Case Report

A Case of Atrial Septal Defect Complicated by Autoimmune Hemolytic Anemia

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Open heart surgery is rarely performed on patients with heart disease complicated by autoimmune hemolytic anemia (AIHA) with the aid of an artificial heart-lung machine. We successfully performed radical open heart surgery on an atrial septal defect (ASD) patient with the complication of AIHA. (Ann Thorac Cardiovasc Surg 2004; 10: 120–3)

Key words: AIHA, CPC

Introduction

Open heart surgery has rarely been performed on patients with autoimmune hemolytic anemia (AIHA) like collagen disease in large part because of a low frequency of complications. It is said that AIHA occurs in 3.4 of every million people, and cardiac surgery is indicated in very few AIHA patients for two main reasons. Firstly, it is not known whether cardiac surgery can be safely performed on AIHA patients. Secondly, the acute aggravation of the underlying disease may be triggered by surgery.[1-3)]

Case Report

A 62-year-old woman (housewife) visited our hospital with chief complaint of dyspnea on effort. She was diagnosed with heart disease when she was in the fifth grade of primary school and developed arrhythmia in her 20s. She received surgery for hysteromyomectomy at the age of 45 years. She was diagnosed as having atrial septal defect (ASD) at the age of 45 years but remained asymptomatic thereafter. The dyspnea on effort appeared at the age of 62. Her height was 142 cm and weight 44 kg. Her blood pressure was 120/70 mmHg and pulse rate 80/min, irregular. Anemia was observed in the palpebral conjunctiva, but jaundice was not noted. A Levine grade III systolic murmur was audible most intensely at the left edge of the third intercostal sternum. Neither hepatomegaly nor splenomegaly was noted, but edema was observed in the lower extremities. In hematological findings, normocytic and normochromic anemia was present as reflected in red blood cell (RBC) of 2.83×10^6, hemoglobin (Hb) of 9.4 g/dl, hematocrit (Ht) of 27.7%, mean corpuscular volume (MCV) of 97.8 μm^3, mean corpuscular hemoglobin (MCH) of 33.8 pg, and mean corpuscular hemoglobin concentration (MCHC) of 34.1%. The reticulocyte count was increased to 38%. Total bilirubin (T-Bil) was increased to 2.2 mg/dl, but direct reacting bilirubin was normal, at 0.8 mg/dl. It appeared that the increase in T-Bil was accounted for by indirect reacting bilirubin. Type I lactase dehydrogenase (LDH) was increased to 42.8%. In immunoserologic findings, both direct and indirect Coomb's tests were positive. The antibody was immunoglobulin G (IgG), and complement binding was not observed. The heptoglobulin level was decreased to 49.8 mg/dl. Other autoantibodies were not detected. Based on the above findings, a diagnosis of AIHA was established. Physical testing of the erythrocyte membrane[4)] (Coli plant centrifuge: CPC) was shown in Fig. 1. Both hemolysis endpoint and maximum point were elevated in CPC. The osmolarity of the erythrocyte membrane was depressed. These findings suggested that the patient was prone to hemolysis. Cardiac dilatation was observed as reflected in cardiothoracic ratio (CTR) of 70%, and pulmonary blood vessels were enhanced in the chest X-ray (Fig. 2). Atrial fibrillation, incomplete right bundle block and biventricular hypertrophy were observed on the electrocardiogram (ECG) (Fig. 3). Preoperative echocardiographic examination revealed the following changes (Fig.

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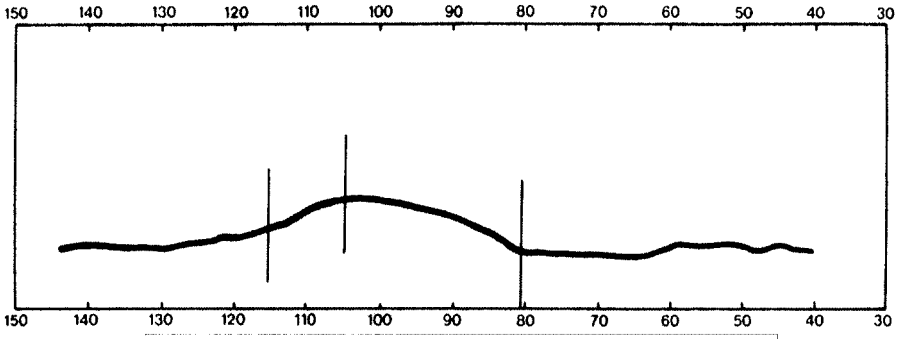

HEP: Hemolysis end point	81mOsm
HMP: Hemolysis maximum point	104mOsm
HSP: Hemolysis start point	115mOsm
HW: Hemolysis width	34mOsm

Fig. 1. Physical testing of the erythrocyte membrane (Coli plant centrifuge: CPC).
These findings suggested that the patient was prone to hemolysis.

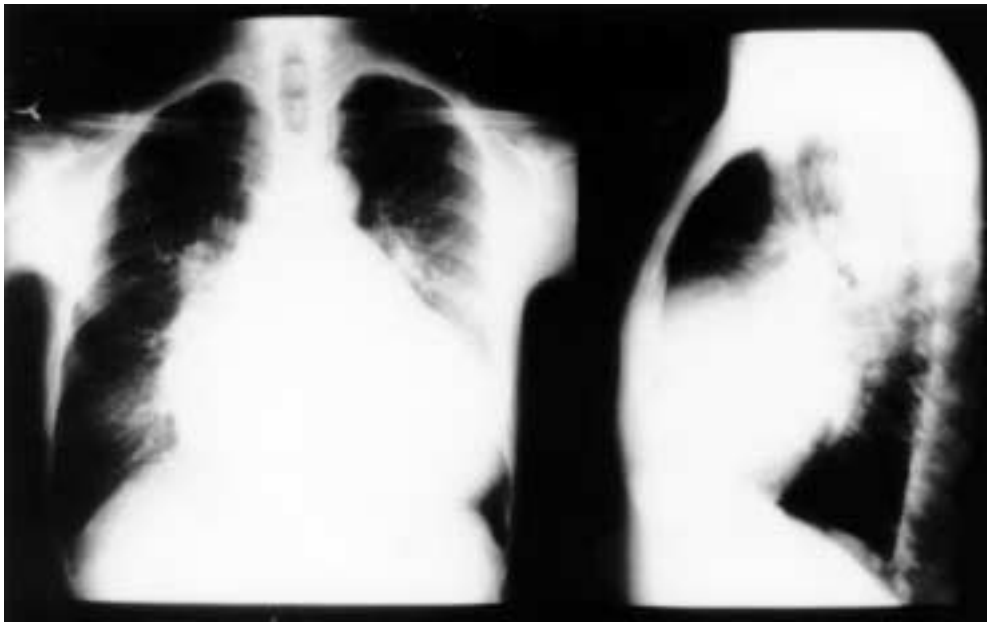

Fig. 2. Chest X-ray findings. CTR of 70%, and pulmonary blood vessels were enhanced.

4). M-mode recordings: pseudo systolic anterior motion (SAM) of the mitral valve leaflet and paradoxical movement of the septum. Color Doppler recordings demonstrated the left-to-right atrial shunt at a high septal level. Cardiac catheterization revealed a pulmonary arterial wedge pressure of 52/13 mmHg and an increase in blood oxygen saturation in the right atrium, and the rate of left-to-right shunt was 53.3%, and Qp/Qs was 2.3. The clinical course after the diagnosis of AIHA is shown in Fig. 5. The Coomb's test reverted to positive after one month of treatment with prednisolone 40 mg. The patient was treated at a reduced dose of 20 mg for two months and was maintained at a dose of 10 mg thereafter. As Hb remained at 11.0 g/dl, radical surgery was performed eight months after the diagnosis of AIHA was made. The dose of prednisolone was increased to 40 mg one week before operation, and washed red cells were used during operation. ASD was fossa ovalis type 1 measuring 28 mm by 20 mm. It was closed with a Xonomedica patch. The tricuspid annulus with 35 mm or larger dilation was sutured by Kay's method. After two weeks of treatment prednisolone was decreased in 10 mg decrements. The patient was discharged six weeks after operation.

Postoperative echocardiographic findings showed that

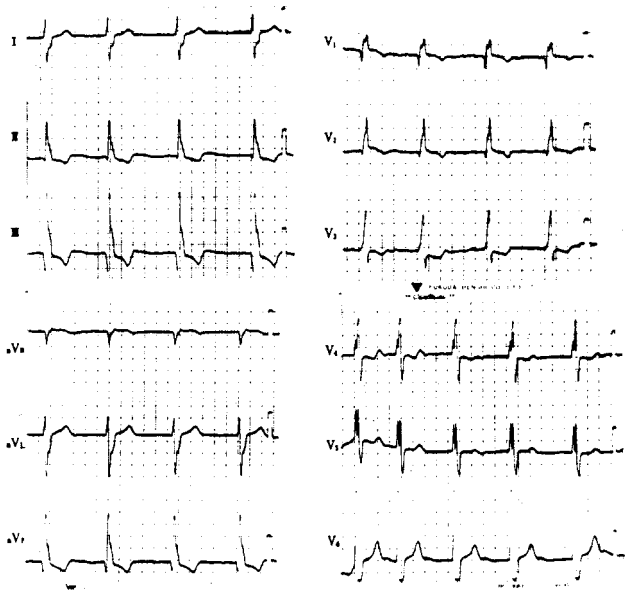

Fig. 3. ECG findings.
Atrial fibrillation, incomplete right bundle block and biventricular hypertrophy were observed on the ECG.

both pseudo SAM of the mitral valve leaflet and paradoxical movement of the septum were absent. The dilation of the tricuspid annulus was reduced to a size of 27×25 mm. Color Doppler recordings showed no shunt of blood.

Discussion

In our patient, however, surgical intervention was successful both presumably because long-term steroid therapy for AIHA inhibited hemolysis and because washed red cells were used during operation.[5)]

In patients with AIHA, caution is required due to (1) increased hemolysis caused by transfusion and (2) the risk of new antibody production.[6)] To date, blood transfusion has been avoided and corticosteroid (prednisolone) therapy has been implemented,[7)] and washed red cells have been used in patients with positive direct and indirect Coomb's test results for complement in the preparation. The introduction of autotransfusion, reduction of hemorrhage through the use of hypotensive anesthesia, and an-

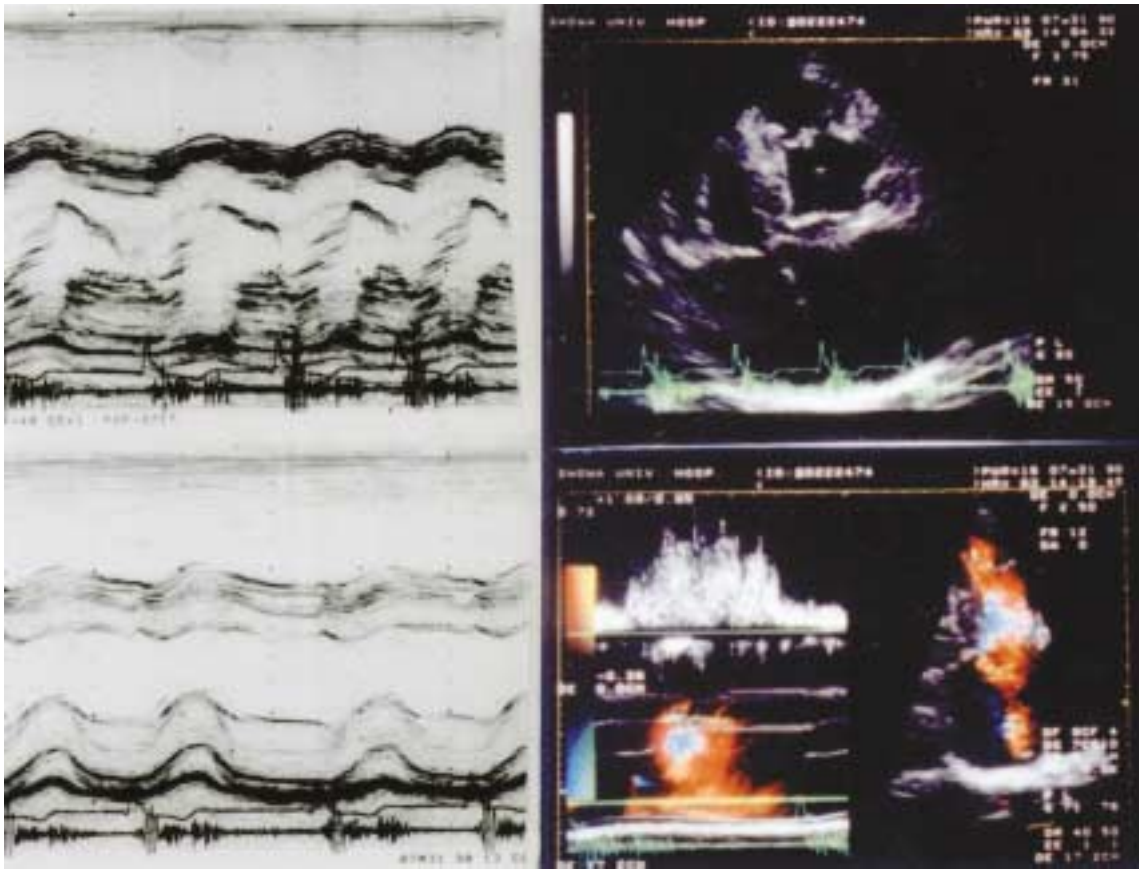

Fig. 4. Preoperative echocardiographic findings.
M-mode recordings: pseudo SAM of the mitral valve leaflet and paradoxical movement of the septum, color Doppler recordings: left-to-right atrial shunt at high septal level.

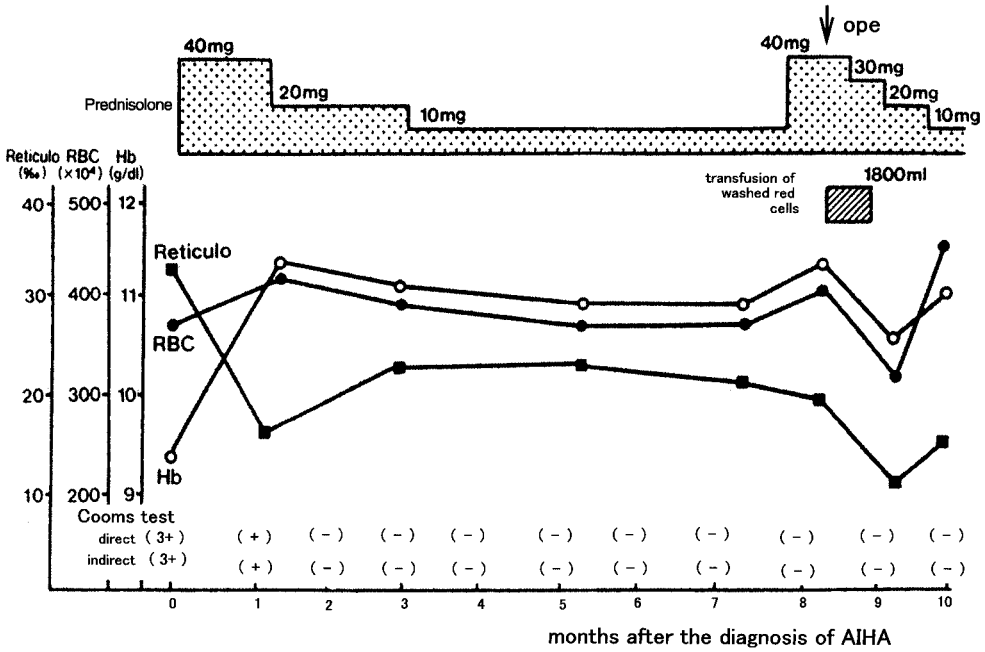

Fig. 5. Clinical course.
The dose of prednisolone was increased to 40 mg one week before operation, and packed red cells were used during operation.

tigen-negative blood transfusion have also been used after surgery in an attempt to eliminate the above problems. However, due care is still needed when performing surgery and blood transfusion in patients with the disease.

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