Severe hemolytic anemia after repair of primum septal defect and cleft mitral valve

Dursun Alehan[1], Rıza Doğan[2], Süheyla Özkutlu[1], Huda Elshershari[1], Fatma Gümrük[3]

Units of [1]Cardiology, and [2]Hematology, Department of Pediatrics and [3]Department of Cardiothoracic Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey

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Two cases are described in which severe mechanical hemolytic anemia developed after surgical repair of primum atrial septal defect (ASD) and cleft mitral valve. In both cases there was residual mitral regurgitation after repair. Moderate mitral regurgitation and collision of the regurgitant jet with the teflon patch used for repair of the primum ASD were detected by color-Doppler echocardiography imaging. Laboratory tests showed normochromic normocytic anemia, increased indirect serum bilirubin, decreased plasma haptoglobin and hemoglobinuria. The peripheral blood smear contained numerous fragmented red cells. Following another surgical correction of the mitral valve (repair or mitral valve replacement), there was no more hemolysis. The two presented cases show that foreign materials in association with localized intracardiac turbulence may cause severe hemolysis.

Key words: hemolytic anemia, primum ASD, cleft mitral valve, mitral regurgitation.

Hemolytic anemia is an infrequent but well documented complication following the insertion of intracardiac or intravascular prosthetic materials. It was first reported by Sayed et al.[1] in a patient who had closure of an ostium primum atrial septal defect with a teflon patch. Hemolytic anemia has also been reported after mitral valve repair[2-9]. With the improvement of surgical materials and techniques, this complication has recently become exceptionally rare. We describe two children who developed severe hemolysis after repair of primum atrial septal defect (ASD) and cleft mitral valve in association with mitral insufficiency. The anemia was corrected by mitral valve replacement or repair.

Case Report

Case 1

An eight-year-old girl diagnosed with a primum ASD and cleft mitral valve underwent an intracardiac repair through median sternotomy using a teflon patch closure of the primum ASD and stitch closure of the cleft in the anterior leaflet of the mitral valve according to Rowlatt formula[10]. Postoperative transthoracic echocardiography revealed good biventricular function and moderate mitral valve regurgitation, but no evidence of residual ASD.

The patient was well until four months postoperatively when she was admitted to our hospital with history of fatigue, lethargy and loss of appetite. Evaluation revealed jaundice without fever or lymphadenopathy. She had an enlarged liver palpable 4 cm below the right costal margin at the midclavicular line. On cardiac examination a regular rhythm, tachycardia and pansystolic murmur over the mitral area were detected.

Laboratory studies revealed a hemoglobin level of 5.0 g/dl. The leukocyte count was 9600/mm^3 with a normal differential count, the platelet count was 242,000/mm^3, red cells were 1.9 x 10^{12}/L, reticulocyte count was 4.4%, and haptoglobin level was low. The peripheral blood smear showed a number of red cell fragments, spherocytes, normoblasts and marked polychromasia. The patient's serum total and direct bilirubin were 6.24 and 0.63 mg/dl, respectively, and lactic dehydrogenase was 5045 IU/L. The urine contained increased amounts of urobilinogen and urobilin. Coombs' test was negative. Transthoracic

color-Doppler echocardiography showed a moderate mitral regurgitation with a central regurgitant jet directly striking the teflon patch. The diagnosis of mechanical severe hemolytic anemia due to mitral regurgitation was suspected. Despite a trial of medical treatment and blood transfusions, the anemia persisted and it was decided to reoperate. The mitral valve was found to be severly deformed; therefore, valve replacement was carried out. The postoperative course was unremarkable. The patient had an uneventful recovery with no further blood transfusion, and hemolytic anemia was cured. The hemoglobin level was 13.8 g/dl and all other tests for hemolysis reverted to normal.

Case 2

A three-year-old girl developed severe hemolytic anemia following surgical correction of a partial atrioventricular canal and closure of a mitral cleft. The patient underwent closure of the ASD with a teflon patch and the mitral cleft was repaired with four sutures.

Two months postoperatively, the patient was admitted to the hospital with severe hemolytic anemia (hemoglobin 6.0 g/dl). All laboratory investigations performed revealed evidence of hemolytic anemia. Transthoracic echocardiography showed good shape and satisfactory function of the reconstructed mitral valve. However, color-Doppler echocardiography revealed a small eccentric high-velocity jet of mitral regurgitation originating from the anterior leaflet of the valve and striking the teflon patch. Reoperation was required for control of the uncompensated hemolysis.

Intraoperative exploration of the mitral valve showed leaking through the stitches of the anterior leaflet. In this case, clinically manifest hemolysis ceased after the mitral valve repair.

Discussion

Hemolytic anemia in association with congenital heart disease is rare and clinically significant. Hemolysis following cardiac surgery is uncommon. Sayed and colleagues[1] described a patient who developed severe intravascular hemolysis after teflon patch closure of a partial atrioventricular canal. At reoperation, a regurgitant jet of blood through a mitral valve cleft was directed against a bare patch of teflon. Covering the teflon with endocardium eliminated the hemolysis. Similar patients were described by Verdon[11] and Sigler et al.[12]. It was concluded that the hemolysis was due to excessive local blood velocity with altered intracardiac morphology resulting in critical turbulence and red cell damage.

Moisey et al.[5] reported a case of severe hemolytic anemia in association with hemodynamically moderate mitral regurgitation. In this case, valve replacement eliminated the hemolysis. Stoschitzky and colleagues[7] reported a patient with hemolytic anemia after mitral valve repair who eventually required valve replacement. The patient had a high velocity jet of mitral regurgitation that struck at a right angle the opposite wall of the left atrium, although the amount of regurgitation was assumed to be hemodynamically insignificant.

A recent report by Yeo et al.[13] suggested that hemolysis after mitral valve repair is due to the high shear stress produced by the regurgitant jet, independent of the severity of mitral regurgitation, and that the most commonly observed mechanism of hemolysis involved direct collision of the regurgitant jet with a prosthetic surface. In vitro studies have domonstrated that shear forces > 3000 dynes/cm^2 are associated with significant red cell destruction[14].

In the present two cases, the most likely source of the hemolysis was the residual mitral regurgitant jet which probably erodes the endothelium, thereby exposing a portion of the teflon surface and hence introducing the risk of hemolysis occurring at that site. This report demonstrates that severe hemolysis may also be caused by mitral regurgitation without hemodynamically significant dysfunction. We suggest that mitral valve repair and mitral regurgitation should be considered in the differential diagnosis of hemolytic anemia even without significant hemodynamic effects. In conclusion, the present two cases confirmed other reports that in the presence of severe hemolytic anemia, a mechanical cause should be investigated. The peresence of cardiac surgery and of intracardiac foreign material makes this diagnosis more likely, and the definitive treatment is surgical repair of the anatomic defect.

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