

Cardiovascular Surgery - Case Report

Incidental Intra-Operative Finding of Large Chronic Pulmonary Embolism in a Patient with Mitral Stenosis Undergoing Mitral Valve Replacement

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Background --- Intraoperative findings may sometimes complicate a previously planned straight forward operation. This paper reports a case of a patient for mitral valve replacement for mitral stenosis with an intraoperative finding of a large chronic pulmonary embolism and the intra-operative decision made to manage this situation.

Case --- We report a case of a 30 year old male patient who has rheumatic mitral stenosis for mitral valve replacement. Intraoperatively, the surgeons found a large chronic embolism occupying approximately 80% of the lumen of the main pulmonary artery, and going towards the right and left pulmonary arteries. He underwent pulmonary endarterectomy, Mitral Valve Replacement and Tricuspid Valve Annuloplasty using a modified De Vega technique.

Conclusions --- The case presented is one of those rare cases in which a patient with valvular disease specifically mitral stenosis had an intra-operative finding of chronic pulmonary emboli. The decision of performing a more tedious and critical surgery such as valve replacement with pulmonary endarterectomy was a critical intra-operative decision that could have meant either the patient would benefit dramatically and survive longer or be no better off than his pre-operative state. *Phil Heart Center J 2013;17(1):50-5.*

Key Words: Mitral stenosis ■ Pulmonary embolism ■ Pulmonary endarterectomy

In developed countries the incidence of Rheumatic Heart Disease (RHD) has substantially decreased due to primary prevention of rheumatic fever.¹ However, a developing country such as the Philippines has RHD as one of its major problems when it comes to heart diseases since majority of the population remain well below the poverty line. In effect, medications needed to treat sore throat are not easily accessible to the average Filipino patient even in government-run hospitals where medications are subsidized. At the Philippine Heart Center, the growing number of patients seen at the outpatient department who are in line for valvular surgery is still that of the hundreds because these patients await funding from charitable organizations or institutions for their operation. The major drawback is that these patients would be admitted already presenting with complications from their disease. As a consequence, a previously planned straight forward operation becomes a more complex one because of

encountered intra-operative findings produced by the progression and gravity of the valvular heart disease. Available literature has taught us that such complications of valvular diseases may happen but how to deal with such a case has not been truly elaborated. The purpose of this paper is to report a case of mitral stenosis with a large chronic pulmonary embolus and the intra-operative decision made to manage this type of situation.

The Case

The patient was admitted for shortness of breath. His condition started two years prior to admission when he was admitted at the PHC due to shortness of breath and pulmonary congestion. During the admission, two-dimensional echocardiography (2DED) revealed severe mitral valve stenosis (MS) with a mitral valve area(MVA) of 0.96 cm² by pressure half time and 0.91 cm² by planimetry and a mitral valve

gradient (MVG) of 15 mmHg. Both the anterior and posterior mitral valve leaflets were thickened and calcified with restricted movement. He had moderate aortic valve regurgitation (AR) 2+, moderate tricuspid valve regurgitation (TR) and pulmonary valve regurgitation (PR) and moderate pulmonary hypertension with systolic pulmonary artery pressure (PAP) of 65 mmHg. The ejection fraction (EF) was within normal limits. No thrombus was noted. He was decongested and advised surgery; but due to financial constraints, he did not comply and was advised to follow up at the out-patient department for proper monitoring in preparation for definitive surgery. In the interim, he had frequent ER visits due to congestion. He was medically managed waiting for his schedule and was maintained on Digoxin, Metoprolol, Furosemide and Penicillin with good compliance. One week PTA, he experienced productive cough with whitish phlegm and bipedal edema. Two days PTA, the patient's symptoms progressed this time accompanied by undocumented fever. A day PTA, he complained of dizziness, generalized weakness and difficulty of breathing, prompting consultation at PHC Emergency Room (ER) where he was admitted. Past health history and family health history were unremarkable. Patient had no vices and was unemployed due to his illness.

During the admission at the ER, he was examined awake, in respiratory distress, and jaundiced. The following are the physical examination findings: pink palpebral conjunctivae; icteric sclerae; neck veins were distended; with hepatojugular reflux; symmetrical chest expansion; no chest retraction; and with fine rales, mid to base of both lung fields. Heart findings showed the following: dynamic precordium, apex beat on the 6th intercostal space on the left anterior axillary line; positive right ventricular heave; irregularly irregular rhythm; positive grade 3/6 diastolic rumbling murmur at the apex; positive grade 3/6 holosystolic murmur on left parasternal border; positive Carvallo's sign and positive opening snap. Abdomen was noted to be flabby with normoactive bowel sounds, soft, nontender, with fluid wave and hepatomegaly. He also had Grade II bipedal edema, no cyanosis, and pulses were fair and equal on both extremities. Admitting impression was Rheumatic Heart Disease, severe mitral stenosis, tricuspid regurgitation in chronic atrial

fibrillation (AF) with chronic passive congestion of the liver. He was admitted to the Medical Intensive Care Unit (MICU) under the service of Critical Care.

At the MICU, he was placed on heart failure regimen with invasive monitoring. Chest x-ray (CXR) revealed cardiomegaly with left atrial and right ventricular prominence, congested lung fields, and minimal bilateral pleural effusion. A repeat 2DED done revealed the MVA to be 0.82 cm² by pressure half time and 0.78 cm² by planimetry and MVG of 4 mmHg. PAP was 95 mmHg by TR jet. EF was 64% by Simpson's. The patient was assessed as having a progression with regards to his MS but still with the same findings of TR and AR. Pulmonary hypertension likewise worsened. Plan at this time was for bridging Percutaneous Transvenous Mitral Commissurotomy (PTMC) and was referred to Invasive Cardiology Service. However, outright Mitral Valve Replacement was suggested by Invasive Cardiology service due to a Wilkins score of ten (10). He was referred then to Thoracic and Cardiovascular Service and assessed for proper timing of surgery. He was evaluated by Pulmonology service and stratified as high risk of developing pulmonary complication post-op. Decongestion and electrolyte replacement were done at the MICU with intense monitoring in preparation for surgery.

He was eventually cleared for surgery with a preoperative plan of mitral valve replacement (MVR) with tricuspid valve annuloplasty (TVA). Standard hospital protocol for heart-lung machine priming and cardioplegia solutions was done. The approach was midline sternotomy. The arterial cannulation was on the aortic root and venous cannulation was on the proximal superior vena cava using a Pacifico cannula and an armored straight cannula on the proximal inferior vena cava. An antegrade and retrograde cannula for delivery of cardioplegia was put in the aortic root and within the coronary sinus respectively. After aortic cross clamping, cardioplegia was given to arrest the heart. After cardiac silence, a biatrial approach was done to inspect both the tricuspid and mitral valves. Intra-operatively, the patient's mitral valve leaflets were severely calcified and leaflet edges were rolled inwards, both commissures were fused, and chordae were shortened. (Figure 1).

The tricuspid valve annulus was enlarged causing none-coaptation of valve leaflets. The right ventricular outlet was inspected and the main pulmonary artery initially palpated and suspected to have a thromboemboli; hence, it was incised vertically. A large chronic emboli occupying approximately 80% of the lumen going towards the right and left pulmonary arteries was noted. (Figure 2). Procedure done was a pulmonary endarterectomy, Mitral Valve Replacement using a mechanical valve (St. Jude Medical) size 27 millimeters and a TVA using a modified De Vega technique. Total bypass time was two hours and forty-one minutes and total cross clamp time was one hour and forty-two minutes. The whole time standard technique of giving antegrade and retro grade cardioplegia was done every 15-20 minutes. There was no problem encountered coming off bypass. After hemostasis and insertion of chest tubes, the chest was closed with standard technique of using sternal wires and layer by layer closure of skin.

At the recovery room, the patient was hemodynamically stable and the supports were slowly tapered. On the 2nd post-op day, he was extubated and transferred to the Surgical Inten-

sive Care Unit (SICU). On the 3rd post-op day, he had minimal drainage from chest tubes, hence, it was removed. Cardiac supports were eventually discontinued. He was transferred to the Adult Service Ward (ASW) on the 5th post-op day after 2DED revealed a normally functioning mitral mechanical valve with effective orifice area of 3.19 cm² with MVG of 5.4 mmHg, PAP decreased to 86 mmHg. On the 7th post op day, there was minimal drainage from the closed suction drain; hence, it was removed together with the myocardial wire. On the 10th hospital day, the sternal wound was noted to be erythematous; thus, antibiotics were resumed. On the 14th hospital day, wound discharge was noted on the sternal incision and wound culture taken. The assessment was surgical site infection. He was then scheduled for emergency wound exploration on the 16th post-op day after loading with antibiotics. Intra-operatively, the wound was classified as a deep infection involving muscle and fascia. Wound debridement was done and drains were inserted. Wound care was done daily. IV antibiotic course was completed and the patient was discharged, recovered after nine days post debridement.

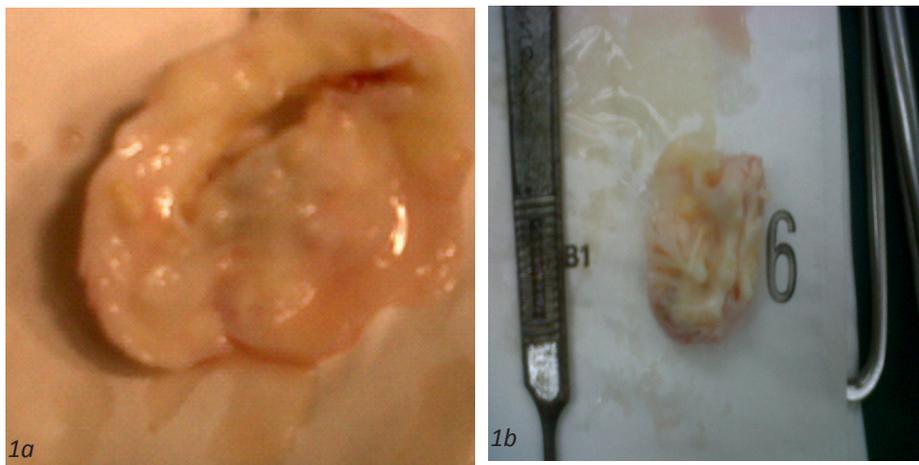


Figure 1. Intraoperative finding of a 30/M with mitral stenosis. 1a-calcified mitral valve. 1b-shortened chordae tendinae of the mitral valve



Pulmonary Thrombus

Figure 2. Intraoperative finding of a 30/M with mitral stenosis. An incidental finding of a large chronic emboli occupying approximately 80% of the lumen going towards the right and left pulmonary arteries

DISCUSSION

Rheumatic cardiac disease is “an immunologic phenomenon that may affect any of the heart valves and the myocardium and is by far the most common cause of mitral stenosis.”² The rheumatic inflammatory process is associated with some degree of pancarditis but permanent injury results predominantly from the endocarditis, along with progressive fibrosis of the valves. Rheumatic valvulitis produces three distinct degrees of pathologic change: fusion of the commissures alone, commissural fusion plus subvalvular shortening of the chordae tendinae, and extensive fixation of the valve and subvalvular apparatus with calcification and scarring of both leaflets and chordae.³ Rheumatic mitral stenosis almost always is acquired before age twenty, and becomes clinically evident one to three decades later. In patients with mild to moderate mitral stenosis, pulmonary vascular resistance and pulmonary arterial pressure may remain within normal limits at rest rising only with exertion or increased heart rate. In contrast to severe chronic mitral stenosis, pulmonary vascular

vascular resistance is increased and pulmonary arterial pressure is elevated at rest and can approach systemic pressure with exercise.⁴

Pulmonary hypertension develops “as a result of passive transmission of high LA pressure, pulmonary venous hypertension, pulmonary arteriolar constriction, and eventually, pulmonary vascular obliterative changes.”⁴ The pulmonary hypertension worsens progressively over time leading to right-sided heart failure, tricuspid insufficiency, and occasionally, pulmonary valve insufficiency like that of the patient.⁴ Severe mitral stenosis ultimately causes irreversible pulmonary vascular changes; cardiac output is low at rest and remains subnormal during exercise and ultimately makes the patient a poor candidate for surgical intervention because prognosis after surgery is similar as to medical management alone. In other words, the patient will not benefit from the surgery and if surgery is done, it would just add to the morbidity of the otherwise morbid state of the patient.

A 2-dimensional echocardiogram is the ancillary procedure of choice when it comes to the diagnosis of MS. The predominant feature of mitral stenosis on 2D echocardiography is “thickening and restriction of motion of both mitral valve leaflets, with the predominant pathologic process being at the tips of the leaflets and proximal chordae.”⁵ Wilkins developed an echocardiographic scoring system, based on the leaflet rigidity, thickening, valvular calcification and subvalvular involvement, to assess suitability for PTMC. Subjects with score of 8 or less have excellent immediate and long-term results with PTMC, while those who score greater than 8 have less impressive results. Slight commissural calcium is not a contraindication for percutaneous mitral commissurotomy, but when the calcium score is more than 2+, the incidence of restenosis is higher and thus surgical repair of the mitral valve is preferable.⁵

In mitral stenosis, progression of the disease is usually slow and complications arise as a result of the abnormal hemodynamic state. Platelet activation has been reported and may contribute to thrombus formation.⁶ Complications such as pulmonary hypertension, atrial fibrillation and thromboembolism may occur. Rarely a pulmonary thromboemboli is encountered and such cases have been reported at University of California at San Diego. Timing of surgical therapy is of importance since numerous studies on early mitral valve surgery have shown excellent long term benefit among patients. Naturally, the longer the time of intervention of an otherwise ailing heart will result in a much longer insult to the valve and therefore affect the overall function of the heart.

In the case presented, the patient may have developed a thromboemboli as a complication of his chronic atrial fibrillation. According to the pathologic study by Berthrong, et.al, if an acute pulmonary emboli does not lyse within 1 to 4 weeks, the embolis material becomes attached to the pulmonary arterial wall at the main, lobar, segmental, or subsequential levels. With time, the initial embolic material progressively becomes converted into connective and elastic tissue.⁷ By a mechanism that is poorly understood, chronic thromboembolic obstruction may also lead to a small vessel arteriolar vasculopathy characterized by excessive proliferation of

smooth muscle cells around small arterioles in the pulmonary circulation. This small vessel vasculopathy may cause pulmonary hypertension due to long exposure at high flow resulting in decreased capacitance of the remaining open bed to absorb the cardiac output.⁸

In a multinational prospective long-term follow-up study done by Pengo and colleagues their findings were that patients who survive an acute pulmonary embolic event will go on to develop chronic pulmonary hypertension within the next 2 years in approximately 3.8%.⁹ This subset of patients will have a poor prognosis especially in the absence of intracardiac shunts. In addition, they fall into a higher-risk category than those with Eisenmenger’s syndrome and encounter a higher mortality rate.⁸ The estimated mortality rate reported by Reidel and colleagues states that once the mean pulmonary pressure in patients with thromboembolic disease exceeds 50mmHg, the 5-year mortality approaches 90%.¹⁰ With the advent of better drugs and advancement of critical care, such an estimation may be lower although statistically still significant.

The primary treatment of patients with chronic thromboembolic pulmonary hypertension is surgical endarterectomy of the pulmonary arterial tree. Medical management is ineffective and used primarily for preparation prior to definitive surgery, while lung or heart-lung transplantation is associated with worse results than those seen with pulmonary endarterectomy and is not feasible here in the Philippines. This procedure is considered in patients who are symptomatic and have evidence of hemodynamic or ventilatory impairment at rest or with exercise. There is no upper limit of PVR or degree of right ventricular dysfunction or tricuspid regurgitation that excludes a patient from operation, even a patient with suprasystemic PAP can safely undergo the surgery.⁸

There are four major types of pulmonary occlusive disease which are based on anatomy and location of thrombus and vessel wall pathology. This intraoperative classification of disease allows for the prediction of patient outcome after pulmonary endarterectomy.¹¹ Type 1 disease (approximately 30% of cases of thromboembolic pulmonary hypertension)) is fresh

thrombus visible in the main or lobar pulmonary arteries. This is the intraoperative classification of the patient. Type 2 disease (approximately 60% of cases) is intimal thickening and fibrosis with or without organized thrombus proximal to segmental arteries. Type 3 disease (approximately 10% of cases) is fibrosis, intimal webbing, and thickening with or without organized thrombus within distal segmental and subsegmental arteries only. Type 4 disease (approximately 1% or less of cases) is microscopic distal arteriolar vasculopathy without visible thromboembolic disease and is inoperable.⁸

In patients with extreme forms of thromboembolic pulmonary hypertension, pulmonary endarterectomy offers hemodynamic benefit that is immediate and curative. Furthermore, even if PAP is suprasystemic, pulmonary endarterectomy in this patient population is still superior to lung transplantation or heart-lung transplantation. In the study of Thistlewaite, PA and colleagues they concluded that survival rate is 89.2% in the perioperative period and approximately 75% at 5 years as compared to survival rates for patients undergoing lung or heart-lung transplantation for this disease, as reported in the 2004 Annual Report of the US Organ Procurement and Transplantation Network to be 74% at 3 months and 39% at 5 years for single- and double-lung transplantation and 80% at 3 months and 48% at 5 years after heart-lung transplantation for pulmonary hypertension.¹²

The case presented is one of those rare cases in which a patient with valvular disease, specifically mitral stenosis, had an intra-operative finding of chronic pulmonary emboli. The decision of performing a more tedious and critical surgery such as valve replacement with pulmonary endarterectomy was a critical intra-operative decision that could have meant either the patient would benefit dramatically and survive longer or be no better off than his preoperative state. The worst case scenario is that the patient may be extremely morbid post-operatively or even die from the procedure. Though this may be just one case, this case report is intended to make surgeons aware that such intra-operative decision-making may be part of his arsenal for future use when faced with a similar situation in the field of Cardiovascular Surgery.

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