A Case of Acute Rheumatic Fever Accompanied by Transient Aortic Regurgitation

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SUMMARY

Though acute rheumatic fever (RF) is now rare in Japan, it continues to be an important disease condition that physicians should be prepared to diagnose and treat. We describe a patient with acute RF accompanied by transient aortic regurgitation (AR). The AR was detected only by echocardiography. There were no other indications, and it disappeared after treatment with prednisolone. The changes in cardiac valves in the early phase of RF have been the subject of only a few case studies. Echocardiography is quite valuable in the workup of patients with acute RF and should be performed even if there are no signs of cardiac involvement. (Jpn Heart J 2003; 44: 291-297)

Key words: Acute rheumatic fever, Aortic regurgitation, Jones criteria

ACUTE rheumatic fever (RF) is characterized by nonsuppurative inflammatory lesions involving primarily the heart, joints, subcutaneous tissues, and central nervous system. Rheumatic valvular heart disease remains the most common acquired heart disease in many developing countries. Here, we describe a young Japanese patient with RF accompanied by transient aortic regurgitation (AR), which was detected only by echocardiography and disappeared after treatment with prednisolone.

CASE REPORT

A 16-year-old boy was admitted to our hospital in July 2000 because of continuous fever and joint pain. He had been healthy until 5 weeks before admission, when he suffered tonsillitis. From that time, he showed a persistent low-grade fever that rose slightly each week. He complained periodically of asymmetric, migratory pain and swelling in his knee, ankle and wrist joints, and he had some difficulty walking.

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When he was admitted, he complained of severe pain in several joints, but there were no cardiac symptoms. His temperature was 40.2°C, his blood pressure was 110/60 mmHg, and his pulse rate was 80 beats per minute. Physical examination revealed tonsillitis and swelling of the ankle and knee joints with tenderness. There was no lymphadenopathy, erythema, or subcutaneous nodules. There were no audible rales. Cardiac auscultation revealed a grade I systolic ejection murmur. No neurologic abnormalities were found. Blood tests showed a white blood cell count of 14,790/µL, erythrocyte sedimentation rate of 104 mm/hr, and a C-reactive protein level of 13.95 mg/dL. The antistreptolysin-O titer was 716 IU/mL, and the antistreptokinase titer was over 20480 IU/mL. A throat culture was positive for group A hemolytic streptococci. The chest x-ray findings were normal. Electrocardiography revealed atrioventricular dissociation (Figure 1) without ST-T wave changes. Echocardiography (Figure 2A) showed trivial AR and mitral regurgitation without apparent valvular abnormalities. Both ventricle size and cardiac function were normal, and there was no pericardial effusion. A diagnosis of acute RF was made on the basis of the migratory polyarthritis and

**Figure 1.** Patient’s electrocardiogram on admission. Twelve-lead electrocardiogram (A) demonstrating no ST-T change. V1 lead (B) revealing atrioventricular dissociation.
Figure 2. Echocardiographically demonstrated aortic regurgitation. Echocardiogram on admission (A) demonstrating trivial aortic regurgitation. Echocardiograms 6 days after admission, para-sternal long axis view (B) and four chamber view (C), showing exacerbation of the aortic regurgitation. Ao = aorta; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.
evidence of streptococcal infection. Oral administration of aspirin at 2500 mg/day and benzylpenicillin benzathine (PC-G) at 1.2 million units/day was begun. The patient's temperature gradually decreased, and the polyarthritis resolved. The atrioventricular dissociation disappeared, and electrocardiography showed no prolongation of the PR interval. The heart murmur remained insignificant. However, 6 days after admission, echocardiography (Figure 2B, C) showed mild to moderate AR with a posterior change in blood flow direction. There were no apparent changes in the thickness or motion of the aortic valve. Oral prednisolone (PSL) was started at 40 mg/day instead of the aspirin (Figure 3). Repeat echocardiography showed gradual improvement in the AR. One month after admission, the patient was afebrile, and no AR was detected. During the course of the illness, there was no sign of heart failure. The PSL was decreased by 5 mg per week, and was withdrawn 8 weeks after administration. The patient was discharged on PC-G at 0.4 million units.

Figure 3. Clinical course. AR = aortic regurgitation; ASO = antistreptolysin-O; BT = body temperature; CRP = C-reactive protein; ESR = erythrocyte sedimentation rate; PC-G = benzylpenicillin benzathine; WBC = white blood cell.
ACUTE RF has become rare in developed countries. The World Health Organization reported an incidence of acute RF of less than 5 per 100,000 population per year in the industrialized world in the 1980s.\(^1\) The Japanese Ministry of Health, Labor and Welfare reported the annual mortality caused by rheumatic heart disease was about 2500 in the 1990s. The age pattern of mortality is concentrated in the elderly over 60 years old who were children or adolescents before 1945.\(^2\) The annual incidence of RF among Japanese schoolchildren is steadily declining as surveyed by pediatric clinics between 1952 and 1980.\(^3\) In contrast, the incidence of acute RF has remained alarmingly high in many developing countries.\(^1,4\) The importance of the disease was underscored during a resurgence of the disease in the United States in the mid-1980s, when a large number of cases occurred in many different states.\(^5-7\) Physicians should note that RF is still an important disease even in developed countries.

Cardiac involvement is the most important aspect of acute RF. RF produces pancarditis, affecting the endocardium, myocardium, and pericardium. Carditis is the only manifestation of acute RF that has the potential to cause long-term disability or death. Thus, the diagnosis of carditis in cases of active RF is critical. The Jones criteria, a clinical guideline introduced in 1944, is now widely used for the diagnosis of RF.\(^8\) The diagnosis of carditis in acute RF is based on the presence of significant apical systolic and/or basal diastolic murmur(s), the presence of pericarditis, or unexplained heart failure. Echocardiography is very useful for detection of subtle morphologic changes in the valve apparatus or valvular regurgitation,\(^9-13\) but the role of echocardiography is not adequately defined in the Jones criteria.\(^8,14\) Ramachandran,\(^13\) reported that the endocarditis involves the mitral valve more frequently than the aortic valve, and that isolated AR is unusual in the first acute RF attack. Our patient had no heart murmur or signs of heart failure that suggested carditis. Only echocardiography revealed the mild AR. Brand,\(^15\) and Minich,\(^16\) reported that trivial AR is frequently observed echocardiographically in the normal population. However, in our case, repeat Doppler echocardiography clearly indicated changes in the grade and direction of the AR. Therefore, we confirmed the presence of carditis associated with acute RF. B-mode echocardiography showed no changes in the aortic or mitral valves. Ozkutlu,\(^17\) reported silent valvitis in patients having RF in the absence of clinical evidence of cardiac involvement. Silent pathologic mitral regurgitation was found in 12 cases, and aortic regurgitation in 2 cases out of 26 patients. After a mean follow-up of 4.52 months, valvular regurgitation disappeared in 4 patients, including the one with migratory arthralgia and no other major criteria. It is possible that inflammation on the tips of the valves impaired
coaptation and caused the valvular regurgitation.\textsuperscript{13,18} Tomaru, \textit{et al}\textsuperscript{19} described that valvular regurgitations were caused by mitral and aortic valve prolapse. They postulated that myxomatous changes in those valves were caused by a post-inflammatory process associated with chronic inflammation, including those after RF. In the Utah outbreak of RF, Veasy, \textit{et al}\textsuperscript{7} echocardiographically demonstrated mitral regurgitation in 14 out of 19 patients with no abnormal murmur. Thus, echocardiographic study may be very important in the evaluation of patients with acute RF.

The American Heart Association recommends prophylaxis against recurrent acute RF.\textsuperscript{20} Acute rheumatic fever without clinical carditis is not a benign entity. If there is evidence of rheumatic heart disease, prolonged secondary prophylaxis is indicated; some authorities recommend lifetime prophylaxis. We will continue prophylactic oral penicillin in our patient as well as careful clinical and laboratory monitoring until he reaches 30 years of age.

\textbf{REFERENCES}