Case report

Mitral stenosis due to rheumatic heart disease - A rare cause of massive hemoptysis

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ABSTRACT

Severe mitral valve stenosis caused by rheumatic heart disease presenting initially as massive hemoptysis has become a rare occurrence in contemporary western medicine. Massive hemoptysis can be due to multiple disease processes including airway diseases such as bronchiectasis, pulmonary parenchymal disease of infectious or autoimmune etiology, pulmonary AVM’s, hematologic disorders, and numerous drugs and iatrogenic injuries. It is less associated with congestion from rheumatic heart disease due to the earlier detection and subsequent management of cardiac valve disease preventing the sequel of more severe disease.

We describe a case of a 59 year-old woman with hemoptysis, who was found to have severe mitral stenosis consistent with rheumatic heart disease. We demonstrate the appearance of pulmonary venous congestion can be seen on bronchoscopic examination in severe mitral stenosis and discuss the significance of the Wilkins score to help guide management.

1. Introduction

While hemoptysis is a well-known and documented sequela of mitral stenosis (MS), its occurrence in contemporary western medicine has become rare [1,2]. Rheumatic heart disease (RHD) is the result of an exaggerated immune response to specific bacterial epitopes in a susceptible host [3]. Chronic RHD affecting the mitral valve apparatus progresses over years to decades and causes a number of pathologic changes, affecting the mitral valve apparatus, which are diagnostic for rheumatic valve disease [4,5]: fusion of the leaflet commissures; thickening, fibrosis, and calcification of the leaflet cusps [6]; and thickening, fusion, and shortening of the chordae tendineae.

According to the WHO, the estimated global prevalence of rheumatic heart disease (RHD) is 15.6 million cases. Of those identified cases, 79% originated in less developed countries, with an estimated prevalence in those countries of 2.5–3.2 cases per 1000 all-age population vs. 0.3 cases per 1000 population in developed countries [7]. The incidence rate of rheumatic fever is < or = 10/100,000 per year in America and Western Europe, while a higher incidence (> 10/100,000) was documented in Eastern Europe, Middle East (highest), Asia and Australasia [8]. The prevalence of Rheumatic Fever, RHD and their mortality rates varied widely between countries and between population groups within the same country; globally, about 2% of deaths from cardiovascular diseases are related to rheumatic heart disease [9].

Despite an apparent fall in incidence over time, Acute Rheumatic Fever (ARF) incidence rates remain relatively high in non-Western countries [8]. In developed countries, the progression of disease is more indolent and manifests at older ages (above 50 years); clinically detected RHD is most commonly diagnosed in individuals aged 20 to 50 years with nearly two-thirds of cases occurring in females [10]. Therefore, the incidence of hemoptysis as the presenting symptom of RHD without associated shortness of breath, chest pain or signs of hypervolemia is limited to case reports [1]. Hemoptysis as a separate entity occurs in around 10% of patients with chronic lung disease [11] and is found in ca. 0.1% of all outpatients [12] and almost 0.2% of all inpatients [13] each year.

The decline in hemoptysis as a presenting symptom of RHD is likely due to the dramatic decline in its prevalence in developed nations and the earlier detection, and subsequent management, of valvular disease preventing the sequel of more severe disease. The work up of hemoptysis is described along with the evaluation of MS.

2. Case report

A 59-year-old African American female with a history of pulmonary tuberculosis treated twenty-one years ago, active tobacco use, and untreated hepatitis C presented with three days of coughing up frank red blood. This was preceded by a month of viral-like upper respiratory
symptoms, chills, unintentional weight loss and night sweats. She denied productive cough, lower extremity swelling, shortness of breath or chest pain. She was an active smoker of 0.25 packs per day for forty-five years (11.25 pack year smoking history) with additional alcohol use estimating four beers per day since she was a teenager. She denied any illicit drug use.

Upon presentation she was in no acute distress with normal mental status. Vitals on admission revealed a temperature of 98.2°F, heart rate of 89 bpm, blood pressure of 144/80 mmHg, respiratory rate of 16 while saturating 96% breathing ambient air. There was a prominent S1, split S2 with prominent P2, and a decrescendo-crescendo diastolic murmur loudest at the apex. Pulmonary exam revealed bibasilar crackles. Her abdomen was soft, non-tender and without organomegaly. There was no cyanosis or clubbing of the extremities.

Complete blood count revealed: WBC of 6.2, Hb of 12.9, Hct of 40.1, PLT of 318. PT was 11.9 seconds, INR was 1.1. Hepatic panel was normal with an AST of 27 U/L, ALT of 23 U/L, Alk phos of 60 U/L & Total Bilirubin of 0.9 mg/dL.

A chest x-ray (CXR) (Fig. 1) revealed fibronodular changes in both apices without infiltrates or effusions. A chest computer tomography angiogram (CTA) (Fig. 2) revealed extensive ground glass opacities, bilateral calcified pulmonary nodules most pronounced in the right upper lobe, and a dilated left atrium. No pulmonary embolism or arteriovenous malformations (AVM) were seen.

A transthoracic echocardiogram (TTE) revealed a left ventricular ejection fraction of 35–45%, mild left ventricular hypertrophy, and severe mitral valve stenosis with moderate mitral valve regurgitation.

Sputum AFB smear, blood cultures, HIV, ANA, ANCA and anti-GBM antibodies were obtained and all negative. Rheumatoid factor was elevated. Bronchoscopy (Fig. 3) revealed engorged veins in the trachea and bilateral proximal bronchial trees.

Results of the clinical work-up made infection, malignancy, TB, vasculitis or AVM unlikely. The dilated left atrium and severe mitral stenosis seen on TTE was confirmed by transesophageal echocardiogram (TEE), which showed a mean gradient of 12 mmHg and Wilkins score of 11 (see Figs. 4–6). Left heart catheterization was performed and showed no evidence of coronary artery disease.

Given these findings, the patient’s massive hemoptysis was attributed to severe mitral valve stenosis in the setting of rheumatic heart disease. Given her Wilkins score of 11, she was not a candidate for balloon valvuloplasty, and ultimately underwent successful mitral valve replacement with a pericardial bioprosthetic valve and resection of anterior and posterior papillary muscles.

3. Discussion

Hemoptysis carries a broad differential including airway diseases such as bronchiectasis, pulmonary parenchymal diseases of infectious or rheumatic cause, pulmonary AVM’s, hematologic disorders, and numerous drugs and iatrogenic injuries [14]. Due to this plurality, a thorough history is critical to guide clinical decision making.

While hemoptysis is a well-known and documented sequela of mitral stenosis (MS), its occurrence in contemporary western medicine has become rare [2]. This is likely due to the dramatic decline in the prevalence of rheumatic heart disease in developed nations and the earlier detection, as well as subsequent management, of valvular disease preventing the sequela of more severe disease. Currently, rheumatic fever (RF) mostly affects children in developing countries, especially in areas of widespread poverty [15].

Common causes of severe mitral stenosis in developed countries include mitral annular calcification, radiation-associated valve disease, Fabry’s disease, Whipple’s disease, mucopolysaccharidosis, methysergide therapy, carcinoid valvular disease, endomyocardial fibrosis, and systemic autoimmune disease (such as systemic lupus erythematosus and rheumatoid arthritis) [16].

Other cardiac conditions may produce hemodynamic abnormalities similar to those of native valvular MS. These include atrial myxomas, large infected vegetations, ball valve thrombi, and degenerated stenotic bioprosthetic mitral valves [17].
Common initial clinical features of MS include dyspnea on exertion (about 70%), decreased exercise tolerance, fatigue, atrial fibrillation, thromboembolism, chest pain, signs of right heart failure, hoarseness (Ortner’s syndrome), and infective endocarditis due to the deformed mitral valve [16]. The symptoms of patients presenting with mitral stenosis are secondary to decreased blood flow over the valve with subsequent congestion and increased pressures in the pulmonary circulation [18]. Hemoptysis, though less common, is due to increased pressures causing rupture of pulmonary vessels [18]. Hemoptysis may present as sudden hemorrhage (pulmonary apoplexy), bloody sputum due to severe coughing associated with paroxysmal nocturnal dyspnea/bronchitis, or pink frothy sputum due to pulmonary edema [18]. Management of massive hemoptysis is focused on stabilization of the airway and the circulatory system. Medications to suppress cough and body positioning to protect the unaffected side are incorporated. Urgent bronchoscopy can be helpful to establish the site of bleeding in cases of a moderate bleed [19]. Definitive treatment depends on the underlying cause. For massive hemoptysis due to severe MS, treatment includes percutaneous mitral balloon valvuloplasty (PMBV) or valve replacement [18].

Certain radiological features are expected in patients with severe MS. These include left atrial dilatation, evidence of pulmonary congestion, and signs of possible hemorrhage [18].

Echocardiography is the test of choice for diagnosis, quantification, and often determination of the etiology of mitral stenosis. The Wilkins (Boston/Abascal/MGH) score allows for grading of mitral valve characteristics from the 2DE examination and is thought to aid in predicting mitral regurgitation after PMBV [20,21]. This scoring system evaluates leaflet thickening, mobility, calcification, and subvalvular involvement on a scale of 0–4 in order to help determine the need for valve replacement. Morphology of the mitral valve is considered the main predictor of a successful balloon mitral valvuloplasty (BMV) [22]. The MV morphology is considered favorable if the mitral echocardiographic score is ≤ 8 [23]. A Wilkins score of > 10 independently predicts severe mitral regurgitation after percutaneous mitral balloon valvuloplasty (PMBV) [21]. However, there are limitations to the 2DE exam using the Wilkins score. It may be semi-quantitative, subject to observer variability, less reliable in classifying patients with scores within the
mid-range, determining fibrosis vs calcification, as well as an uneven distribution of pathology, and it underestimates subvalvular disease [24]. Furthermore, recent literature demonstrates 2D ECHO may overestimate the mitral valve area (MVA) [25].

Three dimensional echocardiography (3DE) allows for more accurate determination of rheumatic mitral stenosis severity. 3DE improves identification of the narrowest part of the MV orifice due to better alignment of the image plane at the mitral tips [26].

Therefore, candidacy for PMBV is determined by multiple factors including a comprehensive echocardiographic evaluation in addition to the Wilkins Score, and degree and mechanism of mitral regurgitation (MR) [22,27]. For patients with a low Wilkins Score (< 8) and not in atrial fibrillation, there is no difference between event-free survival and clinical events for PMBV versus surgery [28]. According to the 2014 AHA/ACC Guidelines for the Management of Patients with Valvular Heart Disease, PMBV is recommended for symptomatic patients with severe MS and favorable valve morphology in the absence of left atrial thrombus or moderate-to-severe MR [Class I, level of evidence (LOE) A] [29]. Mitral valve surgery is indicated for patients with severe symptomatic MS who are not candidates for or have failed percutaneous mitral balloon commissurotomy (Class I, LOE B) [30].

4. Conclusion

Our patient had multiple concerning aspects from her history when evaluating for the cause of her presenting hemoptysis, including a history of pulmonary tuberculosis and longstanding active smoking. She had no known history of streptococcal pharyngitis, acute rheumatic fever or residence in a region where rheumatic heart disease remains endemic. Her cardiac murmur initially raised the possibility of valvular disease. A comprehensive work-up eventually resulted in a diagnosis of severe rheumatic mitral stenosis initially presenting with massive hemoptysis. The patient had sporadic and sparse follow-up with medical providers throughout her life which likely contributed to her mitral stenosis becoming so severe.

This case demonstrates that pulmonary venous congestion can be seen on bronchoscopic examination in severe mitral stenosis and describes the workup and management. Furthermore, this patient’s case exemplifies that despite the decreased incidence of rheumatic heart disease in developed countries, valvular heart disease should remain on the differential for patients presenting with hemoptysis.

Conflict of interest

Nothing to declare.

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