Polyarteritis nodosa

What is PAN?

Diagnosis

Muscle biopsy

Symptomatic muscle is best to biopsy.

Nerve biopsy

Arteriogram

Muscle pain

In PAN, muscle involvement is common and is described as myalgia with muscle weakness, and it may present as muscle pain or claudication.

Creatine kinase can be elevated, but it is usually not high enough to suggest an inflammatory myopathy.

When myalgias are part of PAN, they are classically described as not limited to the shoulder and hip girdle, and may include tenderness of leg muscles.

Differential diagnosis

Decide whether the working diagnosis is atherosclerotic PAD versus vasculitis

Before committing to immunosuppression for suspected medium-vessel vasculitis, important mimics to consider include atherosclerosis, embolic disease, thrombosis, and vasospasm, as well as infection-related mimics.

Among infectious diseases, the following may cause clinical manifestations that either mimic vasculitis or cause vascular inflammation:

Infective endocarditis or other bacteremic disease

Mycotic aneurysm with distal embolization

Hepatitis B or C virus infection

HIV infection

Other disorders that may mimic vasculitis of medium sized arteries include:

Atherosclerosis

Embolic diseases (eg, left atrial myxoma, cholesterol crystals)

Thrombotic disorders (eg, catastrophic antiphospholipid syndrome)

Fibromuscular dysplasia

Ergotism

Radiation fibrosis

Necrotic arachnidism (Loxosceles species spider bites)

Malignant atrophic papulosis (Degos disease or syndrome)

Segmental arterial mediolysis (SAM)

Other causes of malignant hypertension

Other systemic vasculitides to consider are:

Granulomatosis with polyangiitis and microscopic polyangiitis

Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)

Immunoglobulin A vasculitis (Henoch-Schönlein purpura)

Cryoglobulinemic vasculitis

Drug-induced vasculitis

Vasculitis secondary to connective tissue disease (eg, systemic lupus erythematosus, rheumatoid arthritis)

Biopsy

Biopsy examination of the involved tissue is essential for diagnosis of many vasculitides, but is not possible in all cases. For example, a temporal artery biopsy should be considered in cases of suspected GCA and is generally a straightforward procedure. Similarly, skin biopsies of purpuric lesions and kidney biopsies in patients with suspected glomerulonephritis have high diagnostic yields. However, for patients with suspected TAK, except in cases where surgical repair was conducted, tissue biopsy from the aortic arch and its primary branches is not feasible, and the diagnosis is based on other clinical and radiographic findings.

If calf pain is the main manifestation, muscle biopsy can be considered but should be targeted.

Muscle biopsy can show inflammatory changes in blood vessel walls in vasculitic polyneuropathy evaluations.

For muscle biopsy site selection principles: choose a muscle that is weak but not completely atrophic, and avoid muscles recently subjected to EMG.

Calf muscle biopsy is discouraged in myopathy evaluations due to frequent artifactual findings; if a muscle biopsy is pursued for suspected vasculitis, targeting should be deliberate rather than defaulting to calf solely because symptoms are in the calf.

In vasculitis evaluation, combined nerve–muscle biopsy is used in selected cases (often sural nerve with adjacent gastrocnemius muscle), but biopsy can be nondiagnostic due to sampling error.

For percutaneous muscle biopsy in general, UpToDate notes that image-guided biopsy may have fewer complications such as bleeding when ultrasound guidance is used.

Across biopsy procedures that break the skin, common complication categories include pain, bleeding, infection, and scarring or wound issues.

Biopsy is not used for routine follow-up once vasculitis is established, but it can be useful when histopathology is needed to distinguish vasculitis from mimics such as infection or malignancy.

Management of PAN single organ

Unlike cutaneous PAN, single-organ PAN is usually monophasic and does not relapse. The diagnosis of isolated single-organ PAN is typically made by histological examination of surgical specimens. Treatment beyond surgical excision is usually not necessary. However, we fully evaluate such patients initially and provide regular clinical follow-up (every three months in the first year and every 6 to 12 months thereafter) to determine if additional anatomic areas or other clinical features are present or developing.

Methotrexate versus azathioprine

When a decision is made to start an immunosuppressive drug in addition to glucocorticoids for treatment of nonsevere disease, either azathioprine (2 mg/kg daily) or methotrexate (20 to 25 mg once weekly) for at least one year is preferred.

Base the choice between methotrexate and azathioprine on an

evaluation of potential contraindications, and patient and clinician preferences regarding dosing and administration of the drugs. Methotrexate may be faster-acting but should be avoided in patients with kidney disease or hepatitis. Prefer azathioprine in patients with kidney disease and in patients with hepatic disease, although these patients should be closely monitored for hepatotoxicity.

There is limited evidence to support this approach to treatment of nonsevere disease. In a small trial of 95 patients with a case mix of PAN, eosinophilic granulomatosis with polyangiitis (EGPA; Churg-Strauss), and MPA compared the addition of azathioprine or placebo to standard daily oral glucocorticoids for patients classified as having nonsevere disease. The trial did not find a benefit of azathioprine in terms of rates of attaining remission, risk of relapse, or cumulative dose of glucocorticoids. However, only a minority, 19 out of 51 (37 percent) had PAN, limiting the interpretation of these results for patients with PAN. The small sample size of patients with PAN precluded disease-specific subset efficacy analysis. More data are needed on the efficacy of azathioprine and methotrexate as "glucocorticoid-sparing" agents for patients with mild PAN to guide therapy.

References

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