaneous diseases:

Cardiac myxoma can manifest as vasculitis due to peripheral or central embolization. It can manifest as weight loss, low-grade fever, myalgia, arthralgia, and skin rash (58). Potential delayed neurologic complications relevant to patients with tumor embolization include myxoma-induced cerebral aneurysm and myxomatous metastasis, which can mimic the clinical picture of central nervous system vasculitis or infective endocarditis (59). There are also reports of systemic vasculitis as initial presentation of atrial myxoma (60).

Calciphylaxis: There have also been a number of case studies of calcific uremic arteriolopathy, or calciphylaxis (syndrome of vascular calcification, thrombosis and skin necrosis), described within vessels, including patients with chronic renal insufficiency and several forms of vasculitis. Calciphylaxis associated with vasculitis appears to be unique, although relatively uncommon and is likely secondary to a disruption in the calcium-phosphate-parathyroid hormone axis (61). Kyttaris VC et al described a case of chronic kidney disease with intractable lower extremity ulcers and skin findings suggestive of small-vessel disease of the upper extremities, but biopsy of the lesions showed classic calciphylaxis without evidence of vasculitis (62).

Cholesterol Emboli Syndrome: Carlson JA has described Cholesterol Embolization as cause of cutaneous pseudovasculitis (63). Livedo reticularis of the lower extremities and acrocyanosis are the most common cutaneous manifestations. Recently, it has been proposed that cholesterol embolization is associated with vasculitis, and some authors have labeled this condition a "vasculitis look-alike." Potential fatality of this problem is associated with renal involvement (64).

Common Variable Immunodeficiency (CVID): There is a paradoxical relationship between immunodeficiency diseases and autoimmunity. Immunologic defects may result in a failure to exclude microbial antigens, resulting in chronic immunologic activation and autoimmune symptoms. Inherited deficiencies of the complement system have a high incidence of systemic lupus erythematosus (SLE), glomerulonephritis, and vasculitis (65). There are case reports of leukocytoclastic vasculitis and WG with CVID (66,67).

Conclusion:

Primary systemic vasculitis requires early diagnosis and treatment to avoid the complications of disease like ischemia or gangrene of the involved organ. It's necessary to keep vasculitis mimics in mind before starting high dose steroid and toxic immunosuppressive agents.

The immunosuppressive, cytotoxic agents are associated with many adverse effects and can be hazardous if used in setting of infection which is important vasculitis mimicker as discussed here in this review. The uncommon vasculitis mimics like inherited disorders, cardiac myxoma and cholesterol embolization should also be kept in mind before exposing patients to toxic long term therapy.

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