

ACUTE RHEUMATIC FEVER WITH THREE MAJOR CRITERIA: POLYARTHRITIS, CARDITIS AND CHOREA*

A Case Report

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SUMMARY: Günal N, Atakan C, Köse G, Atasay B. (Pediatric Cardiology Unit, Social Security Children's Hospital, Ankara, Turkey). Acute rheumatic fever with three major criteria: polyarthritis, carditis and chorea. A case report. Turk J Pediatr 1998; 40: 585-588.

An eight-year-old girl is presented with three major criteria of acute rheumatic fever: polyarthritis, carditis and chorea. The diagnosis was confirmed with a history of pharyngitis 15 days prior to admission and with the findings of positive acute phase reactants such as elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), elevated anti-streptolysin-O (ASO) titration, and clinical findings of polyarthritis, carditis and chorea. Patient responded well to salicylate and phenobarbital treatment. The rare association of these three major criteria in acute rheumatic fever is emphasized. *Key words:* rheumatic fever, carditis, arthritis, chorea.

Rheumatic fever, the most common cause of acquired heart disease in children and young adults, is a multisystem disease, affecting primarily the heart, the joints, the brain and cutaneous and subcutaneous tissue¹. Particularly in developing countries, the incidence of the disease continues to be high. The diagnosis of rheumatic fever is based on the Jones criteria, in addition to evidence of group A streptococcal pharyngitis². In this report, an eight-year-old girl is presented with three major criteria of acute rheumatic fever: polyarthritis, carditis and chorea.

Case Report

An eight-year-old girl was admitted to the hospital because of arthralgia and involuntary movements. She had had sore throat, fever and pruritic rash 15 days before admission. In another hospital, Group A beta hemolytic streptococcus was isolated from the throat culture and procaine penicillin was administered. She was given two injections of procaine penicillin, but it was discontinued as the symptoms subsided after the second day. Two weeks later, swelling and

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arthralgia began in the knees and ankles in a migratory manner, and the family noticed behavioral changes, involuntary movements and restlessness in the patient.

On physical examination, the patient's body temperature was 37 °C, heart rate was 120/min, and blood pressure was 90/60 mmHg. Involuntary choreic movements of the upper extremities, jerking of the head and tongue fasciculations were detected. The left knee was tender and swollen. A third-degree pansystolic murmur was heard at the apex.

The laboratory findings were as follows: hemoglobin 11 g/dl, leukocyte 10,000/mm³, erythrocyte sedimentation rate (ESR) 78 mm/h, C-reactive protein (CRP) positive, anti-streptolysin O (ASO) titer 800 U, throat culture negative. Telecardiography showed mild cardiomegaly with a cardiothoracic ratio of 0.53, and electrocardiogram showed sinus tachycardia with a heart rate of 125 beats/min, and a PR interval of 0.16 s. Mitral valve regurgitation with a regurgitant flow velocity of 3.46 m/s and mild aortic regurgitation were detected by Doppler echocardiography. According to the above findings, our diagnosis in this patient was acute rheumatic fever with arthritis, carditis and Sydenham's chorea. Therapy was started with salicylate (100 mg/kg, in 4 doses) and phenobarbital (5 mg/kg, 3 times). At the end of the first week of therapy, clinical signs and laboratory findings had subsided. Findings of arthritis disappeared, choreic movements were diminished and the murmur of mitral valve insufficiency decreased. Erythrocyte sedimentation rate decreased to 26 mm/h, ASO to 600 U, and CRP was found to be negative. In the second week, the aspirin dosage was reduced to 75 mg/kg until the end of the third week and then was gradually reduced until it was discontinued at the end of the sixth week. All the laboratory and clinical findings were normal with the exception of a first-degree pansystolic murmur at the apex. The patient was discharged on prophylactic penicillin treatment and was scheduled for periodic control examinations.

Discussion

Rheumatic fever and its major complication, valvular heart disease, are still the most frequent cause of acquired heart diseases in children and adolescents, especially in developing countries. The incidence of rheumatic fever was found to be about 20 per 100,000 in a study from Turkey³. In other study from Turkey, done by İmamoğlu⁴, the prevalence of rheumatic heart disease changed between 10.7 to 2.1 per 1,000, with a higher prevalence in children of low socioeconomic level. Most attacks of rheumatic fever begin with arthritis, or arthritis with carditis. Sydenham's chorea occurs much later, with a much longer latent period than

the other manifestations, usually without any evidence of acute inflammation. In our case, three major manifestations of rheumatic fever were present together with the positive acute phase reactants. To our knowledge, few cases of rheumatic fever presenting with all three major criteria at the same time¹.

Karademir et al.⁵ found 100 patients with arthritis only, 59 with carditis, 40 with chorea and 24 with arthritis and carditis, out of 228 patients.

An epidemiologic study of rheumatic fever from Italy showed a progressive reduction of incidence and prevalence of the disease in the last 10 years. The percentage of carditis in the pediatric age was high (60-70%), often associated with arthritis or chorea. Relapses and residual valve disease were significantly reduced when penicillin prophylaxis was performed correctly⁶.

Carapetis et al.⁷ from Australia reported that Sydenham's chorea was common and associated with later development of rheumatic heart disease in 49 percent of patients, and they also showed the point prevalence of rheumatic heart disease to be 9.6 per 1,000.

Chen et al.⁸ found that 20 of 89 children with rheumatic fever had chorea, and six of them also had mitral insufficiency. Prompt diagnosis and treatment of streptococcal infection and penicillin prophylaxis was stressed.

Carditis may be found without any auscultatory finding in rheumatic fever. Echocardiography can demonstrate inaudible mitral regurgitation in nearly half of the patients with arthritis or chorea⁹. In another report, carditis was found in 36.6 percent of 87 cases of acute rheumatic fever, and was associated with arthritis in 35.3 percent¹⁰.

Carditis is the most serious cause of morbidity in acute rheumatic fever and is seen in about 50 percent of patients with the disease. Arthritis occurs in about 70 percent of patients with rheumatic fever and it is the least specific finding. Sydenham's chorea affects about 15 percent of patients with rheumatic fever¹. The period of latency between the initial streptococcal infection and the symptoms of chorea is about three months or more, much longer than that for arthritis or carditis, which is about three weeks¹¹. For this reason, all three major manifestations rarely occur together in rheumatic fever in the acute attack. Our patient had a rare combination of arthritis, carditis and chorea during the acute attack, after a short latent period of two weeks. The type of streptococcus may be related to the short latent period of the chorea. We conclude that the latent period of Sydenham's chorea may be as short as a few weeks, so this manifestation may occur with the positive laboratory findings of the acute attack, along with arthritis and carditis.

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