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Successful Transcatheter Aortic Valve Replacement for Severe Aortic Stenosis in a Patient with Idiopathic Pulmonary Hemosiderosis: A Case Report

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Abstract

Idiopathic pulmonary hemosiderosis is a chronic respiratory disorder characterized by repeated episodes of alveolar hemorrhage. Patients with this disorder have a risk for acute exacerbation and life threatening respiratory failure. To date, it has not been well elucidated whether patients with idiopathic pulmonary hemosiderosis can safely undergo cardiac valve surgery. Here we describe a case of successful transcatheter aortic valve replacement for severe symptomatic aortic stenosis in a 75-year-old man with idiopathic pulmonary hemosiderosis. Since a wide range of physiological alterations resulting from cardiopulmonary bypass have the potential to trigger life-threatening alveolar hemorrhage, it is prudent to avoid conventional valve replacement surgery in these patients. To maximize patient safety, transcatheter aortic valve replacement is an acceptable alternative.

Keywords: Diffuse alveolar hemorrhage • Idiopathic pulmonary hemosiderosis • Transcatheter aortic valve replacement

Abbreviations: SAVR: Surgical Aortic Valve Replacement; AS: Aortic Stenosis; CPB: Cardio-Pulmonary Bypass; IPH: Idiopathic Pulmonary Hemosiderosis; DAH: Diffuse Alveolar Hemorrhage; TAVR: Transcatheter Aortic Valve Replacement; AF: Atrial Fibrillation

Introduction

Although surgical aortic valve replacement (SAVR) has been the standard of care for patients with severe Aortic Stenosis (AS), increased peri-operative morbidity and mortality is a major concern in some patients with certain preexisting conditions, since a wide range of physiological alterations related to Cardiopulmonary Bypass (CPB) develop during and after surgery. Idiopathic Pulmonary Hemosiderosis (IPH), one of the well-recognized causes of Diffuse Alveolar Hemorrhage (DAH) [1], is a chronic respiratory disorder characterized by repeated episodes of acute exacerbation manifested with dyspnea and hemoptysis [2,3]. If a patient with IPH undergoes SAVR, there is concern whether life-threatening DAH could occur in the peri-operative period. Recently, there is a growing body of evidence related to the efficacy of Transcatheter Aortic Valve Replacement (TAVR). One of the major advantages of TAVR over SAVR is its lesser invasiveness, mainly due to avoiding the use of CPB. To date, however, neither SAVR nor TAVR in patients with IPH has been reported in the literature. In this case report, we will discuss about advantages of TAVR over SAVR in patients with IPH who undergo aortic valve intervention. Written consent to publish this case report was obtained from the patient.

Case Report

A 75-year-old man with symptomatic severe AS was referred to our institution for further evaluation and treatment. Six years before admission, he was hospitalized because of hemoptysis and dyspnea. Computed tomography scan of the chest showed multiple areas of consolidation surrounded by ground-glass opacities with subpleural sparing (Figure 1). He immediately underwent bronchoscopy, and DAH was confirmed. Other causes of acute alveolar hemorrhage, such as collagen disease or vasculitis syndrome were excluded, and the diagnosis of IPH was made. Prednisolone therapy was started. Over the next 6 years, he experienced several acute exacerbations and repeat hospitalizations, all of which were successfully treated with short-term high dose prednisolone therapy. Five months prior to admission, his weight increased over a two-week period, followed by palpitations, dyspnea on exertion, and peripheral edema, although no hemoptysis was observed. Electrocardiography showed Atrial Fibrillation (AF) with moderate ventricular

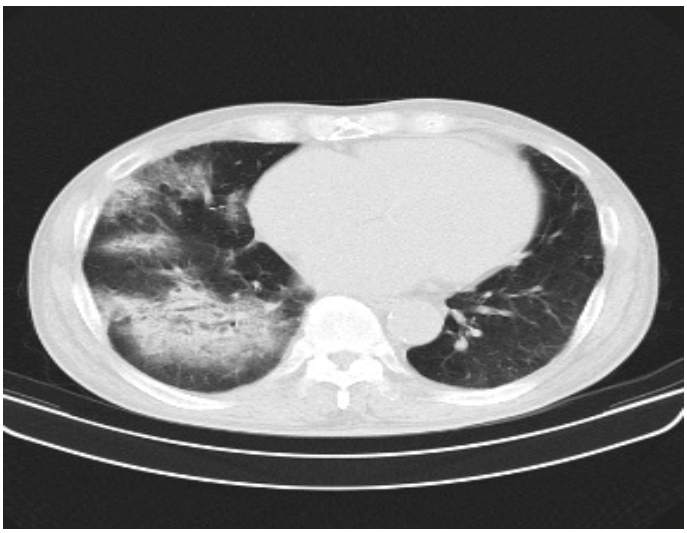

Figure 1. Computed tomography scan of the chest, at the time of initial diagnosis with idiopathic pulmonary hemosiderosis which shows areas of consolidation surrounded by ground-glass opacities with sub-pleural sparing, consistent with diffuse alveolar hemorrhage.

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response. Chest radiography showed bilateral pulmonary edema. Cardiac echocardiography revealed severe AS and moderate mitral regurgitation.

The diagnosis of congestive heart failure due to severe AS was made. He was transferred to our institution for further evaluation and treatment. Physical examination was unremarkable, except for a systolic ejection murmur over the aorta, and peripheral pitting edema of the lower extremities. Oxygen saturation was 96% while breathing ambient air and he had no dyspnea at rest. An electrocardiogram showed AF with a moderate ventricular response. Cardiac echocardiography demonstrated a left ventricular ejection fraction of 47%, a severely calcified aortic valve with a reduced orifice. Estimated aortic valve area was 0.7 cm^2, and a mean aortic trans-valvular pressure gradient of 84.6 mmHg. There was mild aortic regurgitation, moderate mitral regurgitation, and mild tricuspid regurgitation with a dilated right atrium and ventricle. Computed tomography scan of the lung showed subpleural irregular reticular opacities but no evidence of active diffuse alveolar hemorrhage (Figure 2).

The multidisciplinary heart valve team reviewed and evaluated the patient. Since the concern about the risk of perioperative exacerbation of IPH was raised, TAVR was planned instead of SAVR. Aortic root and iliac arterial anatomy were suitable for TAVR. Left sided cardiac catheterization study revealed no significant coronary artery disease. Elective trans-femoral approach TAVR, with a 26 mm Sapien XT prosthesis (Edwards Lifesciences, Irvine, CA, USA), was performed under general anesthesia. The procedure was accomplished without any complications, except for a transient episode of hypotension followed by generalized flushing, possibly due to an anaphylactoid reaction caused by rocuronium. After surgery, in addition to antiplatelet therapy with aspirin and clopidogrel, rivaroxaban was commenced, because a thrombus within the left atrial appendage was incidentally found by transesophageal echocardiography. No postoperative complications occurred and he was discharged home.

Discussion

DAH is defined as bleeding into alveolar spaces from alveolar capillaries [1]. A wide range of diseases cause DAH, and are classified into three major categories including 1) pulmonary capillaritis 2) diffuse alveolar damage, and 3) bland pulmonary hemorrhage [1]. Among these, bland pulmonary hemorrhage is defined as alveolar hemorrhage without inflammation or destroyed alveolar structures [1]. This category includes bleeding disorders or anticoagulation, elevated pulmonary venous capillary pressure, and IPH [1]. IPH is a rare, chronic respiratory disease, characterized by recurrent episodes of DAH [2,3]. Although most patients with IPH cases have childhood onset, about 20% of patients with IPH are adult-onset [3]. The diagnosis of IPH should be made only after other conditions associated with DAH have been ruled out [2,3].

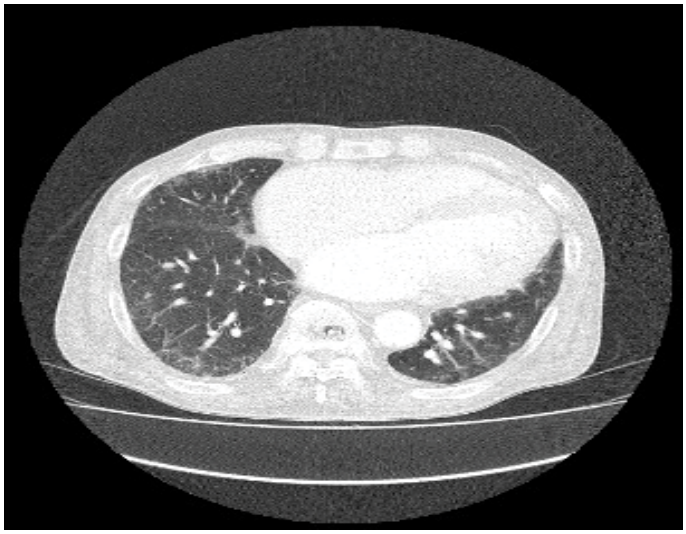

Figure 2. Computed tomography scan of the chest, taken 1 month prior to transcatheter aortic valve replacement which shows sub-pleural irregular reticular opacities in the lower lobe. There is no evidence of active alveolar hemorrhage.

The etiology of IPH is still unknown, however, various immune processes may predispose to this condition [3]. Since acute exacerbations and respiratory failure are the leading causes of death among patients with IPH [2,3], avoidance of acute exacerbations is crucial to improve long-term outcome.

Although the precise mechanism of acute exacerbation of IPH is not well understood, elevated pulmonary venous capillary pressure might have the potential to worsen preexisting alveolar hemorrhage. Several case reports describing acute exacerbation of IPH associated with pregnancy support this hypothesis. According to these studies, worsening of IPH occurred either in the third trimester of pregnancy [4-7] or immediate postpartum period [8], when the maternal blood volume reaches its highest level. Although the relationship between pregnancy and exacerbation of IPH is not well described, the authors suggested that elevated pulmonary capillary venous pressure due to increased blood volume might play a role in developing an acute exacerbation of IPH. On the basis of these findings, a theoretical hypothesis could be made, that elevated pulmonary venous capillary pressure secondary to volume overload after cardiac valve surgery, might exacerbate IPH. Cardiac surgery itself and CPB often result in significant volume shifts to the interstitial space during surgery. As a result, transient hypervolemia and pulmonary venous congestion may develop within a few days after surgery. It is in this environment that exacerbation of IPH might occur in the same way as described in pregnancy.

Impaired hemostasis is also a well-recognized cause of DAH. Several reports of DAH caused by anticoagulation with warfarin [9,10], low molecular weight heparin [11], dual antiplatelet therapy [10,12], and treatment with platelet glycoprotein IIb/IIIa receptor inhibitor [13] have been reported. In some of these cases, patients developed life threatening respiratory failure requiring mechanical ventilatory support and extracorporeal membrane oxygenation [10,12]. In cardiac valve surgery using CPB, anticoagulation with high dose heparin is always necessary. In addition, impaired hemostasis due to CPB, such as platelet dysfunction and depletion of fibrinogen, continues thereafter. Therefore, concern has been raised about the risk of developing life-threatening DAH during and after CPB. Given the fact that DAH associated with anti-coagulation or anti-platelet therapy could occur even in previously healthy individuals, patients with IPH are more likely to develop severe DAH during and after cardiac surgery, with volume overload and impaired hemostasis developing coincidentally.

TAVR is a novel, alternative procedure to treat patients with severe AS that enables valve replacement without CPB. In the early stages, TAVR is indicated for only those who are at high surgical risk, defined as an STS-PROM (Society of Thoracic Surgeons-Predicted Risk of Mortality score) of 8% to 15% [14]. However, recent studies demonstrated no inferiority of TAVR to SAVR, even in patients at intermediate-risk (defined as an STS PROM of 4% to 8%) [15]. Although our patient's STS-PROM was 4.5%, controversy still exists whether TAVR was indicated, because either extreme frailty or prohibitive risk for conventional cardiac surgery other than IPH was not present in this patient. In addition, long term durability of the prosthesis and survival benefit of TAVR has not been fully established [14]. However, we decided to proceed with TAVR, as there are several advantages over SAVR in this patient. First, the risk of fluid overload is less during and after TAVR. Second, a shorter duration of anticoagulation, with less unfractionated heparin, is sufficient to undergo TAVR. While activated coagulation time should be kept above 400 seconds during CPB, only 250 seconds or above is sufficient during TAVR. Finally, avoidance of CPB will be beneficial in preventing impaired hemostasis and significant volume shifts. For these reasons, we decided to perform TAVR.

In summary, cardiac valve surgery with CPB in patients with IPH carries a risk of developing DAH and life-threatening respiratory failure. TAVR is a reasonable procedure for patients with severe aortic stenosis and IPH. The development of apical thrombi frequently occurs after large AMI with LV aneurysm. When transthoracic ultrasonography does not allow adequate characterization of the thrombus and cardiac magnetic resonance is not feasible, CT scan can provide additional definition of cardiac walls and the extent of the thrombus.

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