ELSEVIER

Endovascular Treatment of Symptomatic Pulmonary Sequestration

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Pulmonary sequestration is a rare congenital malformation whose origin is bronchial and arterial simultaneously and its vascularization comes from an anomalous systemic artery. Its clinical presentation includes recurrent pneumopathy in the same anatomic location of the lung and difficult to resolve or recurrent lung abscess. It is usually treated with antibiotherapy and eventual surgical resection. A 23-year-old woman with history of recurrent respiratory infections and three episodes of hemoptysis was admitted at the hospital. Computed tomography and magnetic resonance imaging confirmed diagnosis of pulmonary sequestration. The angiographic study showed the presence of three inflow arteries arising from the thoracic aorta (T10) and supplying the abnormal lung parenchyma at the base of the left hemithorax. The patient underwent endovascular treatment consisting of exclusion of the inflow vessels with Amplatzer occlusive devices and coils. Subsequent computed tomography angiogram confirmed complete infarction of the sequestration. At 7 months, the patient presented with a new episode of bronchial infection. Repeated angiography showed persistence of intermediate small nutrient branches that were treated with coil embolization. The patient is symptom-free at 41 months after this secondary procedure. Endovascular treatment of pulmonary sequestration, with selective embolization of the inflow arteries, is a very attractive minimally invasive therapeutic option, as compared with conventional surgery, and potentially less prone to associated complications.

Normal pulmonary parenchyma has a dual blood supply that comes from two sources: the first origin is the pulmonary artery and the second involves the bronchial arteries directly arising from the aorta. Both sources are connected at the level of the respiratory bronchioles, where pulmonary and bronchial capillaries freely anastomose.

An abnormal communication between the pulmonary and systemic circulations may appear as a result of either congenital or acquired pathologies; most notably, the former include pulmonary sequestration.

This is a rare congenital malformation whose origin is bronchial and arterial simultaneously. It consists of an abnormal pulmonary tissue that has no identifiable connection with the bronchial tree. Its blood supply comes exclusively from one or more anomalous systemic arteries rather than the pulmonary circulation.[1] Its clinical manifestations are recurrent pneumopathy in the same anatomical location. It is usually difficult to be solved, and occasionally presents complications such as recurrent pulmonary abscess or hemoptysis. It is usually treated with antibiotherapy and surgical resection.[2]

We describe the case of a patient with symptomatic pulmonary sequestration treated with selective embolization of the inflow arteries and its subsequent course.

CLINICAL CASE

History and Clinical Presentation

A 23-year-old woman, without a history of congenital or pediatric diseases, was admitted at the hospital because of a 2-month history of purulent sputum and left chest pain, including three recent episodes

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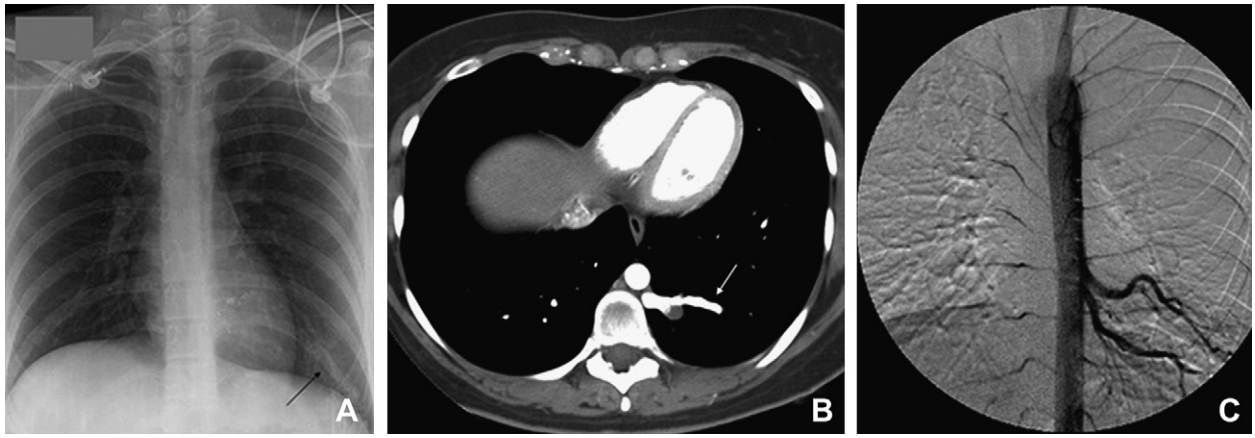

Fig. 1. Initial imaging tests. Anteroposterior thoracic X-ray (**A**), the *black arrow* points to the opacity band at the base of the left hemothorax. Computed tomography angiography (**B**), *white arrow* points to the anomalous inflow vessel arising from the thoracic aorta and crossing at the base of the left hemothorax. Both images correspond to the pulmonary sequestration site. The initial diagnostic aortography (**C**) reveals two major abnormal arteries that originate from the thoracic aorta, supplying abnormal pulmonary parenchyma.

of hemoptysis. The patient reported not having a smoking habit, any previous exposure to air pollution, or the presence of traumatisms or seasonal diseases. She denied experiencing any fever, weight loss, or dyspnea and her physical examination was normal.

Complementary Studies

Routine blood tests of the patient were normal. Chest X-ray showed a band-like condensation at the base of the left hemithorax (Fig. 1A).

Thoracic computed tomography (CT) revealed a slightly increased pulmonary parenchymal density within the left lower lobe suggestive of pulmonary hyperperfusion and a frosted glass image suggestive of a chronic pneumopathy. CT scan imaging of the thorax and abdomen with arterial phase contrast (CT angiography) identified three tortuous arteries coming from the descending thoracic aorta and following a transverse course toward the abnormal pulmonary parenchyma (Fig. 1B).

Echocardiography revealed no evidence of heart disease or congenital malformations.

Multiplanar aortography was then performed (Fig. 1C), which confirmed the presence of two arteries measuring 5 mm in diameter and a smaller intermediate artery, all of them arising from the thoracic aorta immediately above the diaphragm, of transverse pathway toward the left hemithorax (nearly up to the visceral pleura), and supplying the abnormal lung area. In later images, pulmonary sequestration veins could be seen draining in the azygos vein.

Intervention

Endovascular treatment was performed by occluding inflow arteries to induce necrosis in abnormal tissue. Under systemic heparinization, a 7-F Raabe sheath (Cook Medical, Bloomington, IN) was advanced up to the level of the abdominal aorta. Selective catheterization of the upper and lower nutrient arteries was performed, advancing the tip of the sheath into the first centimeter from their origin. Then, a 10-mm Amplatzer Vascular Plug II (AGA Medical, Plymouth, MN) was deployed within the proximal one-third of both nutrient arteries. Postoperative angiography confirmed proper deployment, with occlusion of lower and upper arteries and absence of distal flow. Selective catheterization of smaller intermediate branch was subsequently performed for embolization with two 3-mm Nester coils (Cook Medical, Bloomington, IN) (Fig. 2A). Final angiogram showed occlusion of aberrant arteries (Fig. 2B). No incidents or complications occurred during the procedure.

Postoperative Evolution

In the following days, the patient reported moderate left chest pain treated with celecoxib (200 mg) every 12 hours for 2 days. CT angiography was performed before hospital discharge, which revealed complete

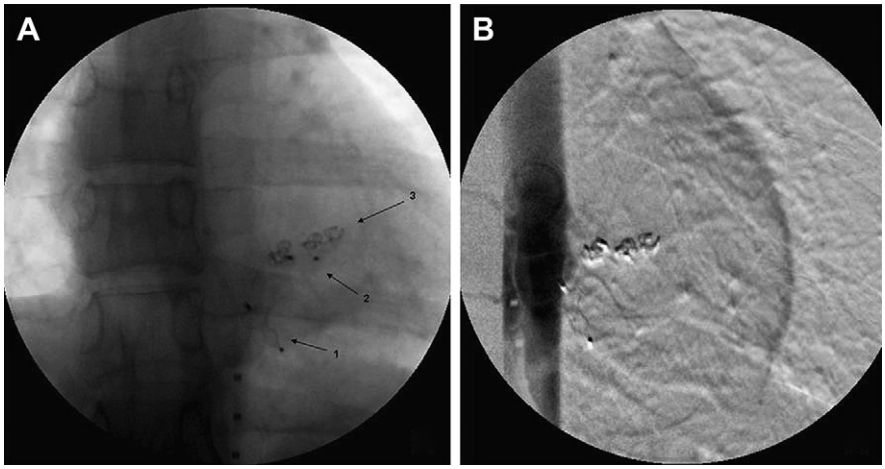

Fig. 2. Postembolization control. (**A**) Fluoroscopy image identifying the devices used during the embolization procedure: *arrows 1* and *2* point to the location of the Amplatzer occluders in the lower and upper branches, respectively, whereas *arrow 3* indicates the location of the coils used in the intermediate branches. Postoperative arteriography (**B**) shows absence of contrast in nutrient arteries.

infarction of the sequestration. Finally, the patient was discharged 5 days after endovascular procedure.

Subsequent Follow-up

At 6 months follow-up, the patient presented new upper respiratory tract infection and respiratory distress without hemoptysis. CT angiogram revealed persistent inflow from a smaller intermediate branch. Therefore, it was decided to perform a new angiography (Fig. 3A) and an additional embolization with three 6-mm Nester coils (Fig. 3B). The patient course was uneventful and she was discharged after confirming branch occlusion in a final CT scan (Fig. 3C).

A total of 41 months after the last intervention, the patient is asymptomatic and follow-up CT angiogram shows involution of the sequestration.

DISCUSSION

Pulmonary sequestration presents differences in its clinical and morphological characteristics, identifying two types according to their relationship with the visceral pleura. Intralobar sequestration, the most frequent, corresponds to abnormal bronchopulmonary tissue contained within the visceral pleura, unlike extralobar sequestration in which the parenchyma is outside of the visceral pleura and can be invested by its own external layer. Both types are clinically different (Table I), but final diagnosis is based on their appearance at the histological examination. In the present case report, the late age of onset, absence of feeding difficulties or a history of associated diseases in childhood or adolescence, the absence of other congenital anomalies, and the presentation with hemoptysis suggested an intralobar sequestration,[1,2] otherwise known to be the most frequently reported type.[2,3] The fact that the patient had been treated with endovascular procedure, without resection, precluded availability of a specimen for histological examination, and did not allow to analyze its relationship with the visceral pleura.

Pulmonary sequestrations do not communicate with the healthy bronchial tree. Clinical presentation with hemoptysis would be explained by recurrent infectious processes that facilitate connections between the sequestration and the normal pulmonary parenchyma[4,5] and the subsequent perfusion of the sequestration at systemic pressure.[6,7]

Conversely, diagnostic imaging studies and its subsequent follow-up have allowed for us to properly define relevant parameters associated with this pathology, regardless of subtype.

Association with other anomalies of embryonic development, such as diaphragmatic hernias, congenital cardiopathies, or cystic adenomatoid malformations, as well as tumoral pathologies, were properly ruled out.[2,8] Angiographic findings confirmed the status of arterial and venous flow, ruling out the presence of other nutrient arteries from the abdomen, as well as fistulas or other arteriovenous anomalies. Selective angiography should be performed in all patients with pulmonary sequestration, especially considering that this abnormal

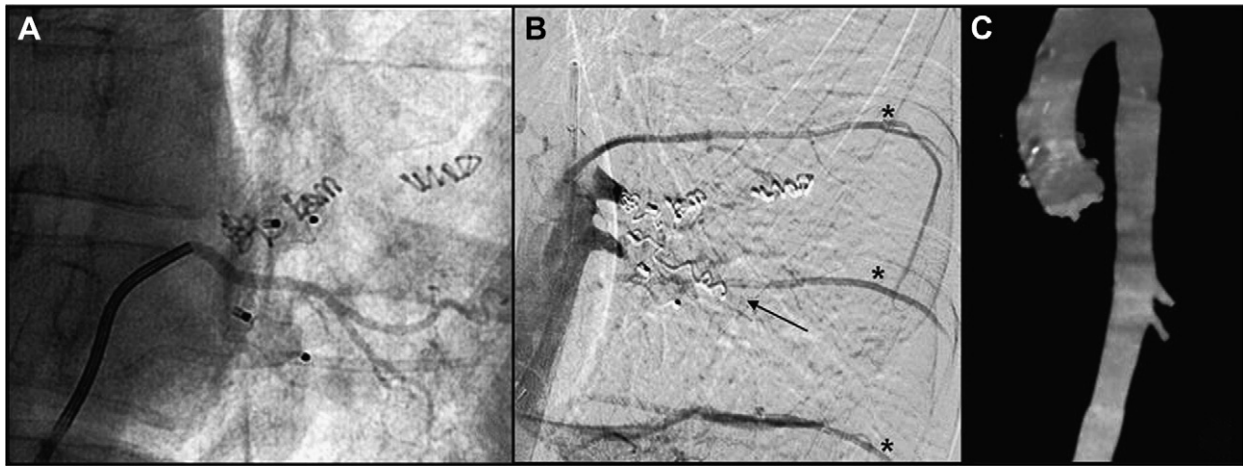

Fig. 3. Second procedure. Selective arteriography (**A**) contrasts previously unappreciated intermediate branch, which is treated with a new coil embolization indicated by the *black arrow* (**B**) *asterisks* indicate normal intercostal arteries), with proper angiographic result and confirmed by postoperative computed tomography angiogram (**C**).

Table I. Comparison of intra/extrapulmonary sequestration

Pulmonary sequestration	Extralobar	Intralobar
Age of onset	Early: 61% in <6 months	Late: rare in <2 years, 20% at 20 years
Gender (male:female)	3-4:1	1:1
Location	Lower lobe: 63% Other: intra-abdominal, mediastinum	Lower lobe: 98%
Side	65% Left	55% Left
Associated anomalies	65%	11%
Symptoms	Respiratory distress Feeding difficulties Fetal hydrops Congestive cardiac failure	Recurrent infection—most frequent Pneumothorax, hemothorax Hemoptysis Congestive cardiac failure

lung area may be supplied by multiple arteries in about 15% of the cases.[9,10]

The standard treatment of pulmonary sequestration has been conventional surgery with resection of abnormal parenchyma through a posterolateral thoracotomy. However, this approach may be the source of additional morbidity as well as aesthetic sequelae. It is even more severe at early ages because it causes skeletal and muscle deformities such as asymmetry of the chest wall, scoliosis caused by rib fusion, or winged scapula. It has been attempted to reduce these collateral complications by performing small incisions[11] or by thoracoscopy,[12,13] although it is not always possible because of variations in its anatomical presentations.

The key point in the surgical approach corresponds to the identification and control of the aberrant arterial supply; therefore, if all the vessels involved are not properly controlled, an exsanguinating hemorrhage may occur even leading to the patient's death. An alternative way to control this situation is to perform selective embolization of the nutrient arteries to avoid recurrent or massive hemoptysis, or to reduce the risk of bleeding during open resection,[14] especially if there is associated heart failure.

Endovascular management has been proposed to avoid the aforementioned risk of morbidity. Yet, there are very few reported cases with this unique therapeutic modality in neonates,[15] infants,[16-18] or adults.[7,19-21] Treatment success in these cases was established by the absence of clinical recurrence. Conversely, because no resection of the pulmonary sequestration was performed, there is a possible risk

of subsequent pulmonary infections; therefore, it is important to wait for late follow-up results to definitely consolidate this therapeutic alternative.

Devices used in this procedure included the Amplatzer Vascular Plug II (AGA Medical), which is a nitinol wire mesh designed to occlude peripheral vessels. Its main advantage is to provide accuracy and control during its placement, allowing recapture after deployment is initiated, so that it can be eventually repositioned before its final release. This feature led us to choose this device among other embolization devices, especially considering its lower risk of accidental deployment in the aorta, as has been previously reported.[22,23]

The present case report describes the successful outcome of endovascular treatment in a patient with symptomatic pulmonary sequestration without clinical relapse at midterm follow-up. Exclusive endovascular treatment enables effective and minimally invasive management in selected cases of pulmonary sequestration.

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