SEVERE HEMOLYSIS FOLLOWING MITRAL VALVE REPAIR

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INTRODUCTION

Hemolysis and valvular heart have been linked for more than 40 years. In 1961, Sayid and associates reported the first case of hemolysis in a patient with ostium primum atrial septal defect and unrepaired mitral cleft.1 Mitral valve repair (MVR) is now a common management strategy for severe mitral regurgitation, a procedure that carries the rare complication of intravascular hemolysis. Between 1981 and 2004, only 33 patients have been reported to present with hemolytic anemia after MVR.[2,3] We present two of them.

CASE REPORTS: CASE #1

A 46-year-old Hispanic female presented co the emergency room with complaints of severe crushing substernal chest discomfort radiating co the neck, back and left arm, shortness of breath, paroxysmal nocturnal dyspnea and orthopnea. Vital signs were stable, her rhythm was atrial fibrillation, and routine laboratory studies were normal. Myocardial infarction was ruled out. A transthoracic echocardiogram revealed an enlarged left ventricle with ejection fraction of 35-39% and severe mitral regurgitation associated with a myxomatous mitral valve. Coronary angiography demonstrated a 60% left anterior descending stenosis. Urgent surgery was perfurmed to repair the mitral valve using a 27 mm St. Jude full-ring mitral valve annuloplasty accompanied by a left internal mammary artery anastomosis to the left anterior descending artery. A postoperative echocardiogram showed no significant mitral regurgitation and a persistent ejection fraction of 35-39%. Coumadin was starred for her atrial fibrillation. She was discharged in stable condition two weeks after surgery.

Two weeks later, the patient presented co the surgical clinic with nausea, fatigue, abdominal discomfort and yellowish skin discoloration. Her rhythm was atrial fibrillation. Vital signs were normal. An apical pansystolic murmur was recorded. Pertinent laboratory studies included: hemoglobin 7.7 g/dL, hematocrit 22, total bilirubin 8.0 mg/dL, direct bilirubin .9 mg/dL, LOH 1911 u/L, haptoglobin 42 ml/dL and erythrocytosis 3%. Hepatitis panel, HIV and Coombs studies were negative. The presumptive diagnosis was intravascular hemolysis secondary to mitral valve regurgitation. Transthoracic and transesophageal echocardiograms showed modest LV enlargement, ejection fraction of 45-49% and severe mitral regurgitation. An eccentric jet was directed posteriorly and thought related to incomplete leaflet coaptation with a restrictive movement of the posterior mitral leaflet.

She underwent a second repair of the mitral valve with the help of an imraaortic balloon pump via the left femoral artery. During surgery, a small area of leakage at the posterior medial commissure was confirmed with no obvious defect in the previous repair. A single stitch was used to plicate approximately 1.0 cm of the posterior medial portion of the posterior annulus. There were a variety of postoperative complications from which she completely recovered. A repeat echocardiogram just prior to discharge revealed modest enlargement of the left ventricle with an ejection fraction of 40-44% and a trace of mitral regurgitation. Subsequently, there was no further evidence of intravascular hemolysis and her remaiming recovery was uneventful.

CASE #2

A 54-year-old male was transferred from an outlying hospital with complaints of fatigue, retrosternal chest tightness, shortness of breath, paroxysmal nocturnal dyspnea and orthopnea. Past history included hyperrension and remote atrial fibrillation. The initial physical examination reported normal sinus rhythm of 68 per minute, blood pressure of 130/96 and an apical pansysmlic murmur. Normal values were obtained for CBC, BUN, creatinine, electrolytes, blood sugar and liver panel. A transthoracic echocardiogram found severe mitral regurgitation from a flail anterior mitral leafier, moderate enlargemem of the left ventricle, estimated pulmonary artery pressure of 43 mmHg and a LV ejection fraction of 70%. Coronary angiography reported left main stenosis of 80-90%, a normal circumflex and a normal right coronary artery.

He underwent MVR by artificial cord insertion (times-4) to the anterior micral leaflet and a full ring annuloplasty with a 29 mm St. Jude ring, plus myocardial revascularization with a left internal mammary artery co the left anterior descending artery and a reverse saphenous vein graft to the obtuse marginal branch. Pulmonary vein isolation with XcraCare radio frequency was performed. A postoperative transthoracic echocardiogram showed mild mitral regurgitation, ejection fraction of 65-69% and an eccentric mitral regurgitant jet directed anteriorly. Postoperative recovery was uneventful, and he was discharged on the eleventh

day poscoperacively.

Six weeks poscoperacively che paciem presemed wich increasing facigue and worsening exertional dyspnea. He was anemic and jaundiced and described his urine as being cola-colored. Physical examination showed normal rhythm (54/min.) and blood pressure ac 130/88. A grade 1 systolic murmur along the left sternal border was recorded. The eleccrocardiogram showed T-wave inversions from V2-V6 and AVL. The hematocrit was 21.5%. Platelets were 249,000, LDH was 2200, Coombs tesc was negative and haproglobin was 6. Peripheral blood smear showed polychromasia, schiscocyces and marked anisocyrosis. A uansthoracic echocardiogram showed moderate mitral valve regurgication and normal LV size with ejection fraction of 64%. A moderace plural effusion, chickened mitral valve leaflets and artificial chordae were seen, and LV filling pressure was increased. A subsequent transesophageal echocardiogram confirmed a normal ejeccion fraction, normal LV chamber size, severe enlargement of the left atrium with normal righc atrial size, and severe mitral regurgication. There was a suggestion of a perforacion or a flail segmem at the medial corner of che micral valve apparatus. Pulmonary artery pressure was 32 mmHg. The clinical impression was that hemolytic anemia had developed due ro MVR dysfunction.

The patient subsequemly required a follow-up mitral valve replacement using a #25 St. Jude expanded cuff valvular prosthesis. There were no posroperacive complications. Thehemolysisdisappeared over the next week, and the patient was subsequently discharged without further incident. No further hemolysis was reported.

DISCUSSION

Hemolytic anemia after mechanical mitral valve replacement is a well recognized bur infrequent complication attributable to traumatic fragmentation of erythrocytes by prosthetic material or by paravalvular jet shear forces.4 It is much less common with tissue or porcine valves. While MVR is an established treatment for micral valve insufficiency, severe mechanical intravascular hemolysis is a rare cause of morbidity and mortality after MVR.5 There are many theories for this rare occurrence, all in the category of intravascular trauma: whiplash motion of disrupted sutures; a dehisced annuloplasty ring producing para-ring regurgitant jets; protruding paravalvular suture material or piedgees that provide an impact site against circulating red blood cells; nonendothelialization of foreign material such as sutures or rings; direction of a small but turbulent regurgitanc jet against che left atrial wall.4

The clinical manifestations of intravascular hemolysis may include easy fatigability, dyspnea, jaundice, cola-colored urine, tachycardia and arrhythmias. Laboratory confirmation may commonly be found from elevated LOH, plasma hemoglobin, indirect bilirubin, depressed or absent hapcoglobin, hemoglobinuria, hemosiderin, erythrocytosis, spherocyrosis, polychromarophilia and schisrocyces.

Patients developing hemolysis after MVR generally present within the first six months.[1] Garcia and associates have verified several hemodynamic paccerns using transesophageal echocardiography and fluid hydrodynamics simulation.6 High-shear stress patterns such as fragmentation, collision and rapid acceleration were more likely to cause hemolysis compared to low-shear stress patterns. In vicro studies have demonstrated that shear forces greater than 3,000 dyne/cm^2 are associated with significant red cell destruction.7 However, hemolysis can occur even without severe micral regurgitacion.4 Low levels of hemolysis chat do not cause severe anemia might not be detected during routine follow-up, and hemolysis may lessen in rime if the direction of the regurgirant jet changes and/or the prosthetic ring or felt pledget becomes endothelialized. Thus, when incravascular hemolysis is first discovered in a patient with minimal evidence of hemolysis, an initial trial of medical therapy may be prudent wirh afterload reducers, iron, folate and vitamin B-12 supplementation. When medical treacmem is unsuccessful, however, reoperation should not be delayed and may involve repeat valve repair or replacement with a prosthetic valve.[8,9]

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