ONE MINUTE CONSULT

BRIEF ANSWERS TO SPECIFIC CLINICAL QUESTIONS

Q: When does pericarditis merit a workup for autoimmune or inflammatory disease?

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A: Pericarditis is in most cases a one-time disease simply treated with anti-inflammatory drugs. It requires no extensive workup for systemic inflammatory or autoimmune disease. Further evaluation is required for patients who have recurrent pericarditis resistant to conventional therapy or pericarditis with manifestations of systemic disease.

ACUTE PERICARDITIS

Pericardial disease has different presentations: acute, recurrent, constrictive, effusive-constrictive, and pericardial effusion with or without tamponade. Acute pericarditis is the most common of these and can affect people of all ages. The typical acute manifestations are chest pain (usually pleuritic), a pericardial friction rub, and widespread ST-segment elevation on the electrocardiogram.[1,2] The chest pain tends to be sharp and long-lasting; it radiates to the trapezius ridge and increases during respiration or body movements.

Acute pericarditis usually responds to an anti-inflammatory drug such as colchicine 0.6 mg/day for 3 months, a nonsteroidal anti-inflammatory drug such as ibuprofen 600 mg three times a day for 10 days, and in advanced resistant cases, an oral corticosteroid.[3,4]

Most often, pericarditis is either idiopathic or occurs after a respiratory viral illness. Much less common causes include bacterial infection, postpericardiotomy syndrome, myocardial infarction, primary or metastatic tumors, trauma, radiation, and uremia. However, pericarditis can also be part of the presentation of systemic inflammatory and autoimmune diseases such as rheumatoid arthritis and systemic lupus erythematosus; hereditary periodic fever syndromes such as familial Mediterranean fever; and systemic-onset juvenile idiopathic arthritis.[1,5]

In acute pericarditis, a complex workup is usually not justified, since the results will have limited usefulness in the clinical management of the patient. It is most often diagnosed by the presenting symptoms, auscultation, electrocardiography, echocardiography, and chest radiography, and by additional basic tests that include a complete blood cell count, complete metabolic profile, erythrocyte sedimentation rate, and C-reactive protein level. However, if pericarditis does not respond to anti-inflammatory treatment and if an autoimmune or infectious disease is suspected, further evaluation may include antinuclear antibody testing and testing for human immunodeficiency virus and tuberculosis. If the diagnosis of acute pericarditis remains uncertain, cardiac magnetic resonance imaging (MRI) may be useful.

RECURRENT PERICARDITIS

Although acute pericarditis most often has a benign course and responds well to anti-inflammatory drugs, 20% to 30% of patients who have a first attack of acute pericarditis have a recurrence, and up to 50% of patients who have one recurrence will have another.[3,4]

Disease activity can be followed with se-

Patients with recurrent pericarditis and pericarditis with manifestations of systemic disease need a thorough workup for autoimmune disease

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rial testing of inflammatory markers—eg, erythrocyte sedimentation rate and C-reactive protein level. Echocardiography, cardiac computed tomography, and cardiac MRI can characterize active inflammation, edema, pericardial thickness, and pericardial effusion.[6–8]

Recurrent pericarditis is often resistant to standard therapy and requires corticosteroids in high doses, which paradoxically can increase the risk of recurrence. Therefore, further workup for underlying autoimmune disease, systemic inflammatory disease, or infection is necessary. More potent immunosuppressive therapy may be required, not only in pericarditis associated with systemic autoimmune or inflammatory conditions, but even in idiopathic recurrent pericarditis, either to control symptoms or to mitigate the effects of corticosteroids.

SYSTEMIC INFLAMMATION

The true prevalence of pericardial disease in most systemic inflammatory and autoimmune diseases is difficult to determine from current data. But advances in serologic testing and imaging techniques have shown cardiac involvement in a number of inflammatory diseases.[9]

In one study, a serologic autoimmune workup in patients with acute pericarditis found that 2% had collagen vascular disease.[9] Pericardial involvement is likely in systemic lupus erythematosus,[10] and a postmortem study of patients with systemic sclerosis found that 72% had pericarditis.[11] Mixed connective tissue disease has been associated with pericarditis in 29% of cases and 56% in autopsy studies.[12,13] Pericarditis may be the initial manifestation of vasculitis—eg, Takayasu arteritis or granulomatosis with polyangiitis (formerly known as Wegener granulomatosis).

Other diseases with pericardial involvement include Still disease, Sjögren syndrome, sarcoidosis, and inflammatory bowel disease. Symptomatic pericarditis occurs in about 25% of patients with Sjögren syndrome and asymptomatic pericardial involvement in more than half. Autopsy studies reported pericardial involvement in up to 80% of patients with systemic lupus erythematosus. Cardiac tamponade occurs in fewer than 2%, and constrictive pericarditis is extremely rare.[5,9–11]

RECOMMENDATIONS

Patients with a first episode of pericarditis should be treated with an anti-inflammatory medication, with no comprehensive testing for autoimmune disease. An evaluation for autoimmune and infectious disease should be carried out in patients with fever (temperature > 38°C; 100.4°F), recurrent pericarditis, recurrent large pericardial effusion or tamponade, or night sweats despite conventional medical therapy. Signs of systemic disease such as renal failure, elevated liver enzymes, or skin rash merit further evaluation.

Prospective studies using appropriate serologic testing and imaging are needed to determine the correlation between myopericardial involvement and inflammatory diseases because of increased morbidity and mortality in several of these diseases.

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