Chronic Chagas' Heart Disease in a Japanese-Brazilian Traveler

A Case Report

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SUMMARY

A 57-year-old Japanese-Brazilian man, visiting Japan for only 9 days, was admitted to our hospital due to syncope and frequent ventricular premature beats. He grew up in a rural area of Brazil and moved to Sao Paulo in 1959 when he was 20 years old. We suspected chronic Chagas' heart disease, ie., dilated cardiomyopathy with apical ventricular aneurysm, right bundle branch block with left anterior fascicular block, and various arrhythmias including supraventricular premature beats, ventricular premature beats and non-sustained ventricular tachycardia because he showed typical echo- and electrocardiographic features of the disease. Coronary arteriograms were normal, and left ventriculogram confirmed the existence of apical ventricular aneurysm. A left ventricle biopsy specimen showed hypertrophic cardiac muscle with mild fibrosis. The diagnosis of chronic Chagas' disease was finally confirmed by the demonstration of *Trypanosoma cruzi* itself in the blood as well as *Trypanosoma cruzi* antibodies. (Jpn Heart J 1999; 40: 375-382)

Key words: Chagas' disease, *Trypanosoma cruzi*, Dilated cardiomyopathy, Ventricular aneurysm, Right bundle branch block, Left anterior fascicular block

CHAGAS' disease, caused by the protozoan parasite *Trypanosoma cruzi*, is endemic over a wide area of South and Central America, where almost 90 million persons are at risk of contracting the disease and 16-18 million people are actually infected.¹⁾ Chronic cardiomyopathy, which appears 5-30 years after primary infection, is the most frequent clinical manifestation of the disease, affecting

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25–30% of infected individuals.²⁾ Chagas' disease is still a leading cause of left ventricular dysfunction in areas where the disease is endemic;³⁾ however, only a few cases of chronic Chagas' disease have been reported in Japan.^{4–6)} The number of visitors and immigrants from South America is increasing in Japan and, along with it, the likelihood of encountering more patients with chronic Chagas' disease. We report here the case of a Japanese-Brazilian traveler whose cardiomyopathy was diagnosed and confirmed as chronic Chagas' heart disease according to the typical clinical manifestations and by direct demonstration of *Trypanosoma cruzi* in the blood.

CASE REPORT

A 57-year-old Japanese-Brazilian man was admitted to Nagasaki University Hospital because of syncope. The patient was born in 1939 in Santa Crus do Riobarto, Brazil, about 500 km north of Sao Paulo, and spent his childhood there. He moved to Sao Paulo in 1959, where he has since been working as a carpenter. His family history was not remarkable, and he had no history of blood transfusion. He had felt well until the day of admission.

On August 29, 1996, he arrived in Japan to visit his brother. On September 6, while he was sitting on a chair, he suddenly lost consciousness, apparently

Figure 1. Electrocardiogram on admission showing an intraventricular conduction defect with a tendency toward low voltage in limb leads.

Figure 2. Supraventricular premature beats (SVP) and ventricular premature beats (VP) on the top tracing, and non-sustained ventricular tachycardia on the bottom tracing were documented in 24-hour monitorings.

without prodromal symptoms. Although he resumed consciousness in a couple of minutes, he sought medical attention, and frequent ventricular premature beats were detected by electrocardiography. He was transferred to our hospital for further examination.

On admission, his blood pressure was 114/78 mmHg. His body temperature was 37.2°C, his pulse was 54 beats/min, and his respiratory rate was 16 breaths/min. He showed clear consciousness. Examination of the heart revealed a third sound; the lungs were clear. The patient showed no lymphadenopathy, and his spleen was not palpable. There was no peripheral edema. Neurological examination was negative. The urine and stool were normal. Hematologic and other blood chemistry tests were normal. Electrocardiography showed a complete right bundle branch block and a left anterior fascicular block with normal sinus rhythm at a rate of 70 beats/min (Figure 1). Various arrhythmias including supraventricular premature beats, ventricular premature beats, and non-sustained ventricular tachycardias were observed in 24-hour monitorings (Figure 2). Chest radiography showed no pulmonary congestion or cardiac enlargement. Echocardiography revealed a dilatation of the left ventricle and poor left ventricular contraction with an apical aneurysm. The left ventricular end-diastolic dimension was 59 mm, the left ventricular end-systolic dimension was 47 mm,

Figure 3. Left ventriculogram. Right anterior oblique view of end-diastole (A) and end-systole (B) revealed a globally dilated and poor contractile left ventricle with an apical aneurysm.

Figure 4. Immuno-double diffusion test for antibodies to *Trypanosoma Cruzi*. Positive bands with titers of 256 were observed between *Trypanosoma cruzi* (trypomastigotes) antigen (Ag) and patient's serum (pt) as well as serums from infected controls (A and B). None = serums from non-infected controls.

Figure 5. Detection of *Trypanosoma cruzi* (epimastigotes) after 7 weeks of blood culture. Right is a magnified view of the red circled area on the left.

and the estimated left ventricular ejection fraction was 42%. Coronary arteriograms showed normal coronary arteries, while a left ventriculogram confirmed the apical aneurysm (Figure 3). The left ventricular ejection fraction was 40%. Histological examination of left ventricular endomyocardial biopsy specimens showed mild fibrosis and hypertrophic cardiomyocytes but no infiltration of inflammatory cells.

Although the patient was not aware of any infestation that could lead to Chagas' disease, we suspected it based both on the fact that he had lived in a high-risk area and on his clinical characteristics, the intraventricular conduction defect, the various arrhythmias, and the globally dilated and poor contractile left ventricle with the apical aneurysm. An immuno-double diffusion test performed for immunological examination of serum antibodies to *Trypanosoma cruzi* showed a positive band, with a titer of 256 (Figure 4). The diagnosis of chronic Chagas' disease was finally confirmed by direct demonstration of *Trypanosoma cruzi* itself in the blood of the patient after 7 weeks of blood culture using LIT medium (Figure 5).

DISCUSSION

The clinical manifestations of chronic Chagas' heart disease vary widely. Some patients are asymptomatic with nearly normal electrocardiograms, while others show advanced disease characterized by cardiomegaly, congestive heart failure, arrhythmias, thromboembolic phenomena, atypical chest pain, right bundle branch block, or even sudden death.⁷⁾ Although our patient had no cardiac symptoms until the day of admission, he showed clinical characteristics of advanced chronic Chagas' heart disease.

The lesioning in chronic Chagas' heart disease has been considered a type of dilated cardiomyopathy,³⁾ and autoimmune myocarditis, parasympathetic denervation and/or microvascular mechanism have been suggested in the pathogenesis of this lesion.⁷⁻⁹⁾ Chronic Chagas' heart disease has distinctive characteristics that could alter the prognosis of the disease unfavorably in comparison to that of non-Chagasic cardiomyopathy. Conduction abnormalities observed by electrocardiography, right bundle branch block and/or left anterior fascicular block, are frequently detected in chagasic patients³⁾ and can develop into complete atrioventricular block. Left ventricular apical aneurysm is found in 20-50% of patients^{3,7)} and has been associated with malignant arrhythmias and thromboembolic phenomena. Histologic examination usually reveals diffuse, multifocal myocytolytic necrosis, extensive fibrosis, and myocellular hypertrophy.^{7,10)} These morphologic changes are not specific, but they are more severe than those observed in non-chagasic cardiomyopathy and may lead to more severe ventricular remodeling in chagasic patients. In fact, it has been reported that chagasic patients with mild to moderate left ventricular dysfunction have a poorer clinical course than patients with similar non-chagasic cardiomyopathies.³⁾ Endomyocardial biopsy findings in our patient were non-specific and did not show the myocytolytic necrosis or intracellular parasites.

Malignant ventricular tachyarrhythmias are the major causes of sudden death and/or syncope among patients with chronic Chagas' heart disease.¹¹⁾ Sudden cardiac death is found more frequently in chagasic patients than in non-chagasic cardiomyopathy patients,³⁾ which might be the consequence of the higher frequency of left ventricular apical aneurysm and right bundle branch block with left anterior fascicular block detected in these patients. Indeed, it has been reported that, in chagasic cardiomyopathy, ventricular tachycardia induced by electrophysiological testing is most common in patients showing conduction abnormalities on the electrocardiogram, low ejection fraction, and left ventricular apical aneurysm.^{11,12)} Further, syncope and sudden death due to malignant arrhythmias develop even before cardiomegaly or heart failure.¹³⁾ Because conduction abnormalities, left ventricular apical aneurysm, frequent ventricular pre-

mature beats, as well as non-sustained ventricular tachycardias were documented in our patient, the syncope in this case may have been due to the ventricular tachyarrhythmia.

In addition to its distinctive clinical features, a diagnosis of chronic Chagas' heart disease is made by several serologic tests for the detection of anti-*Trypanosoma cruzi* antibodies,⁷⁾ although false-positive reactions remain a persistent problem with these assays.¹⁴⁾ In this case, the serologic diagnosis was made by the positive immuno-double diffusion test for anti-*Trypanosoma cruzi* antibodies. Diagnosis was finally confirmed by direct demonstration of *Trypanosoma cruzi* in the blood.

Treatment with anti-*Trypanosoma cruzi* drugs does not abort the progression of chronic Chagas' heart disease, and vaccines are not available.¹⁴⁾ Thus, major efforts are aimed at preventing and interrupting transmission of the parasite. In non-endemic areas such as the United States, the presence of carriers of *Trypanosoma cruzi* poses a significant risk of transmission by blood transfusion.^{1,14)} In addition, it has been suggested that sera from chronically infected rabbits produce acute electrocardiographic abnormalities in conduction, and humoral factors may contribute to the conduction defects in chronic Chagas' heart disease patients.¹⁵⁾ Thus, it is important to note that, even in Japan, blood donations should be accepted from immigrants from regions in which Chagas' disease is endemic only when serologic assays indicate that the donor is not infected with *Trypanosoma cruzi*.

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