An Approach to the Evaluation and Management of Vasculitis

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KEYWORDS

• Vasculitis • Vasculitis syndrome • Primary vasculitis • Secondary vasculitis
• Antineutrophil cytoplasmic antibodies • Immune complex • Granuloma
• Corticosteroids

HOSPITAL MEDICINE CLINICS CHECKLIST

1. Vasculitis is a term representing many heterogeneous and often overlapping groups of clinicopathologic syndromes defined by the presence of inflammatory leukocytes in blood vessel walls that lead to vascular damage and downstream organ dysfunction.
2. Although individual vasculitides are relatively uncommon, this category of disease processes has an incidence of 20 in every million persons.
3. The exact pathophysiologic mechanism is not known, although autoimmunity and immunopathogenic responses to certain stimuli are likely to play key roles.
4. Possible mechanisms of vessel damage implicated in vasculitis include effects of immune complexes, pathogenic T-cell responses with granuloma formation, and autoantibodies such as antineutrophil cytoplasmic antibodies.
5. Vasculitis diagnoses are classically grouped into primary and secondary vasculitides, by size of vessel affected, and by demographics and clinicopathologic characteristics of affected patients.
6. A complete history and physical examination are paramount in the evaluation of vasculitis as typical clinical manifestations are neither sensitive nor specific and frequently mimic other more common disorders.

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7. Vasculitis should be included in the differential diagnosis for any patient presenting with unexplained constitutional symptoms and red-flag examination findings such as mononeuritis multiplex or palpable purpura.
8. Because the differential diagnosis of vasculitis is broad and includes any systemic disease process, the first step in evaluation is exclusion of multisystem mimickers.
9. Many laboratory tests, both general and directed, may be of use in establishing the diagnosis, determining the pattern and extent of disease, and identifying any contributors to the pathogenesis; however, most are nonspecific and not diagnostic.
10. The inpatient diagnostic approach depends on clinical and patient factors including organ(s) involved, caliber of pathologic vessels, and overall risk-benefit ratio of procedures; common studies include histologic biopsy, arteriography, cross-sectional imaging, and electromyography.
11. Inpatient admission and expedited evaluation is typically pursued when a patient's symptoms and/or organ dysfunction are severe or as yet unexplained, and subspecialist consultants are often instrumental in the care of these individuals.
12. Therapy is aimed at inducing and maintaining remission of disease activity and preventing complications arising from treatment.
13. Individual treatment regimens are tailored to patients, and depend on both the category and severity of the disease process; typical therapies for more extensive disease include immunosuppressants such as corticosteroids, and steroid-sparing agents such as methotrexate.
14. An effective transition from inpatient to outpatient setting requires coordination with follow-up providers and excellent patient education through medication reconciliation, review of possible medication adverse effects, and information about relevant signs and symptoms.
15. New or worsening findings in a patient treated for vasculitis does not necessarily represent relapse of disease; hospitalists must be cognizant of other potential causes such as inappropriate therapeutic regimen, drug toxicity, or fibrosis and narrowing of vessels on healing.
16. Regular monitoring of both disease activity and adverse effects from treatments is critical to successful patient care.
17. Prognosis of vasculitis syndromes is variable, although generally favorable; the key determinants of mortality in the acute phase are the effects of untreated vasculitis, whereas late deaths are often attributed to complications of therapy.

DEFINITION

How is vasculitis defined?

The term vasculitis encompasses an expansive and heterogeneous group of clinicopathologic disease processes whereby inflammatory leukocytes are found in blood-vessel walls. These cells initiate a cascade of inflammation, which leads to reactive damage within vascular structures. Subsequent obstruction of the vessel lumen by