Open Med. 2015; 10: 513–518

Case Report **Open Access**

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A case of Multiple Unilateral Pulmonary arteriovenous Malformation Relapse: Efficacy of embolization treatment

DOI: 10.1515/med-2015-0087
received October 26, 2015; accepted November 4, 2015.

Abstract: Pulmonary arteriovenous Malformations (PAVMs) are a rare vascular alteration characterized by abnormal communications between the pulmonary arteries and veins resulting in an extracardiac right-to-left (R-L) shunt. The majority of PAVMs are associated with an autosomal dominant vascular disorder also known as Osler-Weber-Rendu Syndrome. PAVMs appearance can be both single and multiple. Clinical manifestations include hypoxemia, dyspnea cyanosis, hemoptysis and cerebrovascular ischemic events or abscesses. We report a case of an 18 year old female with severe respiratory failure caused by a relapse of multiple unilateral pulmonary arterovenous fistula. Symptoms at admission include dyspnea, cyanosis and clubbing. The patient underwent pulmonary angio-TC scan, brain CT and echocardiography. The thoracic angio-CT scan showed the presence of PAVMs of RUL and RLL; a marked increase of right bronchial artery caliber and its branches with an aneurismatic dilatation was also observed. The patient underwent percutaneous transcatheter embolization using Amplatzer Vascular Plug IV; a relevant clinical and functional improvement was subsequently recorded. Embolization is effective in the treatment of relapsing PAVMS.

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Keywords: Pulmonary arteriovenous Malformations (PAVMs), Embolization, Unilateral fistula, Angio-CT Scan.

1 Introduction

Pulmonary arteriovenous Malformations (PAVMs) are a rare vascular alteration with an overall incidence estimated to be approximately two to three per 100.000 population. PAVMs are abnormal communications between the pulmonary arteries and veins, which bypass the pulmonary capillary bed resulting in an extracardiac right-to-left (R-L) shunt between the pulmonary artery and pulmonary vein [1]. The majority of patients with PAVMs have an hereditary hemorrhagic telangiectasia (HHT), an autosomal dominant vascular disorder also known as Osler-Weber-Rendu Syndrome [2]. The remaining cases are idiopathic, however other causes associated to infections, trauma and Fanconi Syndrome have been described [1]. Diagnosis of PAVMs is not straightforward, as symptoms are common to other cardiovascular and respiratory diseases, related to both individual and environmental factors including cigarette smoking [3-17]. The major clinical manifestations are hypoxemia, dyspnea and cyanosis, the severity of which depend upon the extent of lesions. Serious morbidity and mortality are largely related to complications including paradoxical embolization causing cerebrovascular ischemic events or cerebral abscesses; migraine and hemothorax have also been reported. PAVMs are described according to their anatomical characteristics. Approximately 85% of PAVMs are simple, where the arterial supply arises from one or more branches of a single segmental pulmonary artery [18]. Most of the remainder are complex PAVMs which have multiple arterial feeder vessels from more than one pulmonary segment, while a smaller percentage of PAVMs are diffuse with disseminated involvement of multiple pulmonary segments [19]. The diagnosis of

PAVMs is often made as a result of a chest X-ray or a chest CT scan which is more accurate in differential diagnosis and extension of PAVMs. On chest X-ray, the classic sign of PAVMs is a well-defined round or oval nodule or mass. Additional noninvasive methods to assess PAVMs are quantitative right-to-left shunt studies (100% oxygen method or radionuclide perfusion scanning) and contrast echocardiography. Pulmonary angiography is generally now reserved for post-diagnosis therapeutic purposes [1,3,20]. The fistula-type PAVMs have a feeding artery directly connected to a draining vein, with an intervening single aneurysmal sac while, less commonly, PAVMs are plexiform with a multiseptated aneurysm or a cluster of vascular channels. In the past, symptomatic PAVMs were treated surgically but since the introduction of embolotherapy, percutaneous trans-catheter embolization with coils has significantly decreased the rate of complications [19,21]. According to the International HHT Guidelines, PAVMs should be embolised preventatively to decrease the risk of complications irrespective of symptomatology [22].

2 Case Report

We report a rare case of an 18 year old female with severe respiratory failure caused by Multiple unilateral pulmonary artero-venous fistula. The patient was admitted to the Department of Cardio-Thoracic and Respiratory Science at the Second University of Naples with progressive shortness of breath that had gradually worsened during the previous two month period. Family history was negative for both genetic and cardiovascular diseases. In 2010, following an episode of haemoptysis, the patient underwent an angio-CT scan which revealed marked dilation of bronchial arteries in the right lung as well as dilatation of venous branches approaching superior and inferior pulmonary veins, leading to a diagnosis of artero-venous shunt of right lung with alveolar hemorrhage, involving the majority of the inferior right lobe and lateral segment of median lobe. The patient, then, underwent selective angiography and embolization of the AV fistula using the Gianturco method. Until 2013, the patient was clinically stable, with follow up angio-CT scans showing no disease progression. In 2014 the patient presented with severe dyspnea and was admitted to hospital for assessment. On examination she was found to be dyspneoic at rest with marked cyanosis and finger clubbing. Oxygen saturation on air was 83-84% and blood gas analysis showed pO2: 50 mmHg and pCO2: 33 mmHg with little improvement demonstrated on administration of high flow oxygen. Thoracic angio CT scan, cranial and maxillofacial CT, echocardiogram and abdominal ultrasound were carried out to exclude Rendu-Osler Syndrome. The thoracic angio-CT scan showed the presence of Multiple Artero-Venous Fistulas (MAVFs) of Right Upper Lobe (RUL) and Right Lover Lobe (RLL) with an increase of pulmonary venous caliber compared to left lung field; a marked increase of right bronchial artery caliber and its branches was also observed in addition to an aneurismatic dilatation of 1 cm not previously detected; Web-like appearance was evident in the superior segment of RUL (Figure 1). Pulmonary angiogram, carried out before the embolization procedures, confirmed the presence of MAVFs involving the superior and inferior lobes of the right lung; aortography showed the presence of a systemic-pulmonary bridge arising from the thoracic aorta towards the right lung. Percutaneous transcatheter embolization (TCE) was successfully performed using 8 Amplatzer Vascular Plug IV (St. Jude Medical, Saint Paul - Mn, USA) devices, as well as inserting 2 Amplatzer Vascular Plug IV (St. Jude Medical, Saint Paul - Mn, USA) devices within the systemic-pulmonary bridge (Figure 2). These procedures resulted in a clinical and functional respiratory improvement. Within 90 days, a progressive stabilization of blood gas parameters was achieved. Thoracic angio-CT scan carried out six months post-embolization showed a substantial normalization of the arteriovenous morphology within the right lobes without bronchial pseudo aneurysm; enlargement of some vessels of ventral segment of RUL and few peripheral MAVFs persisted at the level of the superior segment of the RLL (Figure 3).

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

Informed consent: Informed consent has been obtained from all individuals included in this study.

3 Discussion

Our case confirms that patients with PAVMs require lifelong clinical, radiological and cardio-respiratory function assessments as recanalization and collateralization may recur after embolization therapy. We have shown that percutaneous TCE is effective and safe for the treatment of disease relapse. Diagnosis of PAVMs may be intriguing as clinical, physiopatological and radiological characteristics may mimic other cardio-respiratory diseases arising from different cellular and biomolecular pathways [1,3,23-40]. Percutaneous TCE is recognized as the gold standard

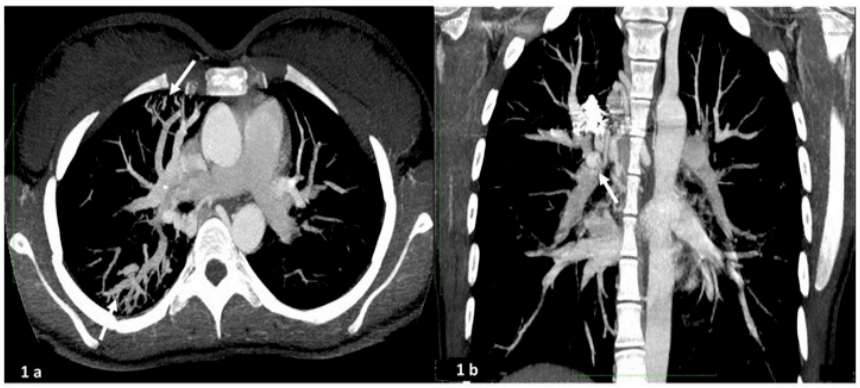

Figure 1: Thoracic angio-CT scan at admission shows: presence of multiple AV Fistulas of RUL and RLL, increase of pulmonary venous and right bronchial artery and its branches caliber associated to aneurismatic dilatation of 1 cm. Web-like appearances are evident in the superior segment of RUL. Transversal (a) and Coronal (b) view.

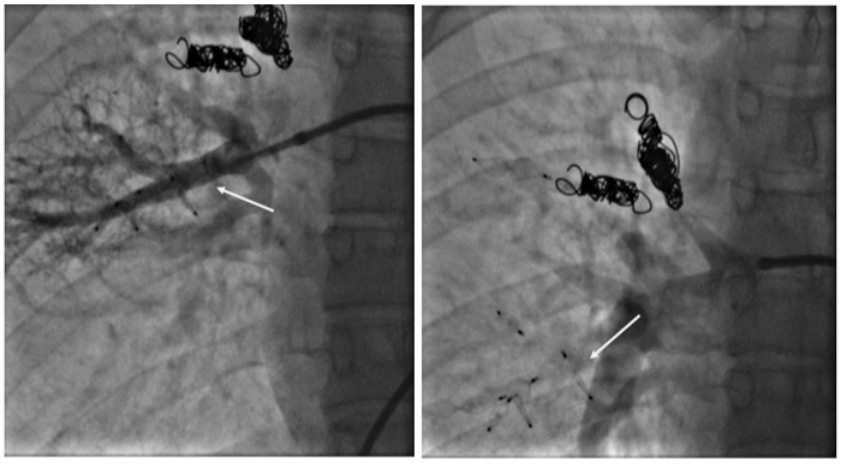

Figure 2: Embolization procedures using the Amplatzer Vascular Plug IV (8 devices) as well as insertion of Amplatzer Vascular Plug IV (2 device) within the systemic-pulmonary bridge.

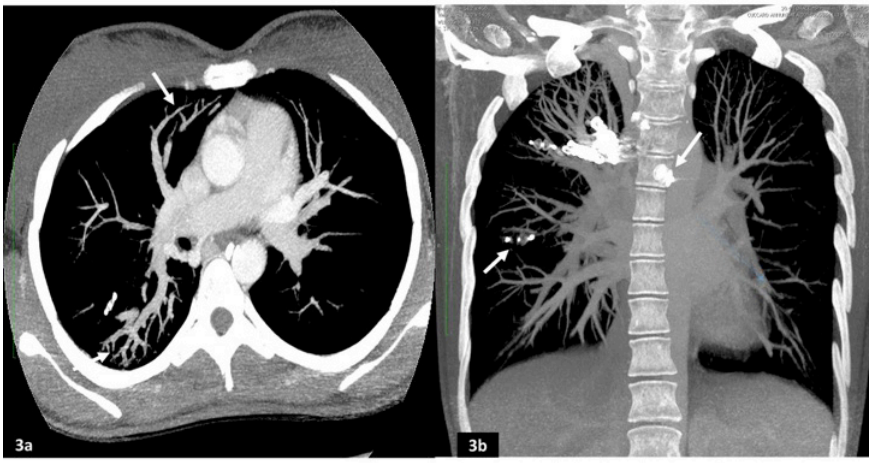

Figure 3: Thoracic angio-CT after embolization. Transversal (a) and Coronal (b) view.

for treatment of PAVM due to its efficacy and safety in reducing the risk of paradoxical embolism and other complications associated with PAVM [41-45]. Major indications for treatment are prevention of neurological complications, including stroke and cerebral abscess from paradoxical embolism, improvement in exercise tolerance, reduction in migraine prevalence, and prevention of lung hemorrhage [1,3]. Advantages over surgical intervention include it being less invasive and easy to repeat, although collateralization and revascularization over time have to be taken into account. Regardless of symptoms, any PAVM with a feeding artery of ≥ 3 mm in diameter detected by CT scan should be considered for therapy using this technique. During embolization, the supplying artery immediately preceding the PAVM is the target for occlusion just proximal to the aneurysmal sac. It is important to achieve an embolization as distal as possible in the feeding artery to avoid occluding branches supplying normal adjacent lung. The choice of embolization device depends on the vascular anatomy of the patient and the practice preference of the operator. A variety of materials are used to embolize PAVMs, depending on the size and complexity of the vessels. In general, PAVM with feeding artery diameters of between 3 and 9 mm are treated with balloons or coils, whereas those with diameters > 8 mm may be treated with either coils alone, an overinflated balloon impacted within a nest of coils, or with vascular plugs. Magnetic resonance-compatible steel or platinum coils are used in the majority of cases. Depending on the anatomy of the specific PAVM, other techniques may be employed. For example, if the neck of the feeding vessel is particularly large or wide, the aneurysm sac may be packed. The deployed coils are designed to coil within the vessel lumen and carry microfibers that activate platelets to generate an occluding platelet plug, whereas Amplatzer vascular plugs and balloon devices provide direct obstruction to vascular flow.

4 Conclusions

Our case describes a relapsing multiple MAVFs within the right lung in an 18 year old woman. The monolaterality of the malformation excludes Rendu-Osler Syndrome; etiopathogenesis remains unknown, although the young age at presentation suggests a miopragia of the vessel walls as possible cause of the vascular malformation. As relapses are not infrequent in PAVMs, patients should undergo regular clinical and radiological assessment. Embolization is effective in the treatment of relapsing disease.

Conflict of interest statement: Authors state no conflict of interest

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