

# Complete Resolution of Bilateral Pulmonary Artery Embolism and Aneurysmal Dilatation with Behcet's Disease after Intravenous Cyclophosphamide Therapy Only: A Case Report and Review of the Literature

Ahmed A. Abd Alkader<sup>1</sup>, Ahmed K. Awad<sup>2</sup>, and Sheref A. Elseidy<sup>2</sup>

<sup>1</sup>Department of Rheumatology, Ain Shams University, Egypt

<sup>2</sup>Department of Cardiovascular Diseases, Ain Shams University, Egypt

## Abstract

Behcet syndrome is a multisystemic illness that leads to vasculitis. The classic constellation of symptoms is recurrent aphthous ulcers of the mouth and genitalia, hyperreactivity of the skin to penetrating trauma (pathergy reaction), other skin lesions, arthritis, and panuveitis. Vascular involvement can involve arteries and veins of all sizes and types. however venous involvement tends to be more common than arterial vasculitis mostly in the form of lower extremity vein thrombosis. Interestingly the disease manifestation of vasculitis itself seems to favor males than females and with a unique geographical distribution. Arterial vasculitis tends to affect the pulmonary vasculature, though Pulmonary involvement in Behcet's disease is rare, occurring 1-7.7 of patients. Pulmonary artery aneurysms with or without "in situ" thrombosis is the most common form of arterial involvement and are the leading causes of increased mortality.

Different management modalities have been proposed. of these immunosuppressive therapies have shown to decrease the inflammation and lead to resolution of the aneurysmal dilatation itself. however, this treatment is reserved for patients with minor to moderate hemoptysis and who are vitally stable.

In this case, we report a patient with Behcet induced pulmonary artery aneurysm and *in situ* thrombosis who presented with massive hemoptysis and was managed with only immunosuppressive therapy after initial stabilization that leads to complete resolution of the aneurysm.

**Keywords:** Behcet • Pulmonary artery aneurysm • Hemoptysis • Immunosuppressive treatment

## Introduction

Behcet is an Immune-mediated multisystemic disease that leads to vasculitis. Pulmonary involvement can range from parenchymal involvement to life-threatening pulmonary arterial vasculitis mostly in the form of aneurysmal dilatation with or without thrombosis that can lead to life-threatening hemoptysis and is considered the leading cause of mortality in those patients if not promptly diagnosed and managed. Management is tailored according to the general condition with which the patient present. Medical management is reserved for patients with non-life treating hemoptysis and good general condition.

## Case Report

A 27 Y/O male presented with a severe attack of hemoptysis of 700 ml since admission and estimated 100 ml prior.

G/A; the patient was pale and in distress

Vitals; Bl/pr.; 100/60, pulse: 110 Bpm

Chest X-ray revealed widened mediastinum, but no patches of pneumonia and no signs of congestion. CT chest (Figure 1) revealed bilateral fusiform aneurysmal dilatation of the main pulmonary arteries with partial mural thrombosis.

**\*Address for Correspondence:** Ahmed K. Awad, Department of Cardiovascular Diseases, Ain Shams University, Egypt, Tel: +0201067071734, E-mail: ahmedkawad@gmail.com

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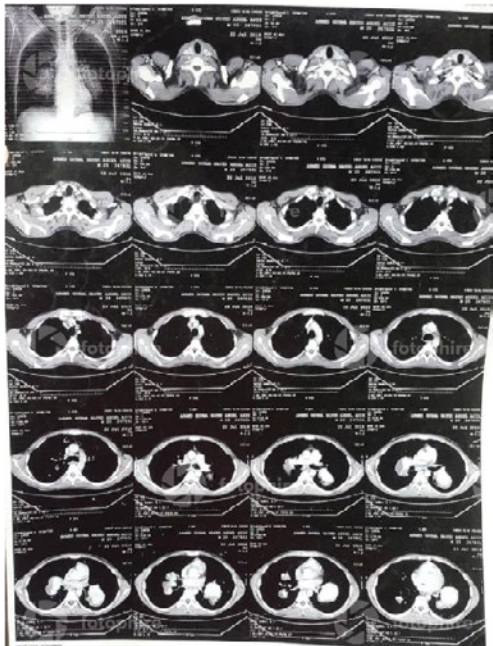


**Figure 1.** Chest CT shows widened mediastinum, but no patches of pneumonia and no signs of congestion with bilateral fusiform aneurysmal dilatation of the main pulmonary arteries with partial mural thrombosis.

On further history taking, the patient told us that he has experienced recurrent oral and genital ulcers with bilateral ankle arthritis. The patient was diagnosed with Behçet's disease.

Resuscitation with 2 units PRBCs and IV crystalloids was done through two large-bore cannulas. Afterward, we started treatment with IV methylprednisolone 1000 mg for 3 successive days, followed by six cycles of IV cyclophosphamide for 6 successive months. 3 months later, there was no improvement and the patient still having scant hemoptysis.

Follow up CT (Figure 2 ) revealed no improvement with the stationary course as regarding the size of aneurysmal dilatation of pulmonary arteries, but progressive and extensive propagating mural thrombosis more on the left side. We continued our management plan with close monitoring. Later, 4 months after the last dose (one year after diagnosis), CT chest (Figure 3) revealed that there was a complete resolution of the pulmonary artery embolism and aneurysm. The patient's condition dramatically improved and with no hemoptysis.



**Figure 2.** Follow up CT reveals no improvement with the stationary as regarding the size of aneurysmal dilatation of pulmonary arteries, but progressive and extensive propagating mural thrombosis more on the left side.



**Figure 3.** Chest CT shows complete resolution of the pulmonary artery embolism and aneurysm 4 months after the last dose of IV cyclophosphamide and one year after diagnosis.

During the disease, the patient had severe bilateral ankle arthritis that improved only with intraarticular steroid injection, but a few months later arthritis recurred. the patient is maintained on oral steroids, parenteral methotrexate, oral cyclosporine and colchicine, and the condition is stable at the time being and the patient has resumed his normal quality of life after being disabled for one year.

## Discussion

Behcet disease is one of the multisystemic affecting types of immune-mediated vasculitis. the disease was found to be HLAB51 (major Histocompatibility Complex, class B) mediated and tends to affect males more than females and with a geographical distribution with rates higher in people with Turkish or north Mediterranean descent [1].

Vascular involvement interestingly is more common and more severe among males and has a predilection of venous than arterial involvement with a significant tendency for thrombosis, and an unfortunately relapsing course. Lower extremity vein thrombosis is the most frequently reported, followed by inferior vena cava thrombosis.

Pulmonary involvement in Behçet's disease is rare, occurring 1-7.7 of patients [2]. And range from recurrent pneumonia, bronchiolitis obliterans and pleurisy to Pulmonary artery aneurysms with or without "in situ" thrombosis. Pulmonary artery aneurysm is reported in 1.5% of adults with Behçet's disease [3,4].

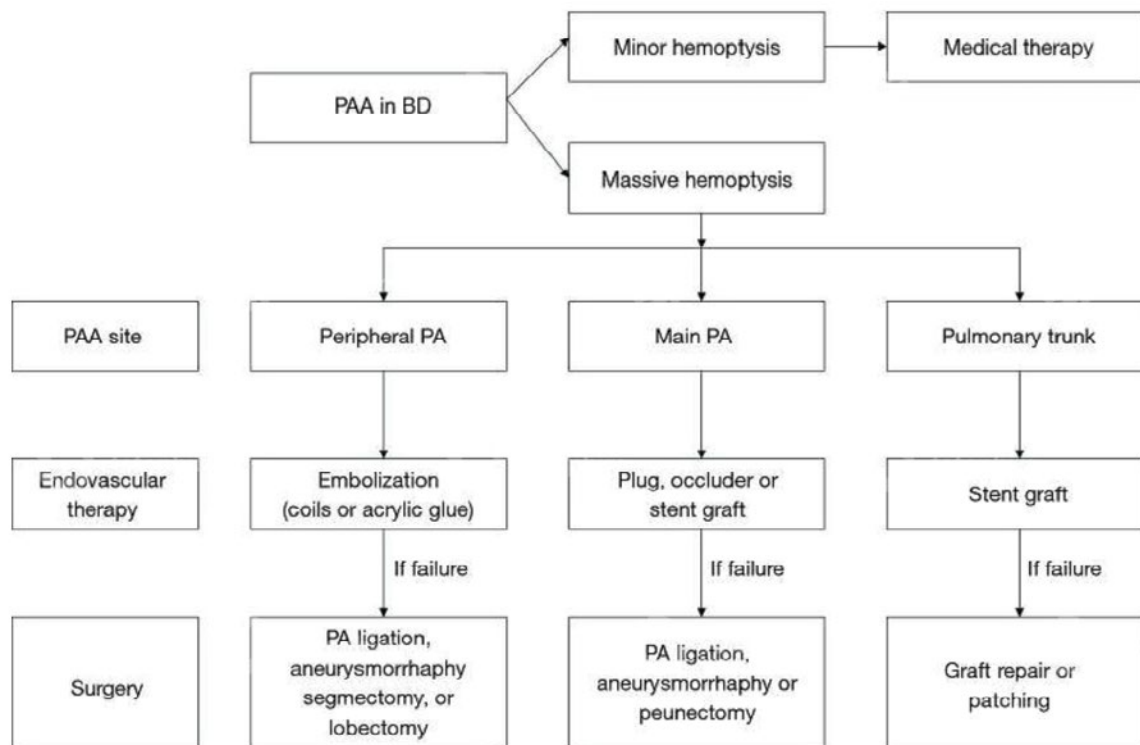
PAI is the leading cause of increased mortality Pulmonary artery aneurysms are the leading cause of mortality in these patients mostly owed to their high tendency to rupture and lead to massive hemoptysis [1-3].

The underlying pathophysiology of aneurysmal formation in Behcet is inflammation of the vasa vasorum of the tunica media, the inflammation and infiltration by lymphocytes, plasma cells, and neutrophils will eventually lead to cytokine-mediated degradation of the elastic fibers of the media and subsequent dilatation of the vessel lumen [5]. Thrombosis of the pulmonary arteries in Behcet's disease is usually *in situ* not a migrating embolus from a lower limb DVT (Deep Venous Thrombosis) as thrombi of the inflamed veins are usually strongly adherent [6].

Hemoptysis is the most common and the most feared presentation of a ruptured aneurysm. Major bleeding is often associated with a poor prognosis with less than 50% 2-year survival, requiring early prompt diagnosis and treatment [7].

Less aggressive manifestations include chest pain, dyspnea, and fever. Our patient was presented with chest pain, intermittent hemoptysis, oral, and genital ulcers [1].

Different treatment modalities, such as surgery, embolization, and various immunosuppressive medications have been used for the management of pulmonary artery aneurysms with variable results [8,9]. However, no randomized controlled study has assessed the efficacy of one plan over the other [10], a strategy was formulated to tailor each modality according to the degree of hemoptysis and the presenting clinical condition with surgery and Endovascular approaches for moderate and severe hemoptysis and immunosuppressive treatment after stabilization for scant and frank hemoptysis (Figure 4). That been said no golden rule for management has been placed and it remained controversial. many cases have been reported with regression of pulmonary aneurysms after treatment with corticosteroids and immunosuppressive agents alone without stating the correlation to the degree of hemoptysis [6,11].



**Figure 4.** Management protocol of PAA in BD to tailor each modality according to the degree of hemoptysis and the presenting clinical condition with surgery and endovascular approaches.

The European League Against Rheumatism currently recommends the utilization of immunosuppressive agents as corticosteroids, cyclophosphamide, and azathioprine for treating major vessel diseases. Anticoagulant therapy is not recommended for the treatment of a Behçet associated DVT (Deep Venous Thrombosis) and/or pulmonary artery thrombosis due to the increased risk of fatal hemoptysis in these cases. Surgery is also discouraged unless performed in an emergency setting, due to the high risk of complications and recurrence [7]. Though immunosuppressive treatment is believed to decrease the inflammation, even the vessel wall [12,13], the exact mechanism of this regression is unknown [13] there is also a chance that the aneurysms may completely resolve with medical therapy alone if no deterioration in the patient's clinical status [13]. the question remains should a patient with severe hemoptysis given a chance of medical management with immunosuppressive therapy if his condition can be initially stabilized remains unanswered. and answering this question would be of great benefit to refrain from operative management strategies that would further endanger a patient with this condition. in our case, the chance of medical management was completed guided by no deterioration in his clinical status and even without initial rapid signs of improvement immunosuppressive treatment has continued that lead to complete resolution of the patient's aneurysm and paved the path for a full recovery till the time being.

## Conclusion

Behçet disease Pulmonary artery aneurysm with or without thrombosis should be given a chance for immunosuppressive therapy alone regardless of the presentation in terms of the degree of hemoptysis guided by no deterioration of clinical status occurs.

## Declaration

### Ethics approval and consent to participate

The case report was performed under the declaration of Helsinki and approved by the Ain-Shams University Hospital ethics committee-Ain Shams institute-according to the international guidelines and ethics.

### Consent to publish

Written consent was taken from the patient's parents.

### Conflict of interests

All authors declared no conflict of interest.

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### Authors' contributions

All the authors have shared work in collecting, analyzing, and writing the research paper. Dr. Ahmed A Abd Alkader, Dr. Sheref A Elseidy were major contributors in writing the manuscript. All authors read and approved the final manuscript.

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Not applicable.

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