






CASE REPORT

Cavitary lung opacity of unusual cause during Behçet disease

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Abstract

Pulmonary artery aneurysm must be evoked in front of any hemoptysis in a patient with Behçet disease as it requires urgent immunosuppressive therapy and often surgery.

KEYWORDS

Behçet disease, cavitary lung opacity, corticosteroids, cyclophosphamide, intracardiac thrombosis, pulmonary artery aneurysm

1 | INTRODUCTION

Cavitary lung opacities are commonly detected on chest imaging, often posing diagnosis challenges to the clinician. When associated with hemoptysis, infectious causes, notably tuberculosis, have to be eliminated—particularly in the Tunisian context. Noninfectious causes of cavitary lesions are less common, including malignancies, rheumatologic diseases, and rarely pulmonary aneurysms.¹

Behçet's Disease (BD) is a multisystemic vasculitis affecting both small and large vessels. Vascular involvement is reported in up to 40% of the cases. The veins are mostly affected, resulting in subcutaneous and deep thrombosis with a relapsing course. However, arterial involvement is less frequent, manifesting as thrombosis, stenosis, and/or aneurysms that are variously associated.^{2,3}

We report an unusual case of BD with pulmonary artery aneurysm (PAA) presented with a cavitary lung mass and hemoptysis.

2 | CASE REPORT

We report the case of a 24-year-old, non-smoking Tunisian man who presented in August 2020 with a one-week history of mild hemoptysis. He had already been followed for 10 years (since 2010) in the department of internal medicine for a BD with severe cardiovascular damage. He has had many relapses since the initial onset of the disease. The first was at the time of diagnosis, when the patient presented recurrent oral and genital aphthous ulcers with a right posterior basal pulmonary embolism, leading to the diagnosis of BD. The ophthalmological examination was normal, and the pathergy test was negative. He received curative anticoagulation for 1 year, with colchicine 1 mg/day, with a good outcome. The second relapse was about 8 years after diagnosis, in 2018. He presented a dry cough with mild hemoptysis. The chest CT angiography showed a bilateral pulmonary embolism and an aneurysm of a distal segmental pulmonary artery branch of the left lower lobe measuring 2 cm in diameter. Three boluses of methyl-prednisone 1 g/day were initiated and

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followed by prednisone 1 mg/kg/day, combined with azathioprine 150 mg/day. Curative anticoagulation was also prescribed, and the colchicine was maintained. The patient was clinically improved, and angiographic images after 6 weeks showed regression of PAA from 2 to 1 cm. However, 2 months later, hemoptysis recurred, accompanied by fever and cough. The CT angiography of the thorax showed multiple bilateral thrombosed PAAs (the right posterior basal, the left lower lobe, and the left paracardiac arteries) and an intracardiac thrombosis of the right ventricle (24 × 15 mm). The latter was confirmed by a cardiac MRI and a transthoracic ultrasound, which also showed pulmonary hypertension estimated at 45 mmHg. Monthly intravenous cyclophosphamide pulses combined with a high dose of corticosteroid therapy (three boluses of methyl-prednisone followed by prednisolone 1 mg/kg/day) were initiated. The azathioprine was stopped. Two months later, the cardiac thrombus became organized, and the PAA size decreased. The patient received 12 monthly and then quarterly pulses of cyclophosphamide. Right at the time of the second quarterly pulse, in August 2020, he developed recurrent mild hemoptysis that had been progressing for nearly 1 week. Clinically, he was afebrile. He had no oral or genital aphthous ulcers. He was eupneic with a correct saturation. His blood pressure was 110/80 mmHg. There was not any other external bleeding. Biology showed a normal blood count, with an international normalized ratio (INR) of 3.4. There was no biological inflammatory syndrome. Chest X-ray showed a 4 cm diameter left basal cavitory lesion with an air-fluid level (Figure 1A). The CT angiography of the thorax showed a regularly shaped, thin-walled cavity with an air-fluid level, connecting with a segmental bronchus of the caudal segment of the lingula, measuring 40 × 44 mm, (Figures 1B and 2). The anti-vitamin K treatment was switched to low-molecular-weight heparin at a curative dose. An infectious complication was feared, but diagnostic investigations for tuberculosis were all negative. Cytobacteriological examination of the sputum, hydatid serology, and aspergillus antigenemia was also negative. However, we prescribed antibiotics: amoxicillin, clavulanic acid, and metronidazole. Two days later, the patient presented massive hemoptysis. Rest was prescribed along with symptomatic treatment based on nebulized adrenaline and injectable etamsylate and simvastatin. Besides, the anticoagulation was stopped. He was then transferred to the cardiovascular surgery department. The patient underwent posterolateral thoracotomy for lingulectomy. Corticosteroid therapy was increased 2 days before the procedure at a dose of 0.5 mg/kg/day of prednisone. The appearance of an aneurysmal sac containing blood and a thrombus and connecting with bronchus via fistula was described intraoperatively (Figure 3). The patient's postoperative course

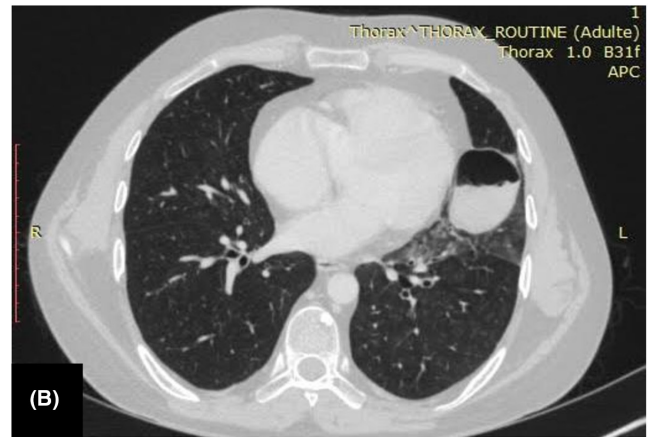
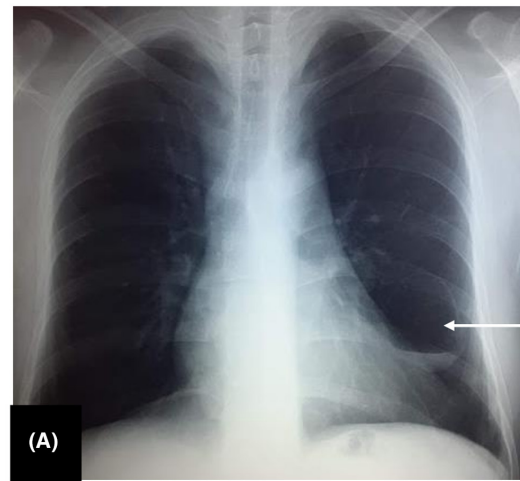


FIGURE 1 (A) Chest radiograph showing a rounded left basal opacity with an air-fluid level (arrow); (B) Chest CT angiography showing a 40 mm × 44 mm left cavity with an air-fluid level

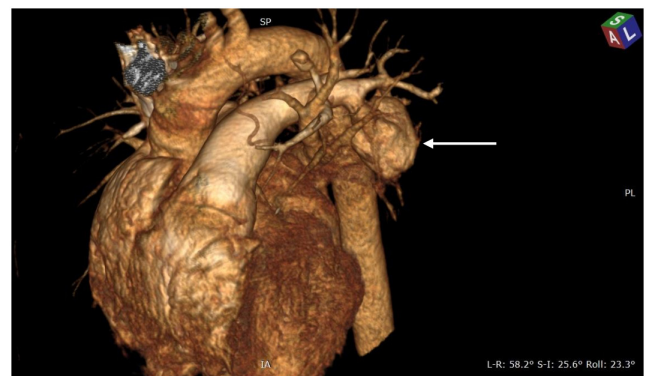


FIGURE 2 Three-dimensional construction of helical CT showing the left pulmonary aneurysm (arrow)

was good. He received a total of 14 days of antibiotics. Anticoagulation has been reintroduced at a curative dose. Hemoptysis did not recur during a 6-month follow-up. An early postoperative X-ray showed no cavitory lesions or operative complications.

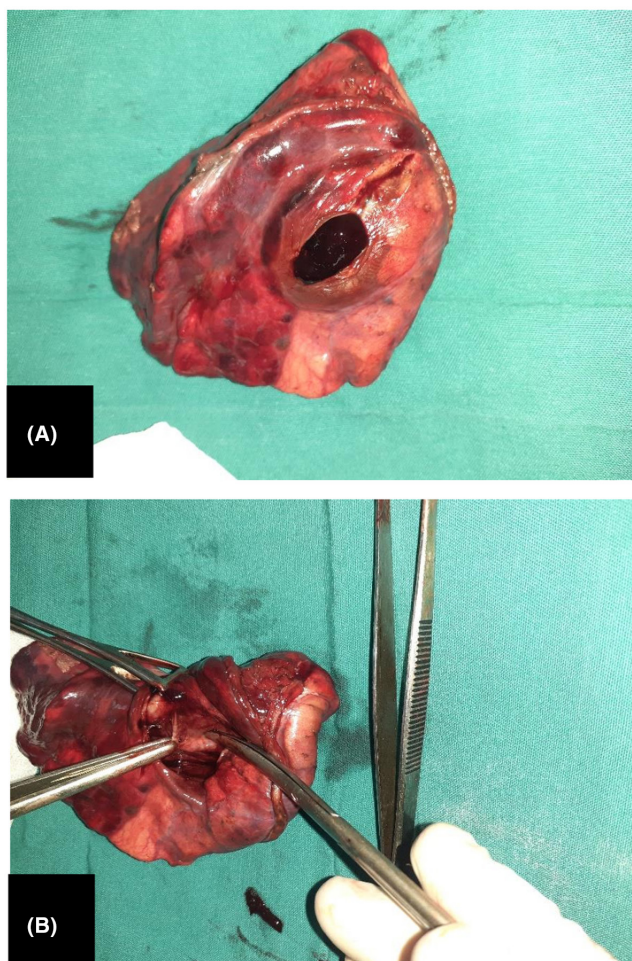


FIGURE 3 Lingulectomy piece: (A) an aneurysmal sac containing blood and a thrombus (arrow); (B) aneurysmal sac connecting with bronchus via fistula (arrow)

3 | DISCUSSION

Behçet's disease is a systemic vasculitis of unknown etiology, with a male preponderance in the Mediterranean region. It is characterized by recurrent oral and genital ulcers, in addition to ocular, cutaneous, and vascular manifestations.⁴ Our patient had confirmed BD according to the revised international criteria⁵ (oral and genital aphthosis with vascular lesions; score = 5).

Vascular involvement concerns up to 40% of the cases, with a predilection for veins. Deep vein thrombosis of the lower extremities is the most frequent vein damage. However, pulmonary artery involvement is rarely observed, with a prevalence rate of less than 5%.⁶ Yet, it is the most common form of arterial involvement. It usually appears within 3–4 years after disease onset. PAA is the most typical form of pulmonary artery involvement and also the most common pulmonary manifestation in BD. Pulmonary artery thrombosis is a less frequent form of pulmonary artery

involvement, observed in about 1/3 of the patients. This prevalence is increasing with the advancing imaging techniques. It is also worth noting that PAA and pulmonary artery thrombosis may coexist and that aneurysms in the BD may be partially or totally thrombosed in 85% of the cases.^{2,7} It is the case of our patient who presented thrombosed PAA.

Clinically, any hemoptysis in a patient with BD should alert the clinician to look for pulmonary artery involvement.⁸ It is, in fact, the most typical and common sign, present in 80% of the cases of pulmonary artery involvement. It can be massive and life-threatening in one-third of the cases, especially in the presence of PAA. Many other non-specific symptoms can be observed according to the severity and the extent of the damage, such as cough, chest pain, dyspnea, and fever. Pulmonary artery involvement manifested in our patient by recurrent hemoptysis in all the relapses, and cough and fever in one relapse.

The characterization of pulmonary lesions is better with thorax computed tomography angiography than with chest X-ray. Therefore, it is the first imaging modality to use in case of suspicion of pulmonary artery involvement. PAAs are usually multiple and bilateral, with a predilection to branches of the lower lobes. This characteristic was found in our case. Other pulmonary lesions are often discovered in the thorax CT scan, such as nodules, consolidations, cavities, and ground glass lesions. They are considered as a part of the pulmonary artery involvement spectrum. They also predominate in the basal lobes, and they are more noted in the active stages of pulmonary artery involvement.^{2,9}

According to the European League Against Rheumatism (EULAR) guidelines for the management of BD, updated in 2018, PAA treatment is based on high-dose glucocorticoids and cyclophosphamide. In refractory cases, monoclonal anti-TNF antibodies should be considered. When the patient has or is at high risk of major bleeding, embolization should be preferred to open surgery (Level of evidence: III; strength of recommendation: C).¹⁰ Surgical lobectomy remains an option in case of massive hemoptysis due to PAA and requiring a definitive solution, especially when the aneurysm is giant.^{2,11} We add that preoperative steroid coverage has been suggested to reduce the risk of complications.¹² In our case, the medical treatment alone with steroids and immunosuppressive drugs (azathioprine then cyclophosphamide) failed in the control of the PAA. Given the episode of massive hemoptysis, an urgent surgical treatment was indicated.

Despite aggressive treatment, pulmonary artery involvement is associated with high mortality reaching 25% of the cases.¹¹ The major factors associated with a poor prognosis in PAA are large aneurysms (diameter >3 cm) and high pulmonary artery pressures. Aneurysms of the pulmonary artery can be complicated by often fatal rupture, or by fistulas of the bronchopulmonary arteries. The

rupture of the pulmonary aneurysms is the main cause of death in BD.

Surviving patients may be asymptomatic or have some symptoms such as dyspnea or hemoptysis.¹³

Our patient presented factors of poor prognosis. In fact, the PAA reached 4 cm in diameter. In addition, he presented pulmonary hypertension. He also developed two complications: aneurysmal thrombosis and broncho-arterial fistula. Fortunately, the postoperative course was satisfactory.

4 | CONCLUSION

A cavitary lung mass with hemoptysis during BD should suggest a PAA. This arterial manifestation deserves to be well recognized because of its severity and its high risk of morbidity and mortality. When diagnosis and management are delayed, PAA can be complicated by in-situ thrombosis, often fatal rupture, or by fistulas of the bronchopulmonary arteries. The prognosis has improved since the use of immunosuppressive drugs. However, embolization or vascular surgery often becomes inevitable in aneurysmal lesions. In our patient, the solution was medico-surgical.

AUTHOR CONTRIBUTIONS

DC involved in data collection, research, and manuscript writing. FF, MS, RBS, and SM contributed to data collection and involvement in the correction/revision of the manuscript. CD and ZB involved in data collection, research, and correction/revision of the manuscript. WA, AH, and MR involved in data collection and operation on the patient.

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CONFLICT OF INTEREST

None declared.

DATA AVAILABILITY STATEMENT

All data are available.

ETHICAL APPROVAL

Published with the consent of the patient.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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