Hemolysis and infective endocarditis in a mitral prosthetic valve

Mitral protez kapakta hemoliz ve enfektif endokardit

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Traumatic intravascular hemolysis after heart valve replacement can be a serious problem. It is commonly associated with either structural deterioration or paravalvular leaks. A 63-year-old woman with a six-year history of surgery for mitral stenosis presented with complaints of weakness and dyspnea. She received treatment at other centers three times in the past six months for dyspnea and anemia requiring transfusion of red blood cells. Transthoracic echocardiography showed a normally functioning mitral mechanic prosthesis. Laboratory findings were abnormal for hemoglobin, hematocrit, white blood cell count, C-reactive protein, serum haptoglobin, and lactate dehydrogenase. Peripheral blood smear showed marked schistocytes, indicative of mechanical erythrocyte destruction. Transesophageal echocardiography demonstrated severe paravalvular leak and a large (9x13 mm) vegetation adhering to the prosthetic valve, protruding into the left atrium. *Enterococcus faecalis* was isolated from blood cultures. Surgery was planned because of large vegetation, repeated hemolysis, and severe paravalvular regurgitation, but the patient refused surgical treatment.

Key words: Anemia, hemolytic; endocarditis, bacterial/complications; heart valve prosthesis/adverse effects; hemolysis.

Kalp kapağı değişiminden sonra gelişen travmatik intravasküler hemoliz ciddi bir sorundur. Genellikle yapısal bozulma veya paravalvüler kaçaktan kaynaklanır. Mİtral darlık için altı yıl önce ameliyat olmuş 63 yaşında kadın hasta halsizlik ve nefes darlığı yakınmalarıyla başvurdu. Son altı ay içinde hasta üç kez, nefes darlığı ve kırmızı kan hücresi transfüzyonu gerektiren anemi nedeniyle başka merkezlerde tedavi görmüştü. Transtorasik ekokardiyografide mitral mekanik protez kapağın normal çalıştığı görüldü. Laboratuvar bulgularından hemoglobin, hematokrit, beyaz hücre sayımı, C-reaktif protein, haptoglobin ve laktat dehidrogenaz değerleri anormal bulundu. Periferik kan yaymasında, mekanik eritrosit yıkımı için gösterge olan parçalanmış eritrositler görüldü. Transözofageal ekokardiyografide ciddi paravalvüler kaçak ve protez kapakta gelişen ve sol atriyum içine sarkan büyük vejetasyon (9x13 mm) saptandı. Kan kültürlerinde *Enterococcus faecalis* üredi. Büyük vejetasyon, tekrarlayan hemoliz ve ciddi paravalvüler yetersizlik nedeniyle hastaya cerrahi planlandı; ancak, hasta cerrahi tedaviyi kabul etmedi.

Anahtar sözcükler: Anemi, hemolitik; endokardit, bakteriyel/komplikasyon; kalp kapağı protezi/yan etki; hemoliz.

Traumatic hemolysis is a potentially serious problem after heart valve replacement. Mild degrees of intravascular hemolysis are common among patients with mechanical prostheses, but red blood cell damage is more pronounced with malfunctioning than with properly working prostheses.[1,2] We report a patient with infective endocarditis, who received multiple blood transfusions due to hemolytic anemia.

CASE REPORT

A 63-year-old female patient presented to our clinic with complaints of weakness and dyspnea of six-month history. She had night sweating, but she did not have fever. She had a six-year history of surgery for mitral stenosis and was admitted to other centers three times in the past six months with dyspnea and received transfusion of red blood cells with the diagnosis of anemia. On physical examination, blood pressure was 110/70 mmHg, heart rate was 90 bpm and irregular. A mechanic valve sound was heard on the mitral valve area. The electrocardiogram showed atrial fibrillation with normal ventricle response. Her temperature was 37.2 °C. On transthoracic echocardiography, functioning of the mitral mechanic prosthesis was normal. Ab-

Received: October 5, 2009 *Accepted:* December 18, 2009

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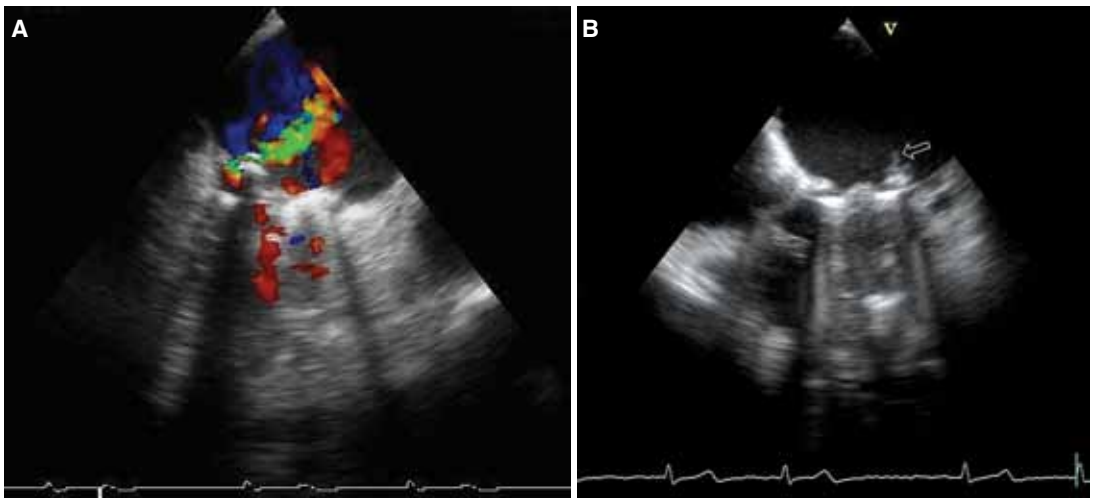

Figure 1. Transesophageal echocardiograms showing **(A)** lateral periprosthetic mitral regurgitation and **(B)** a large vegetation on the mitral valve.

normal laboratory findings were as follows: hemoglobin 8.5 g/dl, hematocrit 25%, white blood cell count 17,400/mm^3, C-reactive protein 74.2 mg/dl (normal 0-5 mg/dl), serum haptoglobin <6 mg/dl (normal 30-200 mg/dl), and lactate dehydrogenase 782 U/l (normal 135-214 U/l). Serum iron, ferritin, vitamin B_{12} and folic acid levels were within normal ranges. Peripheral blood smear showed marked presence of schistocytes, indicative of mechanical erythrocyte destruction. There was no hematuria in simple urine tests. The last INR analyzed was 3.94. Transesophageal echocardiography demonstrated severe paravalvular leak (Fig. 1a) and a large (9 x 13 mm) vegetation adhering to the anterior aspect of the prosthetic valve sewing ring, protruding into the left atrium (Fig. 1b). Blood cultures were taken and empiric antibiotic treatment was initiated. *Enterococcus faecalis* was isolated from all three blood cultures after seven days of incubation. Surgery was planned because of large vegetations, repeated hemolysis, and severe paravalvular regurgitation, but the patient refused treatment.

DISCUSSION

Complications after valve replacement are multiple and include thromboembolism, paravalvular leaks, valve dehiscence, infective endocarditis, and hemolysis.[3] Traumatic intravascular hemolysis after heart valve replacement can be a serious problem. It is commonly associated with either structural deterioration or paravalvular leak.[4] It is reported to occur in 5% to 15% of patients with a ball-cage valve prosthesis, but in most cases is of mild degree and subclinical.[5] Serious hemolysis is rare, but commonly reflects paravalvular leak. The main mechanism is a turbulent flow through the valve or between the sewing ring and the native ring.[4] As suggested by Skoularigis et al.,[6] patients are considered to have intravascular hemolysis under the following conditions: serum lactate dehydrogenase levels greater than 460 U/l and any two of the four criteria including blood hemoglobin <13.8 g/dl for males and <12.4 g/dl for females, serum haptoglobin <50 mg/dl, reticulocyte count >2%, and presence of schistocytes in the peripheral blood smear (normally absent). Hemolysis is probably due to the turbulence of flow with high shear-stress forces and abnormal flow jets through the prosthetic valve. Many factors have been found to influence the degree of hemolysis: site of implant, prosthetic design, size of prosthesis, number of prostheses implanted, presence of atrial fibrillation, heart rate, and paravalvular leaks.[7] With the advent of modern mechanical prosthetic heart valves, clinically significant hemolysis has become relatively rare and occurs mainly with malfunctioning valves accompanied by paravalvular regurgitation. Hemolysis with paravalvular regurgitation results in anemia, and persisting hemolysis can cause organ dysfunction such as renal failure. Detection of subclinical perivalvular regurgitation is important because it contributes to the degree of intravascular hemolysis.[8] Hemolytic anemia is a rare manifestation of infective endocarditis. It has been reported in only a few case reports.[9]

Patients with hemolytic anemia should be given iron and folate supplementation, possibly with blood transfusions.[10] Beta-blocker therapy can be used as well. Its main mechanism is reduction in shearing forces acting on erythrocytes. Pentoxifylline can also be used to minimize hemolysis probably by increasing

erythrocyte durability.[4] Valve replacement should be considered in the presence of severe hemolytic anemia not responding to medical therapy.[10] Reoperation is recommended in the guidelines for paravalvular leaks leading to severe symptoms or hemolytic anemia. The mortality risk of reoperation is about 10%, being much higher than that of the first operation. Because of the increased mortality risk of reoperation, percutaneous closure of paravalvular leaks is an alternative to surgery. Contraindications for percutaneous closure of paravalvular leaks are ongoing infection and the presence of vegetation or thrombus.[4] Thus, in spite of increased mortality risk, reoperation is the optimal treatment option in paravalvular leaks resistant to medical treatment and with large vegetations leading to hemolytic anemia.

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