Challenging Mimickers of Primary Systemic Vasculitis

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KEYWORDS

• Mimicker • Vasculitis • IgG4-related disease • Livedoid vasculopathy
• Segmental arterial mediolysis • Lymphomatoid granulomatosis
• Fibromuscular dysplasia • Degos disease

KEY POINTS

- Immunoglobulin G4–related disease is a common mimicker of small vessel and medium vessel vasculitides, particularly granulomatosis with polyangiitis, but it can also cause a true vasculitis of large vessels, requiring distinction from giant cell arteritis, among other vasculitides.
- Arterial instrumentation should be avoided whenever possible in cases of segmental arterial mediolysis and fibromuscular dysplasia, because such procedures can lead to arterial dissections.
- Calciphylaxis typically involves adipose tissues (eg, the thighs, buttocks, abdomen, and flanks).
- The myeloproliferative form of hypereosinophilic syndrome can be detected with examination of the bone marrow in addition to blood or bone marrow aspirate testing for FIP1L1/PDGFRA fusion, which is present in a subset of such patients.
- Of livedoid vasculitis begin as tender erythematous nodules that then rapidly ulcerate and scar with atrophie blanche. The ulcers have an irregular shape and are extremely painful.

Among the most challenging aspects of evaluating and caring for patients with systemic vasculitis is the need to distinguish rigorously between vasculitis and a host of conditions that can mimic vasculitis closely (**Box 1**). The treatment approaches for vasculitis mimickers are varied and often differ substantially from those required to treat vasculitis. This article reviews 9 challenging vasculitis mimickers: fibromuscular dysplasia (FMD), calciphylaxis, segmental arterial mediolysis, antiphospholipid syndrome (APS), hypereosinophilic syndrome, lymphomatoid granulomatosis (LMPG), malignant atrophic papulosis, livedoid vasculopathy, and immunoglobulin (Ig) G4–related disease (IgG4-RD).

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Box 1
Systemic vasculitis mimickers: a comprehensive list

Conditions mimicking small vessel vasculitis
- Antiphospholipid antibody syndrome[a]
- Atheroembolic disease
- Calciphylaxis[a]
- Hypereosinophilic syndrome[a]
- Emboli (cardiac myxoma, cardiac thrombus, endocarditis, mycotic aneurysm, others)
- Idiopathic diffuse alveolar hemorrhage
- Infection (endocarditis, disseminated intravascular coagulation, Rocky Mountain spotted fever, others)
- Intravascular lymphoma
- Levamisole-induced vasculitis
- Lymphomatoid granulomatosis[a]
- Malignant atrophic papulosis (Degos disease)[a]
- Thrombotic thrombocytopenic purpura[a]

Conditions mimicking medium vessel vasculitis
- Livedoid vasculopathy[a]
- Fibromuscular dysplasia[a]
- Segmental arterial mediolysis[a]
- Thromboangiitis obliterans (Buerger disease)

Conditions mimicking large vessel vasculitis
- IgG4-related disease[a]
- Erdheim-Chester disease
- Ehlers-Danlos type IV
- Loeys-Dietz syndrome
- Marfan syndrome

Conditions affecting a single organ
- Reversible cerebral vasoconstriction syndrome[a]

[a] Discussed in this article.

FIBROMUSCULAR DYSPLASIA

FMD is a noninflammatory vasculopathy of small and medium-sized arteries that can lead to aneurysm, stenosis, occlusion, and dissection.[1,2] This disease may occur in any age group, but mainly affects children and individuals more than 50 years of age.[3,4] The prevalence of FMD in the general population is estimated to be around 2% to 3%.[4,5] Women comprise up to 90% of cases in adults.[3,6] Approximately 10% of patients with FMD report a family member carrying the same diagnosis.[3,7]

The most commonly affected vascular sites are middle and distal portions of the renal, internal carotid, and vertebral arteries (~65% of the cases).[3] Lesions are detected less frequently in the intracranial, common carotid, external carotid,