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Successful endovascular embolization of an intralobar pulmonary sequestration

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ABSTRACT

Pulmonary sequestration is a congenital malformation characterized by dysplastic pulmonary tissue which receives blood supply by arterial systemic system, not in communication with tracheobronchial tree. Although it could be asymptomatic, it can also cause recurrent infections and hemoptysis, rarely massive and fatal.

The conventional treatment consists in surgical resection of the pulmonary sequestration, but in the last few years endovascular embolization has been proposed as a valid therapeutic alternative. In this paper, we report the case of a 43–year-old woman affected by recurrent hemoptysis. Computed tomography angiography of the chest, abdomen, and pelvis was performed in emergency setting. Intralobar pulmonary sequestration in the lower lobe of the right lung was found. A bulky aberrant artery originating from the thoracic aorta supplied the pulmonary sequestration. The interventional radiologist performed an endovascular embolization with coils of the vascular malformation.

The technical success of the procedure was confirmed by computed tomography angiography of the chest performed on the fourth day after procedure. Further examination performed 6 months later showed no complications.

The patient was completely asymptomatic during follow-up. This procedure can demonstrate that arterial embolization is a valid and effective therapeutic alternative to surgical resection in the treatment of pulmonary sequestration.

Competing Interests: All the authors declare that they have no conflicts of interest.

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Introduction

Pulmonary sequestration is a congenital malformation consisting in dysfunctional pulmonary tissue, supplied by aberrant systemic arteries, not in connection with bronchial tree. Its incidence is estimated to be between 0.15% and 1.8% of the general population [1–5]. From a pathologic point of view, there are 2 types of pulmonary sequestration: intralobar and extralobar type.

Intralobar pulmonary sequestration is a dysfunctional pulmonary tissue covered by visceral pleura with venous drainage into pulmonary veins. Usually it is located in the lower lobes, especially on the left side [5]. Malformative tissue is often heterogeneous (with fibrosis, cysts, or bronchiectasis) as a consequence of acute or chronic inflammatory processes [6].

Extralobar pulmonary sequestration is a pulmonary tissue with independent pleura and systemic venous drainage [1,2,4,5].

Clinically, extralobar pulmonary sequestration may become symptomatic in the first weeks of life, because of the presence of left-to-right shunt, whereas intralobar type is more frequently diagnosed in young adults affected by recurrent pneumonia [1,2]. A dangerous complication of intralobar type is hemoptysis, although it rarely becomes massive [1,6].

Surgical resection is currently the gold standard treatment of pulmonary sequestration [7,8]. Recent studies have shown that endovascular embolization is a valid and effective therapeutic option for symptomatic pulmonary sequestration. It is a minimally invasive approach associated with lower morbidity compared with surgical resection [5,8].

Cases of extralobar pulmonary sequestration treated with endovascular embolization have been reported in the literature with positive results [1,7,8]. On the other hand, only few cases of intralobar type treated with endovascular treatment have been reported [1].

In this paper, we report the case of a 43–year-old woman affected by recurrent episodes of hemoptysis, due to a massive intralobar pulmonary sequestration. An endovascular embolization of the malfomation with multiple coils was performed. Patient was successfully treated.

Technique

A 43-year-old woman (no smoking in anamnesis) presented to our emergency room because of repeated episodes of hemoptysis in the last 3 days. No chest pain, dyspnea, fever, weight loss, or recent trauma was reported. Moreover, the patient had no history of anticoagulant therapy or coagulative pathologies. Vital parameters were normal. Computed tomography angiography of chest, abdomen, and pelvis was perfomed. The CT revealed the presence of an arteriovenous malformation in the right lower pulmonary lobe.

The arterial in-flow resulted from an abnormal vessel originating from the descending thoracic aorta (Fig. 1A, B, and C). Venous out-flow was supplied from ipsilateral lower pulmonary vein. The pulmonary parenchyma surrounding vascular malformations was not in communication with bronchial tree and there were small areas of “ground-glass” consolidation (Fig. 1D).

The diagnosis of intralobar pulmonary sequestration was suspected. An angiographic study was performed so as to confirm the diagnosis and to eventually treat the malformation.

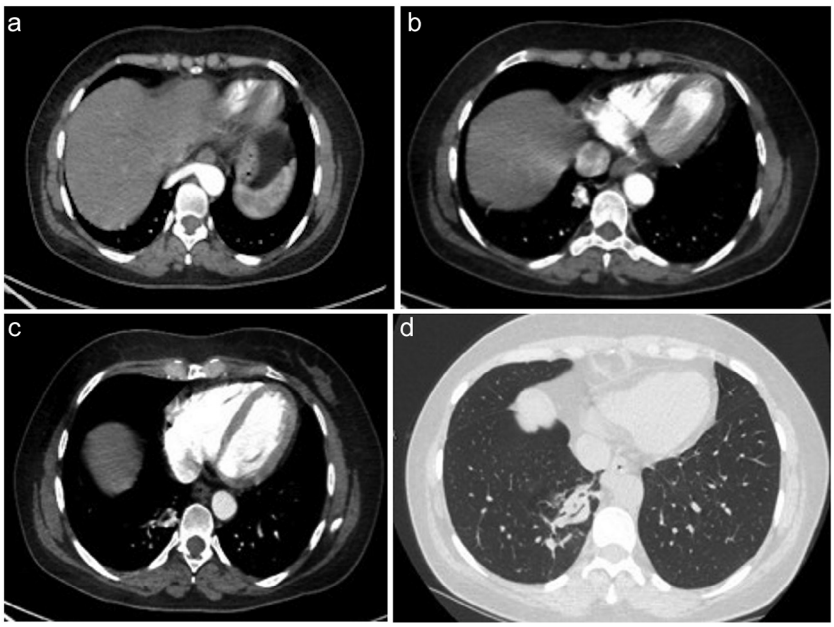

Fig. 1 – (A, B, C, and D) Pre-embolization chest CT examination (arterial phase) on axial planes showing the presence of a large systemic anomalous arterial branch (A) emerging from the right anterolateral wall of the descending thoracic aorta and subdivided in further abnormal arterial branches (B, C) resulting in arteriovenous malformation in the basal medial lobe of the lower lobe of the right lung. The arteriovenous malformation is surrounded by thickened lung parenchyma and areas of “ground glass” (D). CT, computed tomography.

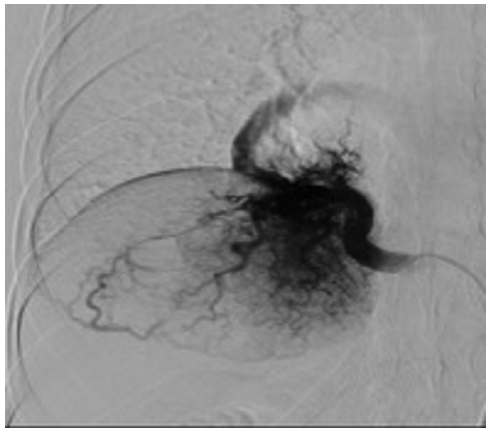

Fig. 2 – Digital angiographic study; selective catheterization of the main vessel emerging from right anterolateral aortic wall with Cobra catheter. Presence of arterial-venous malformation in the lower right lung lobe with blood outflow in the lower right pulmonary vein.

The interventional radiologist performed the endovascular procedure through a right transfemoral arterial access and selective catheterization of arterovenous malformation with vascular catheter (Cobra 5 Fr, Merit-Impress Hydrophilic Catheter, Merit Medical Systems, South Jordan, UT, USA). Diagnostic arteriography confirmed the presence of the abnormal arterial vessel originating from the right anterolateral wall of the descending thoracic aorta. The selective catheterization was performed and confirmed the presence of intralobar pulmonary sequestration (Fig. 2). The endovascular embolization was made through super-selective catheterization of the vascular malformation with coaxial technique by 2.7 Fr micro-catheter (Terumo-Progreat, Lane Somerset, New Jersey, USA). As an embolizing material, multiple metal micro-coils (Nester type, Cook Medical, Nester Embolization coil, Bloomington, Indiana, USA) ranging from 6 to 8 mm inside the arterial vessels of pulmonary sequestration was used (Fig. 3A, B, C, and D).

In conclusion, 3 metal coils (Nester type: 12 mm and 14 mm, Cook Medical) were positioned, directly from the main catheter (Cobra 5 Fr, Merit), isolating the main afferent artery trunk from the descending aorta.

In this case, we considered appropriate the use of metallic coils as embolizing material. Their distal compaction was effective in achieving an adequate slowing down of arterial inflow, avoiding embolization in nontarget vessels and preventing migration and displacement.

Postprocedural angiography showed complete devascularization of the malformation (Fig. 3E). A chest CT examination with intravenous contrast medium was performed 4 days after the procedure and confirmed the complete de-vascularization of the pulmonary sequestration. The patient was asymptomatic.

Patient has never experienced hemoptysis again and remained asymptomatic for 6 months after the procedure. The technical success of the procedure was confirmed by the follow-up chest CT angiography 6 months later (Fig. 4A, B, C, and D).

Discussion

Pulmonary sequestration is a rare, congenital malformation defined as a portion of pulmonary parenchyma which receives abnormal vascularization from systemic arteries and is not in connection with the bronchial tree [1,2]. It is divided in

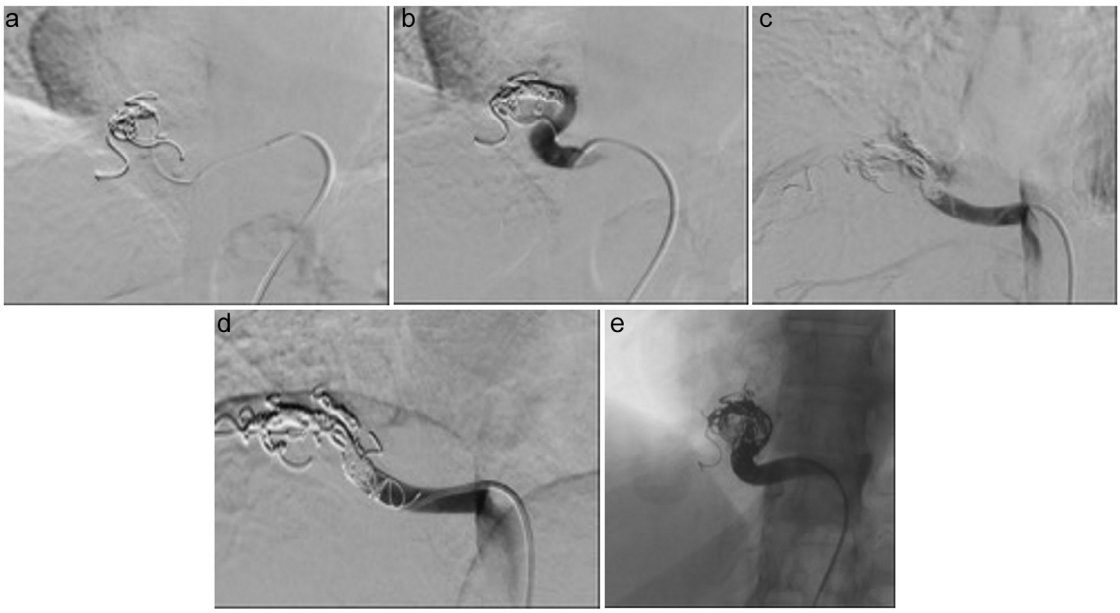

Fig. 3 – (A, B, C, D, and E) Arterial embolization. Coils released in vascular malformation and progressive embolization of vascular malformation with co-axial technique in the "distal-proximal" direction (A). Intra-procedural angiographic checks (B, C, and D). Final angiographic check (E) after release of 3 metallic coils from the Cobra catheter demonstrating complete devascularization of vascular malformation.

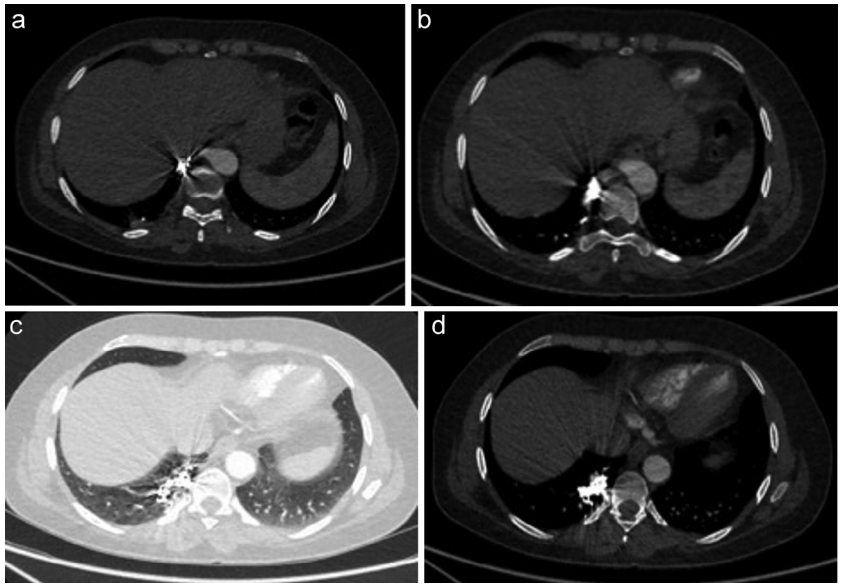

Fig. 4 – (A, B, C, and D) Post-embolization chest CT examination; 6-month follow-up (arterial phase), axial planes. Complete embolization and atrophy of the systemic arterial vessel that supplied the pulmonary sequestration (A, B) and complete devascularization of the vascular malformation (C, D). CT, computed tomography.

2 variants: intralobar and extralobar pulmonary sequestration [1–3,6].

The intralobar variant constitutes the most common form of pulmonary sequestration (between 75% and 86% of pulmonary sequestration), whereas the extralobar variant is less frequent (between 14% and 25%) [1,2,6].

The difference between intra- and extralobar pulmonary sequestration is in the pleural coating: the extralobar variant has an independent pleural coating, whereas the intralobar form shares the pleural coating with the adjacent lobe.

In addition, they have different types of venous drainage: in the intralobar sequestration, venous drainage is typically carried out from a pulmonary vein, whereas in the extralobar sequestration, the venous drainage goes directly in the systemic venous circle (frequently in the lower lobar vein or in azygos vein). In the literature some authors reported cases of subclavian vein or portal vein drainage [1,3,6].

In both cases, arterial vascularization is provided by abnormal systemic vessels that usually originate from descending thoracic aorta (73%), intercostal arteries (4%), or from the cranial portion of the abdominal aorta, the celiac trunk, and splenic artery (21%) [1,2,6]. As far as their clinical manifestation is concerned, extralobar forms are usually present in children, whereas intralobar forms (the most symptomatic) are usually present in adult people.

However, approximately 30% of the pulmonary sequestration are incidental findings [4–6]. Patients affected by intralobar pulmonary sequestration may be asymptomatic or affected by chest pain, dyspnea, recurrent lung infections, hemoptysis (rarely massive and fatal) [1–3,5].

Some authors believe that etiology of hemoptysis could be due to high pressure in arterial circulation deriving from abnormal systemic vessels that supply the pathologic lung tissue [3,5]. Another fatal, but very rare, complication is fibrinoid necrosis that causes hematic pleural effusion [1,2].

First-level imaging examination for patients affected by pulmonary sequestration is CT with endovenous contrast medium [5]. Imaging has 2 main goals:

1. to exclude the presence of other thoracic pathologies;
2. to demonstrate the presence of an abnormal arterial supply.

It is very important to demonstrate the presence of systemic arterial supply to the pulmonary sequestration so as to establish a definitive diagnosis and to exclude the presence of other diseases (such as bronchiectasis or bronchial atresia). Diagnosis, therefore, is based on demonstration of systemic arterial supply to pulmonary sequestration [6]. The gold standard for diagnosis of pulmonary sequestration is angiography. Digital angiography is able to demonstrate the presence of abnormal arterial vessels and pathologic venous drainage. With the development of imaging techniques, less invasive methods have proven to be equally effective and safer than angiography, such as CT and magnetic resonance imaging.

Currently, CT angiography is the most noninvasive method used in clinical practice in case of pulmonary sequestration. CT angiography, indeed, is able to find out pathologic arterial supply and pathologic venous drainage, and it provides an accurate study of pathologic lung parenchyma and airways [1,6].

From a historical point of view, surgical resection of intra- and extralobar pulmonary sequestration is the gold standard treatment. On the one hand, surgery prevents recurrent infections and hemoptysis and preserves the normal adjacent lung. On the other hand, it is associated with morbidity and complications for the patient [1,3,5,6]. Recently, many authors reported that endovascular embolization of systemic arterial

abnormal vessels is a valid alternative therapeutic choice. Endovascular embolization, indeed, reduces arterial inflow to abnormal lung tissue, causing necrosis, and progressive fibrosis and involution.

In addition, pathologic vessels in pulmonary sequestration are more friable and acute or chronic inflammatory processes could cause aneurysms or other vascular malformation. As a consequence, endovascular embolization reduces not only hemoptysis but also bleeding in case of surgery [1,3,5–8].

Few cases of endovascular embolization have been reported in case of intralobar pulmonary sequestration in adult patients. All the cases reported in the literature have been treated with endovascular embolization, without recurrence during follow-up periods [1].

In our case, endovascular embolization was effective and the patient no longer had cases of hemoptysis in the following 6 months.

Recurrence has been reported in 25%-47% of cases treated with endovascular embolization due to an ineffective embolization, an incomplete closing of the vessels, displacement of the embolic agents, or opening of collateral circles and shunts.

Minor complications of endovascular treatment could be migration of embolic materials in nontarget vessels, pain, and infection at site of puncture [1,6].

In our case, we decided to embolize not only the main vessels but also the sub-segmentary vessels of the vascular malformation so as to prevent a belated opening of collateral circles and shunt.

Technical success of the procedure was obtained by performing an embolization with coils and micro-coils, following the "distal-proximal" direction technique. We released micro-coils in the smallest vessels of vascular malformation at first, and in the main arterial trunk at the end. Following this technique, we tried to avoid displacement and migration of coils in the afferent trunk.

Not only coils are the most used embolic agent, but polyvinyl alcohol (PVA) particles, micro-coils, gelatin sponge, alcohol, glue (n-butylcyanacrylate), and Amplatzer plugs have also been reported in the literature [1,3,5].

Combinations of embolic materials have also been reported by authors, such as use of PVA particles together with coils [1]. PVA particles (500-700 μ) slow down the flow in the smaller vessels and prevent migration of the coils.

In conclusion, endovascular arterial embolization is an effective and minimally invasive technique that should be considered among therapeutic options in the treatment of patients with symptomatic pulmonary seizures as an alternative to surgery. However, further prospective studies are needed to determine the long-term outcomes of the procedure in the patients treated.

The main points of this case report are listed below:

- to describe from a clinical and a radiological point of view a rare case of pulmonary sequestration;
- to describe materials and technique of endovascular embolization of the vascular malformation; and
- to review literature about endovascular treatments in case of pulmonary sequestration.

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