CASE REPORT

Adult-onset Acute Rheumatic Fever

Dainari Nakashima¹, Kohei Ueda¹, Kyozo Tsukuda¹, Noriaki Utsu¹, Shimazu Kohki², Hiroaki Fushimi² and Kazuho Miyakoshi¹

Abstract

A 62-year-old man was hospitalized for acute rheumatic fever. He had previously suffered from rheumatic fever at 15 years of age. The rheumatic fever was complicated by carditis, which caused valve disease that required surgical treatment. The incidence of rheumatic fever has decreased in most developed countries with improvements in sanitary conditions. The low incidence of this disease makes a timely and accurate diagnosis difficult. Due to the fact that both the first occurrence and recurrence of acute rheumatic fever can occur in the elderly and adults, this potential disease should not be overlooked when making a differential diagnosis.

Key words: acute rheumatic fever, carditis, developed countries, developing countries

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Introduction

The incidence of acute rheumatic fever has dramatically declined in most developed countries. However, acute rheumatic fever remains a major disease in developing countries, where it is common in both children and adults (1). Although rare, there are still reports of acute rheumatic fever in developed countries. While we found no reports documenting the incidence rate of adult-onset acute rheumatic fever, a few cases of this disease have been reported in Taiwan, Thailand, and the United States (2-4). Therefore, adult-onset acute rheumatic fever should not be overlooked when making a differential diagnosis.

Case Report

On November, 2009, a 62-year-old man visited a clinic in Osaka City, Osaka, with complaints of fever, sore throat, and joint pain that had appeared the day before. He was otherwise in good physical health. He was prescribed oral analgesics and unidentified antibiotics for a few days; however, the symptoms persisted. He visited another clinic (internal medicine) on November, where he was prescribed additional antibiotics (Levofloxacin 300 mg/day for five days). His condition still did not improve. As the joint pain was his primary symptom, he consulted an orthopedic specialist at Minami Osaka Hospital in Osaka City, Osaka on November. When he visited our office, he reported pain in his right shoulder joint and knee joints bilaterally. He also reported that he had experienced symptoms consistent with subcutaneous nodules prior to his visit. He was admitted to the hospital and prescribed oral non-steroidal anti-inflammatory drugs and antibiotics (cefazolin 4.0 g/day). On admission, no chorea, erythema, or other skin lesions were observed. On the 5th day after admission, he developed sudden dyspnea and cyanosis and was placed on a ventilator. He was then referred to the Department of Cardiovascular Internal Medicine. His relevant medical history included rheumatic fever 15 years of age and a subarachnoid hemorrhage 52 years of age.

A physical examination performed on the day of admission revealed a clear consciousness, a blood pressure of 108/76 mmHg, a heart rate of 98 beats per minute and a body temperature of 37.1°C. Auscultation of the heart revealed a Levine grade 5/6 systolic murmur at the apex, a Levine grade 5/6 diastolic murmur at the lower left sternal border, and a third heart sound at the apex. Coarse crackles were heard in the lung fields bilaterally. Laboratory test results indicated a C-reactive protein level of 31.5 mg/dL, a white blood cell count of 21,000/μL, a brain natriuretic peptide concentration of 397.8 pg/mL, an anti-streptolysin O con-

¹Department of Cardiovascular Internal Medicine, Minami Osaka Hospital, Japan and ²Department of Pathology, Osaka General Medical Center, Japan

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Correspondence to Dr. Dainari Nakashima, enomoto0728@yahoo.co.jp
concentration of 1,890 U/mL and a serum iron level of 26 μg/dL. Tests for anti-nuclear antibodies were negative, as were bacterial cultivation tests of sputum, blood and urine. During the initial six days of hospitalization, the patient had intermittent fever with temperatures above 37°C. After the 7th day, no fever symptoms were observed. The chest X-ray radiographs taken on admission were normal. However, on the 5th day, the patient’s condition suddenly changed, and X-ray radiographs indicated an increase in the cardiothoracic ratio (CTR) and bilaterally congested lungs. Electrocardiography performed on admission showed a first-degree atrioventricular block (Fig. 1a). Because transthoracic echocardiography showed hypokinesis of the left ventricular wall, severe aortic valve regurgitation, mild valvular thickening at the mitral valve and no findings at the aortic valve (Fig. 1b), we believed that valve disease had caused the acute heart failure and dyspnea. We administered an intravenous diuretic (Lasix, carperitide) in combination with catecholamines to relieve the cardiovascular hypotension. The patient’s condition improved thereafter and he was weaned from ventilator support on the 8th day after admission (Fig. 2). On the 6th day after admission, we diagnosed the patient with adult-onset acute rheumatic fever, as his symptoms met Jones’ criteria for acute rheumatic fever, including three major manifestations (carditis, polyarthritis and subcutaneous nodules), three minor manifestations (fever, prolonged PR interval on electrocardiogram and elevated C-reactive protein levels) and elevated anti-streptolysin O, thus indicating streptococcal infection (5). Beginning on the 6th day after admission, we administered 500 mg/day of intravenous methylprednisolone and 4.0 g/day of oral aspirin. On the 9th day after admission, we discontinued the methylprednisolone, replacing it with 60 mg/day of oral prednisolone (Fig. 2). The patient’s clinical condition and inflammatory markers gradually improved; however no improvements were seen in the valve disease. Therefore, after the patients recovered from the heart failure experienced on the 5th day after admission, we transferred him to the cardiovascular center for mitral and aortic valve replacement. The prolonged PR intervals persisted after surgery, and the resected valves showed no signs of inflammation (Fig. 3). The patient underwent cardiac valve surgery, and therefore showed a good recovery and was discharged from the hospital 17 days after admission.

**Discussion**

Based on the Jones criteria, we determined the case presented herein to be acute rheumatic fever with valve disease. Acute rheumatic fever can occur in the elderly, with initial attacks reported in patients 90 years of age (3) and 59 years of age (6). Recurrent attacks in patients older than 30 years of age, as was seen in our case, have also been reported (3), and rheumatic fever patients therefore have a lifetime possibility for recurrence. The histological findings of our patient’s resected valves indicated that the carditis had improved prior to surgery, while the aortic and mitral valve re-

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**Figure 1a.** Electrocardiography of a 62-year-old man with acute rheumatic fever indicating a first-degree atrioventricular block.

**Figure 1b.** Transthoracic echocardiography showing mild hypokinesis of the left ventricular wall, severe mitral valve regurgitation, severe aortic valve regurgitation and mitral valve thickening in a 62-year-old man with acute rheumatic fever.

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gurgitation did not change. Therefore, the patient’s valve disease was irreversible and it is likely that the congestive heart failure improved as a result of treatment with diuretics and catecholamines. Acute rheumatic fever is not typically considered in the differential diagnosis when patients visit clinics for treatment of fever or joint pain because the frequency of the disease is low. Even before the advent of antibiotics, the occurrence of acute rheumatic fever was decreasing in developed countries, including Japan, as a result of improvements in sanitary conditions and upgrades to medical care systems (7, 8). The adult-onset form of this disease is rare in developed countries; however, both the child-onset

**Figure 2.** Chest X-ray radiographs of the patient with acute rheumatic fever taken during the course of treatment. (A) The first chest X-ray radiograph on the left was taken on admission. The cardiothoracic ratio (CTR) was 55% and the lung fields were clear bilaterally. (B) The second radiograph on the left was taken on the 5th day after admission when the patient’s condition suddenly changed. The radiograph shows a wide CTR and bilaterally congested lungs. We began administering diuretics and dobutamine on the 5th day and steroids on the 6th day. (C) The second radiograph on the right was taken on the 8th day after admission, and showed an improved CTR and congested lungs. The patient was successfully extubated on the 8th day. (D) The first radiograph on the right was taken on the 13th day after admission and indicated a recovery from heart failure. We began administering piperacillin on the 13th day. The patient’s body temperature was approximately 38°C during the initial six days of hospitalization. The fever improved on the 7th day.

**Figure 3.** Left: The resected aortic valve with a fistula and a fissure (arrow). Right: No inflammatory cells were present in the resected valves.
and adult-onset forms are common in developing countries (9). The incidence of acute rheumatic fever was 0.4 per 1,000 in Japan in 1984 (10). The incidence of rheumatic heart disease was 0.14/1,000 in Japan in 1981 (11) and 1.0-5.4/1,000 in India in the 1980s (12). In the United States, there have been sporadic reports of small endemic areas of acute rheumatic fever since approximately 1980. According to these reports, throat pain is rare in patients with acute rheumatic fever, whereas carditis is frequent. The patients affected are primarily members of the middle class with comparatively easy access to medical treatment (4). We can speculate that rheumatic fever is sometimes not appropriately diagnosed because primary physicians often lack diagnostic experience and because poor patients often have difficulty gaining access to medical care. The rate of carditis as a complication of acute rheumatic fever is between 30% and 50% (13, 14). Therefore, we should consider acute rheumatic fever when diagnosing fever or joint pain and prescribe sufficient antibiotics to patients diagnosed with hemolytic streptococcal infections in order to prevent acute rheumatic fever (15). This is particularly true for patients with a history of acute rheumatic fever, as in the case presented herein.

The authors state that they have no Conflict of Interest (COI).

References


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