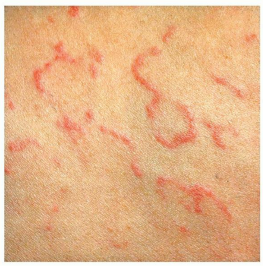

RHEUMATIC FEVER

Acute rheumatic fever (ARF) is a delayed sequel to group A β-hemolytic streptococcal (GAS) infection of the oropharynx. It is an inflammatory disease that can affect the heart, joints, central nervous system, skin, and subcutaneous tissues. There are no specific diagnostic tests, rather it is a clinical diagnosis made on the basis of certain criteria. Treatment of the underlying infection leads to prevention of rheumatic fever.

Epidemiology

ARF is rare and most commonly occurs in children 5 to 15 years of age after clinically severe

tonsillopharyngeal infections. However, it has been reported in patients of all ages and in those with asymptomatic disease. The incidence of rheumatic fever has dropped precipitously since the 1950s with widespread recognition of disease and treatment with penicillin. It has been nearly eradicated from the developed world but remains a serious health problem in developing countries. The World Health Organization estimates that 15.6 million people worldwide are affected by rheumatic heart disease, and that 233,000 people die annually either from rheumatic heart disease or acute renal failure as a result of complicated GAS infection.

Pathogenesis

The exact mechanism by which ARF results from an infection with group A streptococcal bacteria is unclear, but it is thought to be an exaggerated immune response to infection. Some strains of streptococcus are more likely to cause ARF than others. For instance, pharyngeal strains are much more likely to cause ARF than extrapharyngeal strains responsible for impetigo or cellulitis. Molecular mimicry has also been implicated in the etiology of ARF. M protein has structural similarity to cardiac myosin. Studies have demonstrated that rats immunized with streptococcal M protein develop myocarditis. Oddly, however, the morbidity and mortality of rheumatic fever results from valvular disease rather than acute carditis. Familial clustering of disease, and the linkage of HLA-DR4 and -DR2 to ARF in Caucasian and African American patients demonstrates that host properties also affect the susceptibility of individual patients in the development of disease.

Clinical Features

The clinical features of ARF vary widely and are largely outlined in the Jones criteria used to aid in diagnosis of the disease. The Jones criteria, updated in 1992, emphasize five characteristic major manifestations: polyarthritis, carditis, chorea, subcutaneous nodules, and erythema marginatum . Non-specific signs of systemic inflammation are considered minor criteria and include arthralgia, fever, elevated acute phase reactants, and a prolonged P-R interval by echocardiogram.

Diagnosis is based on the presence of two major criteria and one minor criteria plus evidence of preceding infection with group A streptococcus. If, however, chorea and carditis are present then demonstration of antecedent strep infection is not required to make the diagnosis.

Rheumatic carditis usually involves the endocardium, myocardium, and pericardium and almost always is associated with a murmur, either mitral or aortic regurgitation. Carditis, if present, usually presents in the first 3 weeks of the illness. There has been much debate about the use of echocardiography to aid in diagnosis of rheumatic heart disease without clinical evidence of a murmur; however, thus far its routine use has not been advocated. Its use has been accepted in patients with polyarthritis where other diagnostic criteria have not yet been met and in indistinguishable heart murmurs.

The Jones Criteria for Rheumatic Fever, Updated 1992

MAJOR CRITERIA

MINOR CRITERIA

□

Carditis

□

Clinical

□

Migratory polyarthritis

• Fever

□

Sydenham chorea

• Arthralgia

□

Subcutaneous nodules

□

Laboratory

□

Erythema marginatum

• Elevated acute phase reactants

• Prolonged P-R interval

plus

Supportive evidence of a recent group A streptococcal infection (e.g., positive throat culture or rapid anti

From Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. JAMA 268, 1992. Special Writing Grou

Migratory polyarthritis can affect any joint but most often involves the large joints. Early administration of anti-inflammatory medications does mitigate the arthritis and may therefore mask it. In addition, post-streptococcal reactive arthritis exists and can confound the picture. The diagnosis of chorea is based entirely on clinical signs, which include fleeting local muscular weakness, emotional lability, personality changes, and erratic, jerky, purposeless movements. Chorea does not occur in adult men.

The dermatologic manifestations of ARF are characteristic but rare. Subcutaneous nodules are small, painless, and localized over bony prominences and in tendon sheaths. Typically, they last for 1 to 2 weeks and spontaneously resolve. Erythema marginatum begins as an erythematous macule or papule extending outward while the central skin returns to normal. The border is pink and serpiginous, is not indurated, and blanches with pressure . Patients are often unaware of its presence. Histopathologically, there is a sparse superficial perivascular infiltrate of lymphocytes and neutrophils.[10]

Treatment

Treatment is directed at eradication of GAS from the oropharynx. Penicillin is the antibiotic of choice, is inexpensive, and can be administered orally or intramuscularly. Oral therapy should be continued for at least 10 days. Penicillinallergic patients can be treated with cephalosporins or macrolide antibiotics. Due to the high rate of recurrence in previously affected people, antibiotic prophylaxis is recommended, especially in endemic areas.

The acute inflammatory process is largely treated symptomatically. Anti-inflammatory medications including salicylates and nonsteroidal agents are effective for the arthritis, fever, and arthralgia. However, salicylates alone have not been shown to be effective in decreasing the incidence of rheumatic heart disease after ARF. Corticosteroids are believed to be beneficial in the treatment of carditis; though this has never been shown in a randomized controlled trial. Lastly, intravenous Ig has also been tested but not shown any benefit. Approximately 80 percent of patients who fulfill the criteria for this disorder recover spontaneously.