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CASE REPORT

Aneurysm of an Anomalous Systemic Artery Supplying the Normal Basal Segments of the Left Lower Lobe: Endovascular Treatment with the Amplatzer Vascular Plug II and Coils

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Abstract An anomalous systemic artery originating from the descending thoracic aorta supplying the normal basal segments of the lower lobe of the left lung without sequestration is a rare congenital anomaly. The published surgical treatments include lobectomy, segmentectomy, anastomosis, and ligation. In addition, endovascular treatment with coils has been reported. A second-generation occluder, the Amplatzer Vascular Plug II (AVP II), has a central plug and two occlusion disks and a finer, more densely woven nitinol wire, thus enabling faster embolization. This published case is the first successful occlusion of an aneurysm of an anomalous systemic artery with the AVP II and fibered coils, with 10 months of follow-up.

Keywords Systemic arterial supply to the lung · Anomalous artery · Aneurysm · Amplatzer Vascular Plug

Introduction

A systemic artery supplying the basal segments of the lower lobe which has no structural or bronchial abnormalities is a rare congenital anomaly and also the rarest form of congenital anomalous systemic arterial supply to the lungs [1]. Aneurysm of the anomalous artery has been reported before [2]. This congenital anomaly was previously classified as Pryce type I sequestration [3]. Recently some synonyms such as systemic arterial supply to the basal segments of the lung, systemic origin of the sole artery to the basal segments of the lung, systemic arterialization of the lung without sequestration, and pseudosequestration of the lung have been used for this congenital anomaly [4–7].

Reported surgical treatments include lobectomy, segmentectomy, anastomosis between the anomalous artery and the pulmonary artery, and ligation of the anomalous artery [8–10]. In addition, transcatheter closure with coils has been reported [7, 11, 12]. The Amplatzer Vascular Plug II (AVP II; AGA Medical Corp.n, Golden Valley, MN, USA) is a second-generation occluder with a finer, more densely woven nitinol wire and a three-segment design for arterial and venous embolization, which enables faster occlusion [13, 14].

This published case is the first successful occlusion of an aneurysm of the anomalous systemic artery arising from the descending aorta with the AVP II and fibered coils.

Case Report

A 53-year-old man was admitted to our hospital with a new episode of acute chest pain. His pain intensifed with inspiration on the left side. He had a history of smoking for about 23 years (one pack per day) and occasional coughing. He had not experienced hemoptysis. The only remarkable finding in the physical examination was some rales in the lower zone of the left lung. Laboratory tests were within normal range except for D-dimer, which was 1747 ng/ml (normal range, 0–500 ng/ml).

Doppler ultrasonography was performed and acute thrombosis was observed in the left great saphenous vein.

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Multidedector computed tomography (MDCT) examination of the thorax, which was performed with a 64-dedector scanner (Aquillion 64; Toshiba Medical Systems, Tokyo) to rule out pulmonary embolism, revealed thromboembolic occlusive disease in the left upper segmental pulmonary arteries. In addition, an aneurysm (50 × 48 × 43 mm) with mural calcification and partial thrombosis originating from a large, tortuous anomalous vessel, which arose from the descending aorta, and a ground-glass appearance of the basal segments of the lower lobe of the left lung were observed. The artery distal to the aneurysm gave off branches to the normal basal segments of the lower lobe of the left lung. Transverse images with a lung window setting demonstrated normal bronchial branching of the left lower lobe and absence of the pulmonary artery to the basal segments (Fig. 1).

In the third month of medical treatment with anticoagulation, a control MDCT showed complete improvement of the thromboembolism in the pulmonary arteries. Also, no change in aneurysm diameters was observed, but a decreased volume of the partial thrombosis and increased diameters of the filling lumen with blood into the aneurysm were noted. The ground-glass opacity in the affected segment was also clarified properly. Because of the clinical history and the potential risk of massive hemoptysis due to aneurysm and pulmonary hypertension, we decided to perform endovascular treatment.

Before the procedure, informed consent was obtained. During the procedure, the patient received 100 IU/kg heparin and the activated clotting time was kept at 200–250 s. Under local anesthesia, a 6-Fr, 90-cm straight guiding sheath (Destination; Terumo Co., Tokyo), with the assistance of a 5-Fr diagnostic catheter (Cobra-1; Terumo Co.) and a 0.035-in. angled guidewire (Radifocus; Terumo Co.), was advanced into the anomalous artery arising from the descending thoracic aorta via the right brachial artery, just proximal to the aneurysm. A diagnostic angiogram was obtained, in which the aneurysm, the distal arterial branches that were supplying the left lower basal segments, and venous drainage via the inferior pulmonary vein, subsequently, were seen (Fig. 2A). A 2.7-Fr microcatheter (Progreat; Terumo Co.) was carefully passed across the aneurysm into the distal branches. First, distal branches were embolized with multiple 0.018-in. fibered coils (Vortex-18 fibered platinum coils; Boston Scientific/Target Vascular, Cork, Ireland) to decrease the blood flow and to prevent retrograde filling by unexpected collaterals. Then a 14-mm AVP II was deployed in the artery proximal to the aneurysm, which was measured as 10 mm on MDCT. At first deployment, the proximal occlusion disk of the device prolapsed inward to the aorta. At this stage, the plug was recaptured by pulling the delivery wire back slightly and advancing the delivery system back over the plug until the entire device was within the delivery system. Then the guiding sheath was repositioned and the delivery cable was pushed forward during the second deployment, and this enabled more precise placement (Fig. 2B). A control angiogram was obtained via the guiding sheath, confirming appropriate device position, and it was released by rotating the delivery cable in counterclockwise fashion. Finally, thoracic aortography was obtained just after the embolization, which showed total occlusion of the embolized anomalous artery (Fig. 2C). The postprocedure course was uneventful and the patient was discharged 1 day later on antibiotic therapy to prevent superinfection.

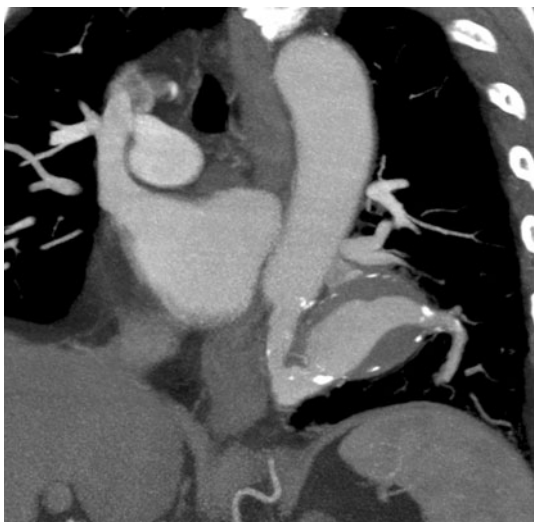

Fig. 1 Multiplanar reconstructed image shows an aneurysm with mural calcification and partial thrombosis originating from a large, tortuous anomalous vessel, which arises from the descending thoracic aorta. The artery distal to the aneurysm gives off branches to the normal basal segments of the lower lobe of the left lung

In the first week after embolization, the patient was admitted to our hospital with chest pain and coughing. MDCT examination of the thorax to rule out recurrent pulmonary embolism revealed no thromboembolic occlusive disease. Total occlusion of the anomalous artery and aneurysm, as well as consolidation and ground-glass appearance in the basal segments of the lower lobe of the left lung, was seen (Fig. 3A). In addition, a volume loss in the lower lobe of the left lung and minimal pleural effusion adjacent to the affected basal segments of the lower lobe were demonstrated. Postembolization syndrome was considered and the patient was treated conservatively with analgesics.

A follow-up MDCT angiography scan 10 months later demonstrated no recanalization of the embolized artery and improved consolidation and ground-glass appearance (Figs. 3B, C). The patient remained asymptomatic.

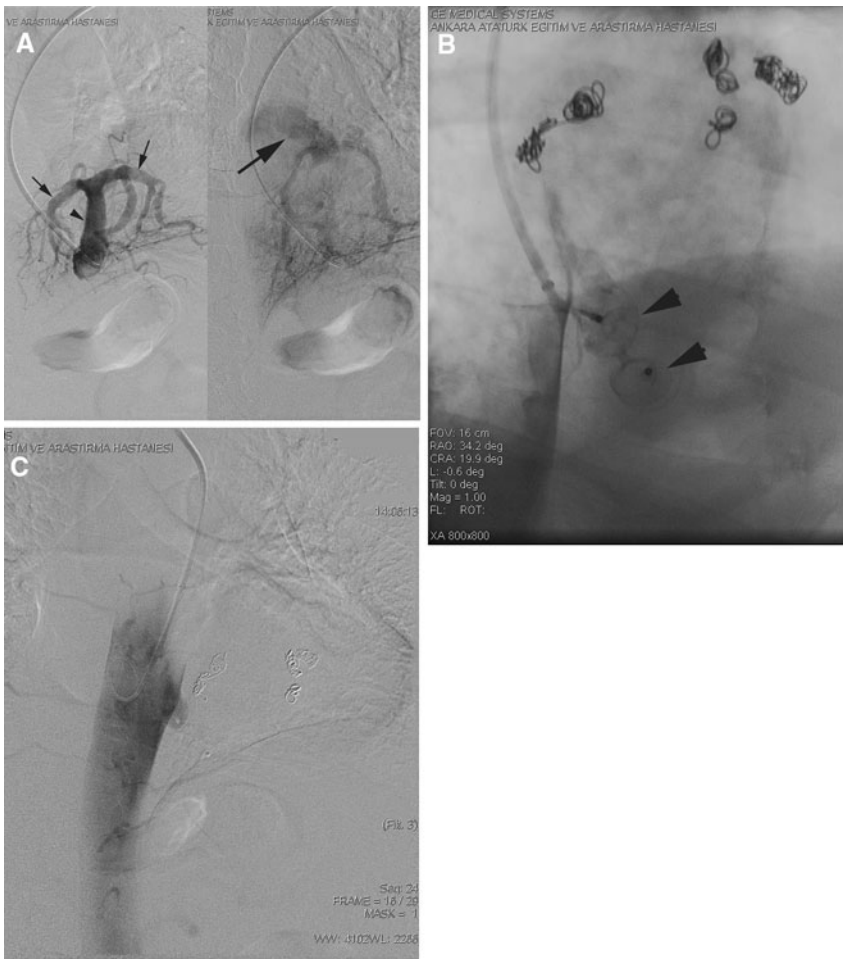

Fig. 2 **A** Diagnostic angiogram shows the filling lumen of the aneurysm (*small arrowhead*) and distal arterial branches (*small arrows*) which supply the left lower basal segments, and venous drainage via the inferior left pulmonary vein (*large arrow*), subsequently. **B** Test injection through the guiding sheath after the second deployment shows proper positioning of the AVP II (*arrowheads*) in the targeted vessel. **C** An arteriogram obtained just after embolization demonstrates complete occlusion of the aneurysm

Discussion

An anomalous systemic artery arising from the thoracic aorta supplying the normal basal segments of the lower lobe, absence of a pulmonary arterial supply, and a normal bronchial system are the characteristic imaging findings of this anomaly [1]. The left lower lobe is more frequently involved than the right lower lobe. On the left side the anomalous systemic artery originates from the descending thoracic aorta, whereas on the right side it usually arises from the abdominal aorta or the celiac artery [7, 9]. Although there have been a few case reports of aneurysm of the anomalous artery, endovascular treatment of it has not been reported previously as far as we know [2].

Although some authors place this entity within the spectrum of pulmonary sequestration such as Pryce et al. classification (type I, abnormal artery without sequestration; type II, abnormal artery to sequestrated mass and adjacent normal lung; type III, abnormal artery confined to sequestrated mass), this entity differs from classic bronchopulmonary sequestration in that the involved lung tissue includes a normal connection to the bronchial tree. In addition, the absence of the pulmonary artery to the basal segment of the left lower lobe is considered an important differentiation from bronchopulmonary sequestration [1–12].

Although the etiology of this anomaly is unknown, the most likely theory is the persistence of an embryonic connection between the aorta and the pulmonary parenchyma. At the fourth week of embryonic life, the lung bud is fed by systemic capillaries, the splanchnic plexus or primitive bronchial arteries, which have lots of connections with the primitive dorsal aorta. The ventral and dorsal sixth branch is connected and forms the sixth arch. At the fifth week, the pulmonary arteries (plexiform vessels) appear and grow toward the lung bud from the sixth aortic arch. At the sixth week the primitive aortic postbranchial branches regress and the pulmonary arterial system becomes established. The eventual bronchial arteries develop between the 9th and the 12th week and communicate with the capillary bed in the airway walls. Inefficacy of passage from the systemic to the pulmonary artery circulation may cause a persistent systemic arterial supply to an otherwise normal lung [9, 15].

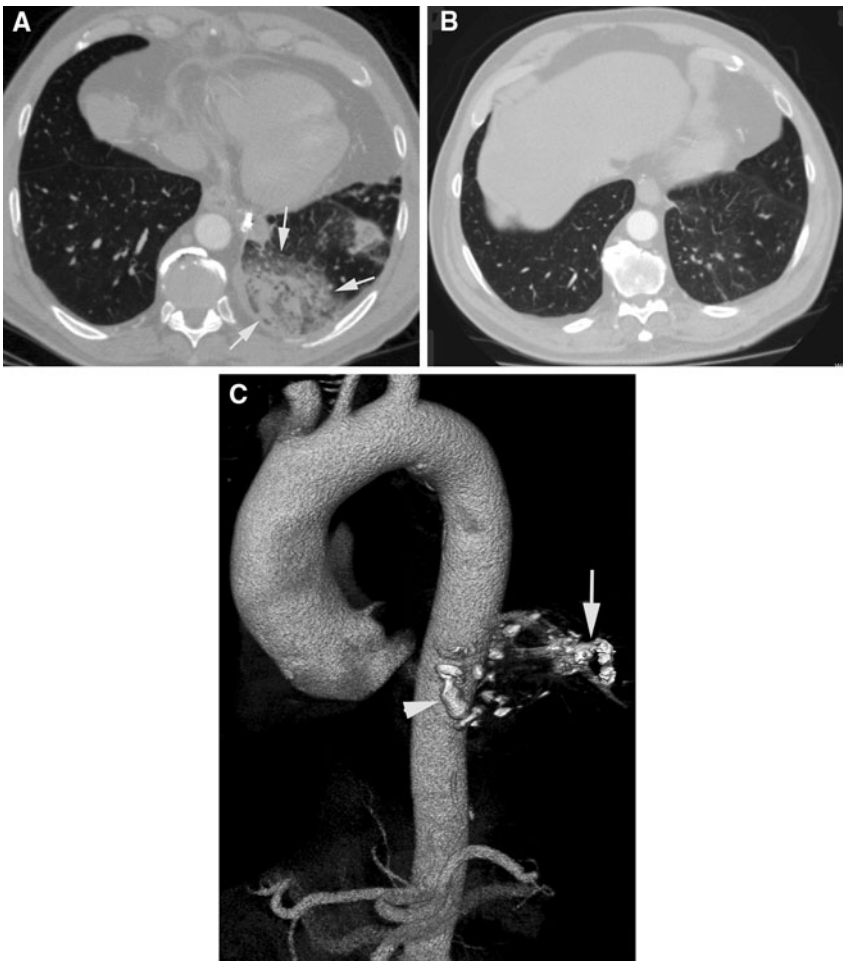

Fig. 3 **A** One-week follow-up contrast-enhanced MDCT examination shows consolidation and ground-glass appearance (*arrows*) in the basal segments of the lower lobe of the left lung owing to pulmonary infarction following the vaso-occlusion. **B** A subsequent MDCT examination at 10 months demonstrates improvement of the consolidation and ground-glass appearance. **C** Three-dimensional volume-rendered image shows complete occlusion of the anomalous artery with the AVP II (*arrowhead*) and coils (*arrow*)

Conversely, many theories have suggested that anomalous persistent systemic arteries may have a developmental or acquired rather than a congenital etiology, for example, in chronic inflammation hypertrophy of normal vessels can develop within the inferior pulmonary ligament [15].

The main symptoms are continuous murmur in pediatric age groups and episodes of hemoptysis in adulthood. However, adult patients are usually asymptomatic as in our case [4–9]. Because of the long-standing exposure of these vessels to high systemic arterial pressure, diffuse dilatation of the peripheral pulmonary vasculature and areas of ground-glass opacity with a mild volume decrease are almost always noted in the involved basal segments on CT scans. Regions of ground-glass opacity in the involved basal segments are thought to indicate a mild degree of pulmonary congestion. The congestion may or may not be associated with alveolar hemorrhage induced by the high systemic arterial pressure [1].

Traditionally the treatment is surgery, which is indicated for all patients with this anomaly because it has potential risks such as congestive heart failure caused by left-sided cardiac overload, recurrent severe infections, and massive hemoptysis due to pulmonary hypertension [8]. In the literature, lobectomy or segmentectomy has been performed in most cases with this condition [9]. Besides, anastomosis between the anomalous artery and the pulmonary artery has been described in managing this condition [8, 9]. Another choice of treatment is ligation of the anomalous artery [10].

Successful endovascular treatment with coil embolization of the systemic anomalous artery has also been reported in two adult patients who presented with hemoptysis and chest pain [11, 12]. Although there were no major complications in these two cases, and there was the advantage of lack of surgery, one of the cases had postembolization syndrome after the procedure as in our case. The authors concluded that the postembolization syndrome could be explained by a pulmonary infarction following the vaso-occlusion [11].

The first-generation AVP is an alternative permanently occluding self-expanding cylindrical device made of a single layer of nitinol wire mesh and secured on both ends with platinum marker bands similar to the Amplatzer septal and ductal occluding devices, which has been used increasingly for different applications in interventional

radiology since 2004 [16–18]. The second-generation plug, the AVP II, which was introduced in 2007, is a cylindrically shaped, more densely woven nitinol-mesh occluder with a central plug and two occlusion disks, thus enabling faster embolization [13, 14]. Additionally, the AVP II is available in small and large diameters (11 sizes, from 3 to 22 mm), in contrast to the original plug (7 sizes, from 4 to 16 mm). It is recommended to select a device approximately 20–50% larger than the vessel diameter for secure fitting, prevention of device migration, and total occlusion. The position of the device can be easily verified with a test injection through the guiding sheath. If the position is not satisfactory, as in our case, the plug can be recaptured by pulling back slightly on the delivery wire and advancing the delivery system back over the plug until the entire device is within the delivery system, then it can be repositioned and redeployed.

In this case, the AVP II was chosen for permanent occlusion of the anomalous artery proximal to the aneurysm due to sufficient length and proper size of the targeted vessel and because it allows for targeted delivery, enabling more precise placement within the artery. First, distal branches of the artery are embolized with coils to decrease blood flow and retrograde filling by unexpected collaterals. If the aneurysm is very close to the aorta, an aortic stent-graft may be considered as another choice for endovascular treatment.

In conclusion, in selected cases, the endovascular treatment of an aneurysm of the anomalous systemic artery arising from the descending aorta with the newly introduced AVP II and coils is simple, effective, and an alternative method for surgery.

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