Life-threatening hemoptysis following mitral valvuloplasty for rheumatic mitral stenosis

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ABSTRACT

Background: Massive hemoptysis is a rare complication of rheumatic mitral valve stenosis. Its recurrence following successful initial treatment of the stenosis has not, to our knowledge, been described.

Case report: We describe a 58-year-old African American woman with a history of balloon valvuloplasty for the treatment of severe rheumatic mitral valve stenosis who presented to our institution with massive life-threatening hemoptysis due to recurrent mitral valve stenosis. Repeat balloon valvuloplasty was complicated postoperatively by severe mitral regurgitation and the patient expired from refractory cardiopulmonary collapse not amenable to further intervention.

Conclusion: Life-threatening hemoptysis is a medical emergency requiring rapid source identification and treatment of the underlying etiology. A high degree of suspicion should be maintained for recurrence of mitral valve stenosis in patients presenting with life-threatening hemoptysis and risk factors of rheumatic heart disease, regardless of previous surgical management or unilateral chest x-ray signs.

1. Background

"Rheum" is a Greek term that refers to the flow of watery fluid, and was used in the 17th century to signify the flow of fluid into joints. Differentiating rheumatic fever (previously known as acute rheumatism) from rheumatoid arthritis (previously known as chronic rheumatism) was challenging because the conditions were thought to be a spectrum of one disorder. David Pitcairn linked rheumatism and cardiac disease in 1788 when he described “rheumatism of the heart” [1]. While the term “rheumatic fever” was coined in 1806, physicians continued to describe it as “acute rheumatism” for another 100 years [2].

In the 20th century, rheumatic fever was established as a separate entity linked to the bacterium group A streptococcus. The introduction of antibiotics for streptococcal pharyngeal infections greatly reduced the incidence of acute rheumatic fever (ARF), which now affects fewer than 10 per 100,000 people in the United States [3]. Despite our current understanding of the etiology, pathophysiology, and management of ARF, managing its complications can often challenge even the most seasoned clinician. Rheumatic heart disease (RHD), a late sequel of ARF, remains one of the most common causes of acquired heart valve disease in resource-limited countries [3]. Rheumatic heart disease presents differently with age, shifting from mitral regurgitation in those younger than 30 years, to mitral stenosis in middle aged patients, to mixed mitral valve diseases in the older population [4]. In this case, we report on a rare presentation of complicated rheumatic mitral valve disease, years after successful initial management.

2. Case report

A 58-year-old African American woman without any prior foreign travel was admitted to our hospital with dyspnea and scant hemoptysis that later progressed to massive life-threatening hemoptysis. Her past medical history was significant for chronic obstructive lung disease, hepatitis C, complicated rheumatic heart disease, congestive heart failure, and atrial fibrillation. Her medication list included metoprolol, amiodarone, and warfarin. She was a regular smoker with a 42-pack-
year history but had no history of alcohol abuse or dependence. Her only prior surgery was a mechanical aortic valve replacement (AVR) at the age of 36, secondary to longstanding RHD. At the age of 53, seventeen years after her AVR, she experienced episodic hemoptysis (~0.5 L total), which was attributed to newly developed mitral stenosis. Transthoracic echocardiogram (TTE) estimated a peak mitral valve gradient of 35 mmHg, with a mean gradient of 17 mmHg, mitral valve area of 0.8 cm², and moderate pulmonary artery hypertension. Despite brief success with medical therapy as an outpatient, she was readmitted with massive hemoptysis requiring intubation. Laboratory studies showed an INR of 4.8, PTT of 57, WBC of 21.7, hemoglobin of 10.8, hematocrit of 33.7, and platelet count of 323. She underwent a balloon valvuloplasty that resulted in immediate resolution of the bleeding. Estimated mean gradient across the valve fell to 9.5 mmHg and the mitral valve area increased to 2.3 cm². The patient and the surgical team involved at that time elected against proceeding with a valve replacement.

She remained well for five years until her presentation at our hospital with dyspnea and streaky hemoptysis. Shortly after admission, she was intubated for life-threatening hemoptysis, at which time her chest x-ray demonstrated unilateral signs of heart failure, including increased prominence and cephalization of the pulmonary arteries, silhouetting of the cardiac borders, and a right-sided effusion and infiltrates obscuring the right hemidiaphragm (Fig. 1). Post-intubation bronchoscopy revealed large airway submucosal varices bilaterally and a clot in the left mainstem bronchus, which was extracted. A TTE showed anterior and posterior mitral valve leaflet thickening and fusion of the commissures causing severely restricted movement; estimated mitral valve peak gradient was 47 mmHg, mean gradient was 21 mmHg (Fig. 2), and mitral valve area had fallen to 0.5 cm². Severe pulmonary hypertension was also evident. The patient’s mitral valve stenosis and elevated right heart pressures were subsequently confirmed with right heart catheterization.

Transesophageal echocardiography (TEE) findings in the setting of RHD-induced mitral valve stenosis tend to have a characteristic appearance and help to guide management decisions. One stereotypical TEE finding is the “hockey stick” configuration of the anterior leaflet, which exhibits an outward “doming” with a partially or completely immobile posterior leaflet. The likelihood of a successful percutaneous valvuloplasty was estimated using the Wilkens scoring system, which assesses echocardiographic features of the mitral valve such as mobility, thickness, calcification, and subvalvular thickening [5]. Our patient’s valve score during her presentation was 6 out of possible 16, with higher scores portending worse clinical outcomes with valvuloplasty. Her high risk status for surgery as well as her favorable valve score suggested a more positive outcome with balloon valvuloplasty [5].

Given these findings, she underwent an urgent repeat balloon mitral valvuloplasty for recurrent rheumatic mitral stenosis. Her post-procedure course was complicated by the development of severe mitral regurgitation, causing both refractory hypoxic respiratory failure and severe pulmonary hypertension that resulted in cardiovascular collapse and multiorgan failure. Based on her clinical instability, prognosis, and family preferences, she was not considered a candidate for valve replacement and subsequently expired.

3. Discussion

Approximately half of patients with AFE develop cardiac manifestations [6], and of those affected, only about half ever manifest cardiac valvular pathology appreciable by auscultation later in life [7]. In these cases, mitral valvular disease represents the most commonly affected heart valve [6]. Mild hemoptysis is frequently seen in rheumatic mitral valve stenosis. However, massive hemoptysis is an uncommon but life-threatening complication that occurs in fewer than 3 in 1000 patients [8]. In such rare cases, the source of the bleed must be correctly identified, as massive gastrointestinal bleeding or bleeding from the oropharynx can be confused with hemoptysis. Bronchoscopy, which confirmed our patient’s site of bleeding, can assist to further differentiate between an airway and a focal or diffuse parenchymal bleed. In cases of massive hemoptysis secondary to RHD, urgent intervention (e.g., mitral valve replacement, commissurotomy, or valvuloplasty) can halt bleeding [9]. In our report, we identified the occurrence of massive hemoptysis as a presenting complaint secondary to a recurring mitral stenosis.

The unilateral signs of heart failure on chest x-ray represent another informative presenting feature in this case. Unilateral chest x-ray signs are uncommon in heart failure and can mislead physicians to consider solely unilateral pathological processes. Mitral disease, however, should remain in the differential diagnosis of such a radiographic appearance. In one case series, two out of five cases of unilateral chest x-ray signs of heart failure were due to recurrent rheumatic mitral valve stenosis following balloon valvuloplasty, similar to our case, but presenting more overtly with symptoms of heart failure [10]. The propensity of recurrent mitral valve stenosis to cause unilateral pulmonary edema has not been reported on in the literature, but according to our knowledge, this case constitutes the third reported incident.

We based our patient’s treatment on case reports and small studies showing immediate resolution of hemoptysis with correction of the mitral valve stenosis [9–12]. Given the need for rapid intervention and available resources at the time, valvuloplasty was the therapeutic option that provided the most immediate pragmatic treatment with the greatest chance of success. Despite good response to valvuloplasty after her first presentation for mitral stenosis five years prior, her recurrence proved fatal due to the development of hemodynamic decomposition progressing to multiorgan failure.

To our knowledge, no other report documents rheumatic mitral valve stenosis-induced hemoptysis following a prior mitral valvuloplasty. Other cases describing patients with hemoptysis after either mitral valve repair or radioablation of atrial fibrillation exist, although these cases appear to be related to pulmonary vein stenosis and not actual recurrence of valve stenosis [13]. Reports also exist of massive hemoptysis as the initial presentation of mitral stenosis [14], which our patient experienced as well. We present this case to highlight that mitral stenosis can constitute the third reported incident.

![Fig. 1. Portable AP chest x-ray obtained on re-presentation with massive hemoptysis (five years after valvuloplasty) demonstrating a moderate right pleural effusion (black arrow) and right mid- and lower-lung opacities obscuring the right hemidiaphragm and right heart border. Vascular congestion can be seen with cephalization of the vascular pattern (red arrows). The left hemidiaphragm is clear while the left heart boarder is silhouetted with a suggestion of air bronchograms in the lingula (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)](image-url)
valve stenosis has the potential to recur even after an initially successful intervention.

4. Conclusions

Clinicians should consider mitral valvulopathies in the differential for patients presenting with hemoptysis, especially if risk factors for valvular pathology exist. Additionally, as illustrated in this case, neither prior valvuloplasty nor unilateral chest x-ray signs should exclude the diagnostic possibility of mitral valve stenosis causing life-threatening hemoptysis.

Conflicts of interest

The authors declare that they have no competing interests, real or perceived.

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