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Case Report

Transcatheter occlusion of complex pulmonary arteriovenous malformations in a cyanotic child

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ABSTRACT

This case report presents a 16-month-old boy with isolated mild cyanosis diagnosed to have complex pulmonary arteriovenous malformations. Three-dimensional computed tomography with volume rendering has provided the diagnosis, enabled detailed imaging, and facilitated transcatheter device occlusion of the complex arteriovenous malformations by vascular plugs and coils. Magnetic resonance of the brain revealed associated dural arteriovenous malformation. Genetic testing showed a missense disease-causing variant in the *ENG* gene that encodes endoglin, and the diagnosis of hereditary hemorrhagic telangiectasia was made.

<**Learning objective:** The initial clinical presentation of isolated mild cyanosis in a child with pulmonary arteriovenous Malformations (PAVMs) has led to the diagnosis of hereditary hemorrhagic telangiectasia may pose a diagnostic challenge. The use of three-dimensional computed tomography with volume rendering enables the diagnosis of complex PAVMs and facilitates the planning of transcatheter device occlusion.>

Introduction

Pulmonary arteriovenous malformations (PAVMs) occur with an incidence of 2 to 3 per 100,000 [1]. While the age at first presentation varies widely, the majority of patients is diagnosed within the first three decades of life. About 80–95% of PAVMs are associated with hereditary hemorrhagic telangiectasia (HHT) [2]. The clinical presentation depends on the magnitude of intra-pulmonary right-to-left shunting, which is related to the number and size of PAVMs. Patients may therefore be completely asymptomatic or present with cyanosis at rest or during exercise. Paradoxical embolization through PAVMs with or without coexisting cerebral arteriovenous malformations account for the neurological events, which include migraine, transient ischemic attack, stroke, cerebral abscess formation, and seizure [3]. Less common but acute life-threatening complications include hemoptysis and hemothorax due to intra-bronchial rupture of PAVMs or endobronchial telangiectasia and rupture of sub-pleural PAVMs [4]. We present the case of a child with complex PAVMs that required a combination of vascular plugs and coils for closure.

Case report

A 16-month-old boy was incidentally found to have mild cyanosis during preoperative assessment for repair of his right hydrocele. His oxygen saturation in room air ranged from 82 to 90%. The child was otherwise asymptomatic with no history of recurrent epistaxis. Physical examination revealed mild central cyanosis. There was no detectable cutaneous telangiectasia. His hemoglobin level was 16.4 g/dL. Chest radiograph showed an abnormal opacity over the upper lobe of the left lung. Two-dimensional echocardiographic examination of the heart was normal.

High-resolution computed tomography (CT) of the thorax showed a cluster of abnormally dilated vessels in the superior segment of the left upper lobe, and reformatted images showed connection of the left pulmonary artery to vein through these vessels (Fig. 1). The early arterial phase scans demonstrated strong enhancement of most of the abnormal vessels and left upper pulmonary vein. These findings were suggestive of fast and high flowing PAVMs. Further magnetic resonance imaging (MRI) of the brain and liver was found to be normal. The parents were explained

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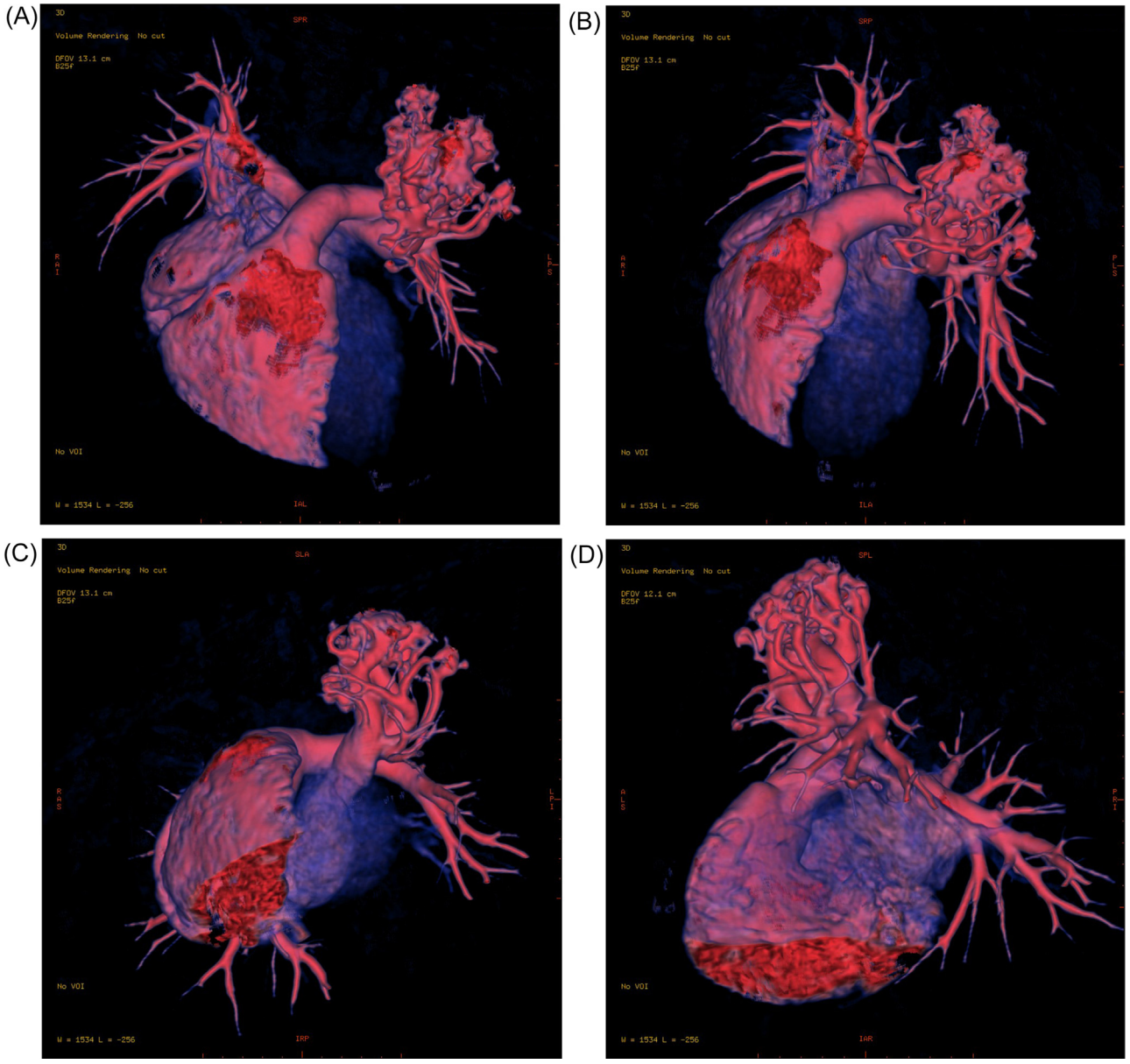

Fig. 1. Volume-rendered three-dimensional computed tomography images of the complex pulmonary arteriovenous malformations in our patient in different orientations (A, anterior, B, anterolateral, C, lateral, D posterior). A cluster of abnormally dilated vessels, connecting the left pulmonary artery to the left pulmonary vein, is shown in the superior segment of the left upper lobe.

about the findings and, given their concerns of the potential risks of intervention in the young, transcatheter closure of the PAVMs was planned at an older age or when the child became more cyanotic.

After a minor head injury at the age of 4 years 10 months, the patient had another MRI of the brain, which revealed a meshwork of tortuous high-flow vascular structure at the cortical surface of the anterior left frontal region consistent with dural arteriovenous malformation (Fig. 2). Cerebral digital subtraction angiography showed two arteriovenous malformations at the left frontal region. Neurosurgical intervention at an older age was planned. The child was otherwise free of cardiac symptoms and having satisfactory exercise capacity with an oxygen saturation well above 80%.

Transcatheter embolization of PAVMs was performed at 5 years of age when the child weighed 19.8 kg and his oxygen saturation was lowered to about 80%. Pulmonary angiography showed extensive PAVMs malformations involving the left upper lobe (Fig. 3). The feeding arteries were occluded by seven Amplatzer vascular plug IV with diameters of 4–8 mm (Abbott Vascular, Saint Paul, MN, USA) and two Flipper detachable embolization coils (Cook Medical Inc., Bloomington, IN, USA). Our selection of the devices was based on the consideration of (i) the ease of the delivery catheters to negotiate through the tortuous large feeding vessels, (ii) the avoidance of the need to manipulate the stiffer guiding catheters if Amplatzer vascular plugs I and II were to be used, and (iii) the size of the target vessels. Systemic arterial oxygen saturation rose from 75% to 93% immediately after the procedure. The pulmonary arterial pressures were normal before and after the embolization of the PAVMs. There were no peri-procedural complications and antiplatelet or anticoagulation therapy was not used. At 1 month and 1 year of follow up, the oxygen saturation was 95% and 96%, respectively.

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