Surgical Intervention for Giant Pulmonary Artery Aneurysm in Behçet Disease: A Case Report

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Patient: Female, 37-year-old
Final Diagnosis: Pulmonary aneurysm
Symptoms: Hemoptisis
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease
Background: Pulmonary artery aneurysm (PAA), defined as a pathologic dilatation of the PA greater than 1.5-fold the normal diameter, is a rare complication of Behçet disease. It is due to a weakening of the vessel wall for a great vessels' vasculitis, often asymptomatic and incidentally diagnosed on imaging studies. However, if ignored, it can lead to life-threatening complications such as rupture and massive hemoptysis. We report the case of a giant fast-growing PAA in a young patient with a history of Behçet disease in which an inadequate follow-up and poor patient information could had led to life-threatening complications.

Case Report: A 37-year-old man with a history of Behçet disease presented to our Emergency Department with hemoptysis due to a right inferior lobar artery aneurysm measuring 52×33 mm. The aneurysm was detected years before, measuring 18 mm, but the patient and physicians missed the subsequent follow-up. After several attempts at embolization, the multidisciplinary board suggested to proceed with surgical intervention. Surgery was performed with an extracorporeal circulation system kept on stand-by due to the high hemorrhagic risk. By opening the fissure, the dilatation of the inferior lobar artery was clearly identified up to the origin of the middle lobar branch. Thus, a lower-middle bilobectomy was performed after the introduction of a suction cannula in the aneurysm, which facilitated its emptying and the subsequent maneuvers.

Conclusions: PAA is a rare disease, generally treated with medical therapy or interventional procedures. However, giant and fast-growing aneurysms are more likely to entail complications and often required immediate treatment. In this case, primary surgical intervention with a pulmonary bilobectomy appeared mandatory to avoid life-threatening events.

Keywords: Aneurysm • Behçet Syndrome • Thoracic Surgery

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Introduction

Pulmonary artery aneurysm (PAA) is a rare disease of the pulmonary circulation. It is defined as a pathologic dilatation of the PA to a diameter of at least 1.5-fold the normal diameter and involving all 3 layers of the vessel wall. Based on recent literature, main PA aneurysms in men are defined by a diameter larger than 43.4 mm and in females larger than 40.4 mm [1]. Acquired causes include infections, such as tuberculosis and syphilis, pulmonary arterial hypertension, chronic pulmonary embolism, lung cancer, medical procedures, and vasculitis, especially Behçet disease (BD) [2].

BD is an autoimmune multisystemic chronic inflammatory disease, first describe by Hulusi Behçet in 1924, generally involving the eyes, mouth, and genitals [3,4]. Large-vessel vasculitis affects only about 15% of Behçet’s patients, leading from thrombophlebitis to great artery aneurysms. The latter is due to a weakening of the arterial wall caused by disruption of the elastic lamina interna and is one of the leading causes of morbidity and mortality in these patients [3]. PAAas are rare, frequently asymptomatic, and incidentally diagnosed on imaging studies performed for other reasons. However, they can lead to life-threatening complications such as massive hemoptysis from rupture, coronary artery compression leading to acute coronary syndrome, and dissection [5]. In complicated and refractory cases, surgical intervention is required to prevent life-threatening events.

Case Report

A 37-year-old man presented to our Emergency Department reporting massive hemoptysis and dyspnea for a few hours. His medical history showed the diagnosis of BD in 2017 treated with colchicine and endovascular implantation of aortic prosthesis because of a giant abdominal aortic aneurysm. At that time, chest CT showed also a right pulmonary artery aneurysm of 1.8 cm diameter, and anticoagulant therapy with apixaban was started. The patient was not followed up.

At the Emergency Department, 5 years after the diagnosis of BD, the patient had a chest X-ray that showed a right parahilar opacity. Thus, a chest CT scan was performed, showing a right inferior lobar artery aneurysm measuring 52×33 mm with irregular walls, concentric thrombotic appositions, and ground-glass areas in the right lung (Figure 1). Anticoagulant therapy was stopped and the patient was initially treated with endovenous tranexamic acid. No anemia was found. After stabilization, he underwent PA angiography, which confirmed the inferior lobar artery aneurysm. Several attempts at embolization were unsuccessful. Considering the dimension of the aneurysm, the high risk of rupture, and the possible fatal complications, the multidisciplinary board suggested to proceed with surgical intervention according to the guidelines. In fact, another endovascular procedure was considered unfeasible and only providing medical therapy could have led to rupture and death.

Surgery was performed through a right posterolateral thoracotomy. An extracorporeal circulation system was kept on stand-by due to the high hemorrhagic risk. A voluminous hyper-vascularized and pulsatile mass appeared within the lung parenchyma. After opening the mediastinal pleura, the inferior pulmonary

![Figure 1](image-url)
vein was detected and prepared. By opening the fissure, the dilatation of the inferior lobar artery was clearly identified up to the origin of the middle lobar branch. Thus, a lower-middle bilobectomy was performed (Figure 2). The intermediate artery was carefully prepared, taking care of the ascending arterial branch for the superior lobe, and was sutured with a vascular Endo-Gia. The aneurysm was excluded from circulation and the introduction of a suction cannula facilitated its emptying and the subsequent manipulation maneuvers of the parenchyma. The pathology examination showed a thin wall of the pulmonary artery with rarefaction of the elastic fibers of the wall and the presence of an inflammatory infiltrate associated with fibrinoid necrosis.

The patient was discharged on postoperative day 6 without complications. One month after surgery, a chest CT scan control examination showed radiological stability.

**Discussion**

Vascular involvement in BD occurs in up to 30% of patients [3]. These are generally venous disorders such as thrombosis or venous occlusions. However, up to 20% of these patients have arterial lesions, especially aneurysm or pseudoaneurysm of the aorta, carotid, subclavian, brachial, renal, cerebral, and popliteal arteries. These lesions are due to the disruption of elastic fibers of the vessel wall from the related vasculitis. Arterial involvement is one of the main causes of mortality in patients with Behçet’s disease [3,4]. In particular, PAA is reported in less than 5% of cases, but these patients have a poor prognosis due to the high risk of rupture [2-6]. In fact, pulmonary circulation is characterized by low pressures and thinner arterial wall that facilitate the onset of rupture. Mortality reaches 50% when there are complications [7]. Hemoptysis occurs due to erosion of the inflamed arterial and bronchial walls and creation of a fistula between bronchi and pulmonary artery, which is often fatal. Therefore, prompt surgical intervention can prevent life-threatening events and it is indicated when there is disease evolution or after failure of other treatments [8]. Overall, the management strategies for PAA are variable and are based on the underlying etiology, hemodynamics, and associated comorbidities. There are still no general guidelines due to the limited experience and the rarity of the disease. Conservative treatment options include medical management aimed at the underlying disease. In BD, the first treatment options include corticosteroids associated with immunosuppressors (eg, azathioprine, cyclophosphamide, or anti-TNF). However, when these treatments fail, invasive treatment must be considered, including endovascular repair, surgical repair, or PA replacement with grafts.

Patients with larger aneurysms (>3 cm) are more likely to die [9]. In these patients, especially when the PAA is >5.5 cm, PAA embolization is frequently unsuccessful and surgical treatment is indicated. Surgical treatment of distal PAA may be more difficult, often requiring lung resections. Reisenauer et al suggested a therapeutic management based on the PAA dimension: PAAAs greater than 8 cm should undergo primary surgical intervention independently; PAAAs of 5-8 cm should be surgically treated if fast growing or symptomatic; and PAAAs smaller than 5 cm are candidates for conservative treatment and radiological follow-up [9]. In the latter, regular follow-up should be scheduled and treatment changes are recommended in case of disease worsening such as thrombus formation, compression of adjacent structures, growing greater than 5 mm in 6 months, hemoptysis, and onset of pulmonary artery hypertension [10].

We report this clinical case to show the role of surgery in treatment of pulmonary artery aneurysms. In our opinion, this case...
shows some inappropriate medical behavior. After the first finding of PAA, despite the absence of symptoms, the patient should have started close follow-up to monitor the effects of the medical therapy. However, the next radiological follow-up was missed. After 5 years, the aneurysm presented clinically with hemoptysis. The patient risked death by delaying coming to the hospital for 5 days. Fortunately, the hemoptysis was self-limiting.

At admission, a chest CT scan showed massive enlargement of the PAA, growing from 18 mm to 52 mm. Aneurysms of this size require urgent surgery since other interventional treatments often fail. However, the patient was exposed to the risks of PAA rupture due to an unsuccessful angiography procedure [8]. Surgery in this case required a middle-lower bi-lobectomy because the PAA involved the intermediate artery up to the origin of the middle lobar branch. However, lobectomies or segmentectomies can be feasible in presence of smaller and more peripheral aneurysms and should be preferred if possible due to the lower incidence of morbidity and mortality. The goal of surgical treatment in case of a giant PAA in a patient with a systemic disease such as BD is to eliminate the risk of acute rupture but not to cure the underlying disease. For this reason, medical treatment with immunosuppressor and radiological follow-up is essential for the disease control.

Conclusions

Giant PAA in patients with BD can lead to fatal complications, especially if fast growing, and may require primary surgical intervention. Here, we described a difficult case that involved some critical managerial and surgical mistakes. Lessons learned from this case include:

1. In case of symptomatic fast-growing aneurysm or refractory cases, surgery is the best treatment option to reduce the risk of rupture.
2. An extracorporeal circulation system in the operating room must be ready to manage any risk of PAA rupture.
3. Posterolateral thoracotomy to the fifth intercostal space allows for greater control of all elements of the mediastinum and is recommended.
4. Before starting PAA preparation, the main PA should be prepared and encircled to be ready in case of bleeding.
5. After adequate preparation, it is possible to exclude the vascularization of the aneurysm and to obtain a volume reduction by introducing an aspirating cannula into it, facilitating further resection maneuvers.
6. Involvement of a proximal part of the PA may require a more extensive resection such as a lower-middle bilobectomy or pneumonectomy.

Surgery is the best strategy in case of a giant PAA or complicated cases. Correct multidisciplinary management is necessary to determine the optimal timing of surgery and to avoid undertaking surgery only in emergency situations.

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