CLINICAL VIGNETTE

Aortopulmonary Fistula in a Patient Presenting with Hemoptysis, Fevers, and Dyspnea after a Chest Deceleration Injury

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Introduction

Fistulas between the aorta and adjacent structures are rare but can be fatal if not promptly identified and treated. Most new diagnoses of aortopulmonary fistulas in adults are associated with a history of trauma or vascular intervention. The fistula creates backflow from the systemic to the pulmonary circulation which results in elevated pulmonary artery pressure, increased right heart strain and ultimately right heart failure. Presenting symptoms include hemoptysis, fever, chest pain, and shortness of breath. Clinicians should have a high suspicion for this entity in patients who have a history of aneurysms, vascular repair, or trauma and present with bleeding manifestations, or right heart failure.

Case Report

We present a 61-year-old male with history of bicuspid aortic valve requiring aortic valve replacement x 3, nonischemic cardiomyopathy, atrial fibrillation/flutter, and ascending aorta repair with Hemashield graft placed in 2018. The patient presented to his local hospital with 9 days of chills and fevers up to 102°F. He denied chest pain, abdominal pain, or urinary symptoms, but reported occasional dry cough as well as malaise, fevers, and dyspnea. His initial vital signs included temperature of 36.7°C, blood pressure was 101/61 mm Hg, and heart rate 80 bpm, O2 saturation was 96% on room air. Exam included a previously known pansystolic II/IV murmur at the right sternal border, but was otherwise unrevealing. Chest radiograph showed minimal patchy airspace opacities at the bases, likely atelectasis and no osseous abnormalities. Urinalysis was normal. COVID and influenza PCR tests were negative. Electrocardiogram showed no evidence of acute ischemia or arrhythmia. The patient was admitted and started on Vancomycin and Ceftriaxone empirically for endocarditis, though no further fevers were recorded during his hospitalization.

After blood cultures remained negative after 48 hours, a transthoracic echocardiogram (TTE) was discussed to evaluate for culture-negative endocarditis given his extensive cardiac history. On further questioning the patient reported a deceleration injury several weeks prior. He described being 'thrown backwards' during a boating incident and striking his back on a cooler. He presented to the ED. Chest radiograph was

negative for acute osseous abnormalities, but he was told he had posterior rib fractures and sent home with pain medications. Since this accident he had been taking shallow breaths and occasionally coughing up blood-tinged phlegm. He was able to visit a local theme park 3 weeks after the injury and walk about 5 miles, although he felt fatigued. However, his symptoms progressed until his ED presentation. Although his clinical picture was consistent with pulmonary contusion/pneumonia, given his significant cardiac history a TTE was obtained. The American College of Cardiology Foundation (ACCF), in partnership with the American Society of Echocardiography (ASE), advise TTE evaluation after chest trauma when valve or cardiac injury are possible.⁵

TTE revealed a new aortopulmonary fistula, severely elevated pulmonary pressures (PASP 69 mm Hg), moderate to severe pulmonary regurgitation, severely dilated bilateral atria, and <50% IVC collapse during respiration. Subsequent computed tomography (CT) angiogram revealed an ascending aortic to main pulmonary artery shunt measuring approximately 16 x 16 mm, focal dissection along anterior aspect of ascending thoracic aorta, progressive ascending aortic root dilation (reported 4.6 cm in 2017, now measuring 5.7 cm), and a retrosternal fluid collection measuring 1.3 x 3.6 x 12 cm superficial to the anterior pericardium which was inseparable from the anterior ascending aorta. Notably, the CT angiogram also revealed mildly displaced subacute right 7th-11th rib fractures. Transesophageal echocardiogram showed no valvular vegetations.

The patient was emergently transferred to a tertiary care center for cardiac surgical evaluation. Infectious Diseases obtained Karius pathogen blood test and tagged white blood cell scan which were negative for any infection. As endocarditis had been ruled out, the patient was cleared to proceed with surgery. Given 3 prior sternotomies and clinical complexity, the shunt was addressed endoscopically and a 5 cm covered stent was placed in the pulmonary artery with reduction in angiographic shunting from the aorta to the pulmonary artery from moderately severe to minimal. Simultaneously, moderately severe pulmonary homograft regurgitation was addressed with transcatheter pulmonary valve replacement with subsequent minimal residual regurgitation. Mild distal homograft stenosis

at baseline with a peak-to-peak gradient of 12 mmHg was reduced to negligible after stent and valve placement.

Two clinical issues remained after stent placement. First the 6cm neo-aortic root dilation and secondly, focal dissection above the right coronary artery which could enlarge. Given prior surgically challenging aortic repair, the Adult Congenital Heart Disease (ACHD) and the cardiac surgery team decided to defer surgery and repeat imaging in 3 months to assess for interval change. The patient was discharged on enoxaparin with ACHD clinic follow up. Today the patient is doing well, walks up to a mile a day, and follows regularly with his cardiology specialists.

Discussion

Aortopulmonary fistulas are rare complications of ascending aorta pathology and have been sporadically reported. Our patient with extensive cardiac surgery history who presented with fevers and hemoptysis was investigated for endocarditis, only to discover a new diagnosis of an aortopulmonary fistula. Aortopulmonary fistula can be found during evaluation of chronic diseases such as pneumonia, lung abscess, and mycotic aortic aneurysm. However, they usually occur after erosion or rupture of a degenerative or false aneurysm of the distal aortic arch or descending thoracic aorta into the lung. They can also be caused by trauma or postoperative complications after aortic surgery, including aortic valve replacement.

The symptoms of aortopulmonary fistula commonly include chest pain, hemoptysis, and dyspnea or other respiratory symptoms. Hemoptysis, which is characteristically intermittent or recurrent, occurs when the hematoma "leaks" into the bronchopulmonary tree due to aortic rupture. Interestingly, our patient presented with no clinical signs of heart failure despite severely elevated pulmonary pressures on echocardiogram.

The hemodynamic consequences of a large aortopulmonary fistula include rapid cardiac decompensation and prompt diagnosis is crucial. Transthoracic echocardiography is a useful diagnostic method. High oxygen saturation in the PA blood and an oxygen step-up between the right atrium and the PA, as determined during cardiac catheterization, are confirmatory. As a rule, aortopulmonary fistula should be treated surgically and percutaneous coil embolization has shown favorable outcomes. In addition, endovascular stent-graft repair of aortopulmonary fistula appears to be safe and well tolerated.

Summary

In this patient the dissection and aortopulmonary fistula were likely due to a contained rupture sustained after the deceleration injury in the setting of known aortic atherosclerotic disease. They were hypothesized to be subacute. The traumatic event may have led to local disruption and the formation of an aortopulmonary fistula that gradually enlarged with progressive dyspnea and hemoptysis. While chest contusion with pneumonia could have explained this clinical picture, given his

significant cardiac history, TTE to rule out culture negative endocarditis vs cardiac damage turned out to be a revealing and likely lifesaving intervention.

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