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Case Report

Percutaneous closure of a giant pulmonary arteriovenous malformation using multiple devices

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ABSTRACT

Percutaneous device closure is an established method to treat pulmonary arteriovenous malformation (PAVM). This report describes the case of a 23-year-old man with hereditary hemorrhagic telangiectasia (HHT) presenting with dyspnea and hypoxia. The patient was found to have a giant left-sided PAVM. The patient underwent percutaneous closure of PAVM with multiple devices with a good outcome. At 12-month follow up, the patient was asymptomatic with near complete obliteration of the arteriovenous malformation.

<**Learning objective:** This case demonstrates the feasibility and safety of using multiple devices to treat pulmonary arteriovenous malformation percutaneously. Good short-term (12-month) outcomes are demonstrated and there were no complications.>

Introduction

Pulmonary arteriovenous malformations (PAVMs) are rare, yet an important cause of extracardiac right to left shunt. More than 50% of PAVMs are associated with hereditary hemorrhagic telangiectasia or Osler–Weber–Rendu syndrome. Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominantly inherited condition characterized by visceral arteriovenous malformations. About three-fourths of the patients will have abnormal physical findings including pulmonary bruit, clubbing, cyanosis, or mucocutaneous telangiectasia [1]. The right to left shunting in PAVMs has been associated with multiple complications including brain abscess, seizures, and stroke [2]. In addition, spontaneous hemothorax [3] and massive hemoptysis [4] may occur secondary to the rupture of these arteriovenous malformations, especially in large lesions. If left untreated 25–35% of patients will have a stroke or brain abscess [5]. All PAVMs of size at least 3 mm have to be treated to prevent these potentially lethal complications. The treatment options include surgical resection of the involved lung segment or percutaneous occlusive device therapy. Percutaneous embolization therapy has a high success rate in the closure of PAVMs with low complication rate.

Case report

A 23-year-old male presented with symptoms of exertional dyspnea of New York Heart Association Class II, intermittent headache and easy fatiguability for a duration of two years. The patient had symptoms of recurrent spontaneous epistaxis until three years before presentation. The patient's mother and elder brother also had symptoms of recurrent spontaneous epistaxes. On examination, the patient was deeply cyanosed with an arterial oxygen saturation of 82% while breathing room air. He had pandigital grade III clubbing (Fig. 1a). Telangiectatic lesions were noted in the oral cavity. His right arm blood pressure was 110/76 mmHg. The precordial examination was unremarkable. On further evaluation, his chest X-ray revealed a well defined homogenous opacity in the middle of his left lung field suggestive of pulmonary arteriovenous malformation (Fig. 1b). The patient was clinically diagnosed to have HHT based on Curaçao criteria. Contrast echocardiography was performed which showed almost immediate opacification of left-sided chambers. A 64-slice computed tomography of the thorax confirmed the presence of PAVM consisting of a conglomeration of multiple, tortuous, dilated, and intensely enhancing vascular channels in inferior lingular segment of left upper lobe (Fig. 1c). The lesion measured

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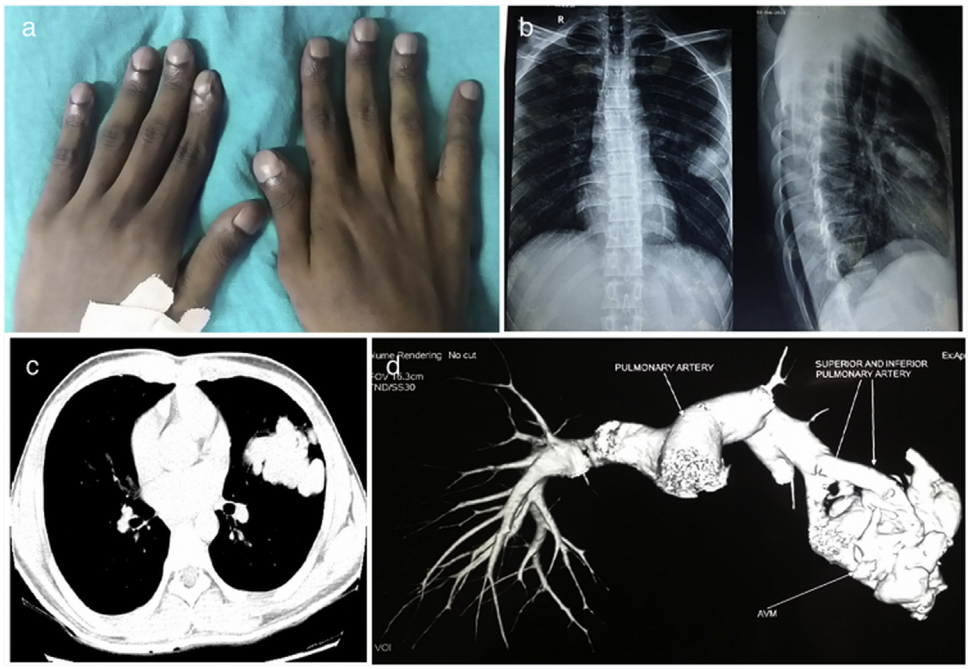

Fig. 1. (a) Pandigital uniform clubbing with cyanosis. (b) Chest X-ray showing homogenous radio-dense lesion in the left lung field suggestive of pulmonary arteriovenous (AV) malformation. (c) Computed tomography (CT) pulmonary angiogram showing pulmonary AV malformation. (d) 3D reconstruction of CT pulmonary angiogram showing the feeders of the malformation.

63 mm × 49 mm × 47 mm. Feeding arteries originated from the superior and inferior lingular segmental branches which measured 13 and 10.5 mm respectively (Fig. 1d). The two major feeders coursing into the lesion measured 8 and 7 mm respectively. The two draining veins (14 and 7 mm) emptied into the superior pulmonary vein. Contrast enhanced computed tomography of the abdomen revealed heterogenous enhancement of liver parenchyma suggestive of multiple telangiectasias and also evidence of a small splenic hemangioma. Brain imaging did not reveal any arteriovenous malformations. Upper gastrointestinal endoscopy showed telangiectatic lesions in fundus, cardia, and body of stomach. After discussing with the patient, the options of surgery versus percutaneous procedure, percutaneous device closure of PAVM was planned.

Procedure notes

Under local infiltrative anesthesia, a 7French introducer sheath was placed in the right common femoral vein. The right heart was catheterized with a 6F multipurpose (MPA) catheter. The right ventricular systolic pressure was 20 mmHg and the mean pulmonary artery pressure was 16 mmHg. The catheter was then directed to engage the left pulmonary artery and a pulmonary angiogram showed the two major feeding arteries supplying the PAVM. This was followed by selective cannulation of the descending branch of left pulmonary artery into the anomalous vessels. Successive selective contrast injection into the major feeders revealed multiple small secondary channels supplying the arteriovenous malformation and two large draining veins (Fig. 2a and b). The target vessels for occlusion were the superior (7 mm) and inferior (8 mm) major feeders. The inferior major feeding artery was wired first with 0.035″ Hiwire (Cook Medical, Bloomington, IN, USA). This was followed by a 7F SteerEase (Lifetech Scientific Co., Shenzhen, P.R. China) delivery sheath into the inferior feeder. A 12-mm vascular plug system (Cera, Lifetech Scientific Co., Shenzhen, P.R. China) was delivered into the neck of the major inferior feeder (Fig. 2c). The device was positioned to occlude all the secondary channels. However, post deployment, a

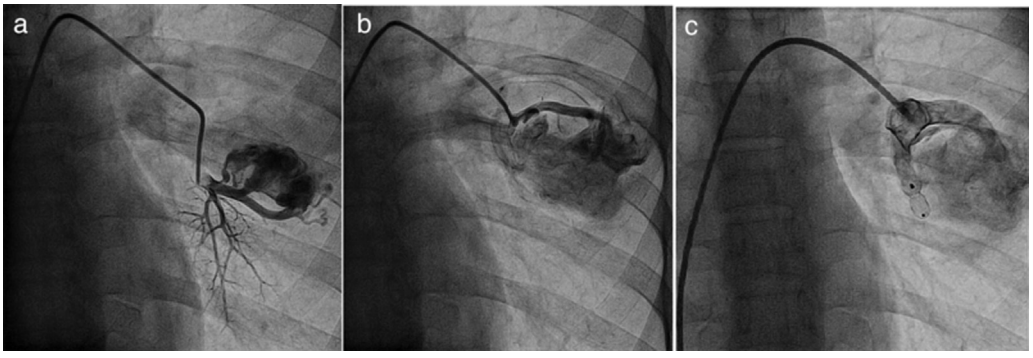

Fig. 2. (a) Selective injection of the inferior feeder showing the multiple small secondary feeders. (b) Selective injection of the superior feeder showing the arteriovenous malformation and the draining vein. (c) Fluoroscopic image after the deployment of a vascular plug system in the major inferior feeder.

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