The disease that “bites the heart and licks the joints”

A recently published report of rheumatic fever in Nova Scotia serves as a reminder that this disease, once thought to have virtually disappeared from North America, is still with us and can pose a diagnostic challenge for physicians.

In early July 1997 a 10-year-old girl in Nova Scotia experienced upper respiratory tract symptoms, which were followed a week later by arthritis in the right first metacarpophalangeal joint. She was given antibiotics on the basis of a presumptive diagnosis of septic arthritis but subsequently experienced migratory arthritis of both knees and ankles and the right foot. On admission, on July 21, she had acute arthritis of both knees and ankles, tachycardia and a systolic murmur. Electrocardiography showed a prolonged PR interval, and echocardiography confirmed mitral and aortic valve insufficiency. The antistreptolysin-O level was elevated (to 600 IU/mL), and the erythrocyte sedimentation rate was 119 mm/hour.

The child’s illness met the current case definition for acute rheumatic fever because she had at least 2 major criteria (migratory polyarthritis and carditis) and there was evidence of an antecedent streptococcal infection (although throat culture at the time of admission was negative for streptococci, likely because of the recent antibiotic therapy). Two other cases were confirmed in Nova Scotia in the same month.

Rheumatic fever remains a major health problem in developing countries. In North America its incidence declined dramatically after World War II. However, in the mid-1980s there were several reports of large outbreaks, for example in the western US in 1985–1986. The reasons for the North American resurgence of rheumatic fever are unclear. Although once a disease strongly associated with overcrowding, inadequate housing and poverty, rheumatic fever is now disdainful of social class. At about the same time as rheumatic fever began to recur, cases of “toxic-strep syndrome” (life-threatening systemic infections caused by streptococci) were widely reported. It is likely that these developments represent a fundamental change in the virulence of this genus of bacteria.

Children and teenagers are the most likely to have acute streptococcal pharyngitis (which may be asymptomatic) and, not unexpectedly, are at highest risk for rheumatic fever. The disease usually begins 3 to 4 weeks after acute streptococcal pharyngitis. In about 70% of patients migratory arthritis develops, usually involving the large joints of the extremities and sometimes accompanied by signs and symptoms of a febrile illness. Most of the patients without arthritis have arthralgias, considered a minor manifestation. About 50% to 60% of patients with rheumatic fever exhibit evidence of pericarditis, myocarditis or endocarditis. The last of these is the most serious manifestation of rheumatic fever and usually involves the mitral or aortic valves. The joint symptoms are milder than the heart problems in children, in whom the disease is said to “bite the heart and lick the joints.” The reverse holds true in adults, in whom the disease is generally rare and milder. Other manifestations are less common; these consist of erythema marginatum, subcutaneous nodules, chorea (major), fever and an elevated C-reactive protein level (minor).

The acute respiratory tract infection should be treated to prevent an initial attack of rheumatic fever. A recently published clinical aid for the correct diagnosis of acute streptococcal pharyngitis should be of assistance in this regard. Once the diagnosis has been established, salicylates can be given to manage the fever and joint symptoms. Patients with cardiac involvement may require bed rest. There is a 50% chance of recurrence of rheumatic fever, so prophylactic treatment with appropriate antibiotics should be continued for at least 5 years after the acute episode.

References