Case Report

Pulmonary artery dissection complicating aortic dissection in a patient with bicuspid aortic valve and aortic coarctation

Minhaj S. Khaja MD, MBAa,*, Richard L. Hallett MDb

a Vascular & Interventional Radiology, Department of Radiology, University of Michigan Health System, 1500 E. Medical Center Drive, Ann Arbor, MI, USA
b Department of Medical Imaging, St. Vincent Hospital Indianapolis, Stanford University School of Medicine, Indianapolis, IN, USA

ABSTRACT

We report a case of a type A aortic dissection with extension into the main pulmonary artery through a sinus of Valsalva fistula. Echocardiography and computed tomographic angiography of the chest were performed and bicuspid aortic valve, hemopericardium, and dilatation of the aortic root were found. A Stanford type A dissection was seen, extending to the distal transverse thoracic aorta, and there was a communication between the dissection at the left sinus of Valsalva and the main pulmonary artery, where a dissection flap was detected at computed tomographic angiography. This case report reviews the rare diagnosis of pulmonary artery dissection, multimodality imaging findings, and a brief review of etiology and management.

© 2017 the Authors. Published by Elsevier Inc. under copyright license from the University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Introduction

There is a wide differential diagnosis to consider when evaluating patients with chest pain. Among the more concerning diagnoses are myocardial infarction, pulmonary embolism, and aortic dissection. In many cases, a more benign cause such as a primary pulmonary infection or musculoskeletal etiology is found. We report a case of a type A aortic dissection with extension into the main pulmonary artery through a sinus of Valsalva fistula.

Case report

Institutional review board's approval is not required for preparation of a case report at our institution. A 25-year-old previously healthy man presented to the emergency department complaining of chest pain, shortness of breath, and throat tightness. He had a normal chest x-ray and laboratory tests at that visit and was sent home with symptomatic treatment for a viral syndrome. He returned 6 days later with visual loss in his right eye and leg weakness in addition to chest pain and shortness of breath. Physical examination revealed a loud, holosystolic murmur and decreased radial and femoral pulses. The rest of the physical examination was unremarkable.

A ventilation-perfusion scan showed a significant perfusion mismatch in the entire left lung, interpreted as high probability for pulmonary embolus (Fig. 1). Echocardiography and computed tomographic angiography (CTA) of the chest were performed (Figs. 2 and 3). A bicuspid aortic valve (BAV),
hemopericardium, and dilatation of the aortic root were found. A Stanford type A dissection was seen, extending to the distal transverse thoracic aorta. In addition, there was a communication between the dissection at the left sinus of Valsalva and the main pulmonary artery, where a dissection flap was detected at CTA. Juxtaductal aortic coarctation was also demonstrated, with dilated intercostal and internal mammary collaterals. No pulmonary embolism was found.

The findings at echocardiography and CTA were confirmed at surgery, where an intimomedial tear was seen extending into the left sinus of Valsalva, near the origin of the left coronary artery. The tear extended through the sinus into the main pulmonary artery, causing the pulmonary dissection. Hemopericardium and left hemothorax were also confirmed. Aortic valve resuspension and graft replacement of the ascending aorta and arch were performed. Graft repair was also extended to the descending aorta to repair the aortic coarctation. The fistula from the sinus of Valsalva and the pulmonary dissection were repaired. The patient was transferred to the Cardiac Recovery Unit in stable condition.

Fig. 1 – (A and B) There is homogeneously decreased perfusion throughout the left lung (arrows), without focal defects. RPO, right posterior oblique; LAO, left anterior oblique; RL, right lateral; LL, left lateral; LPO, left posterior oblique; RAO, right anterior oblique; RT, right.

Fig. 2 – (A) Axial CTA image shows the ascending aortic dissection flap (arrow), as well as the true and false (arrowheads) lumens of the pulmonary dissection. Pericardial effusion (H) shown to be hemopericardium at surgery is seen, as are bilateral pleural effusions (E). (B) Oblique coronal CTA reconstruction demonstrates rupture of the left sinus of Valsalva into the pulmonary trunk (arrow), causing pulmonary dissection. The true lumen of the pulmonary dissection is identified (*). (C) Oblique sagittal volume-rendered CTA image demonstrates postductal coarctation (arrowheads), multiple intercostal collaterals (arrows), dilated internal mammary artery collateral (**), and dilated aortic root.
Discussion

Pulmonary artery dissection is a rare disease with a high mortality, with predominately case reports noted in the literature [1–5]. It most commonly occurs secondary to pulmonary arterial hypertension, but has been reported with right heart endocarditis, amyloidosis, catheterization, and chronic pulmonary artery inflammation [1–3]. As the condition is extremely rare, the diagnosis is frequently not considered in most differential lists. However, BAV has a well-known association with aortic coarctation, and patients with BAV and coarctation show significantly increased incidence of aneurysm and dissection compared with BAV alone. Aortopathy with BAV is linked to decreased metalloproteinase activity and vascular smooth muscle apoptosis, causing increased risk of aneurysm and dissection [4].

Treatment of pulmonary aortic dissection is not well defined. However, medical management of resulting pulmonary hypertension, surgical repair of entry site, lobectomy, and transplantation have all been briefly described [2,3,5]. Aortic and pulmonary artery dissection should be considered in patients with chest pain or shortness of breath with any of the above risk factors. Multimodality imaging may aid in confirming the diagnosis and treatment planning.

References

[1] Neimatallah MA, Hassan W, Moursi M, Al Kadhi Y. CT findings of pulmonary artery dissection. Br J Radiol 2007;80:e61–3.
[2] Mutlu H, Demir IE, Mutlu LN, Doyle D. Pulmonary artery dissection mimicking mediastinal mass. Heart Lung 2009;38:233–7.
[3] Inayama Y, Nakatani Y, Kitamura H. Pulmonary artery dissection in patients without underlying pulmonary hypertension. Histopathology 2001;38:435–42.
[4] Braverman AC, Guven H, Beardslee MA, Makan M, Kates AM, Moon MR. The bicuspid aortic valve. Curr Probl Cardiol 2005;30:470–522.
[5] Adodo DK, Kloekker M, Bergoend E, Couetil JP. Pulmonary artery dissection: a case treated by homograft replacement. Ann Thorac Surg 2017;103:e47–9.