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CASE REPORT

Pulmonary aneurysms associated with thrombosis in Behçet's disease: a case report

Aneurismas pulmonares associados a trombose na doença de Behçet: um relato de caso

Renato Lucindo Bolelli de Oliveira[1], Ruben Horst Duque[1], Isac Ribeiro Moulaz[1], Anna Carolina Simões Moulin[1], Rafael Burgomeister Lourenço[1], Bruna Costa da Mata Moura[1], Ketty Lysie Libardi Lira Machado[1].

[1] Hospital Universitário Cassiano Antonio Moraes, Universidade Federal do Espírito Santo, Vitória/ES, Brasil.

E-mail:
renatobolelli@hotmail.com

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ABSTRACT

Introduction: Behçet's disease is a chronic, multisystemic, inflammatory syndrome characterized by recurrent attacks of oral and genital ulcers, in addition to cutaneous, ophthalmic, central nervous system, gastrointestinal and vascular involvement. Other manifestations may occur more rarely, such as pulmonary artery aneurysm and pulmonary artery thrombosis. **Case report:** We report a case of a woman presenting with dyspnea and hemoptysis secondary to pulmonary artery aneurysms and pulmonary artery thrombosis caused by Behçet's disease as the first clinical manifestations. **Conclusion:** Anticoagulation in thrombosis, without the use of immunosuppressants in patients with Behçet's Disease, may lead to a poor outcome, as the underlying vasculitis will not be treated. The clinical suspicion of this condition has prognostic implications, especially in atypical presentations, emphasizing the relevance of the theme exemplified in this case report.

Keywords: Behçet's disease; Pulmonary artery aneurysm; pulmonary artery thrombosis.

RESUMO

Introdução: A doença de Behçet é uma síndrome inflamatória crônica multissistêmica, caracterizada por crises recorrentes de úlceras orais e genitais, além de envolvimento cutâneo, oftálmico, do sistema nervoso central, gastrointestinal e vascular. Outras manifestações podem ocorrer mais raramente, como aneurisma de artéria pulmonar e trombose em artéria pulmonar. **Relato de Caso:** Relatamos o caso de uma mulher que apresentou dispneia e hemoptise secundárias a aneurismas de artéria pulmonar e trombose na artéria pulmonar causado pela doença de Behçet como primeiras manifestações clínicas. **Conclusão:** A anticoagulação na trombose pulmonar, sem o uso de imunossupressores em pacientes com doença de Behçet, pode levar a um desfecho desfavorável, pois a vasculite subjacente não será tratada. A suspeita clínica desta condição tem implicações prognósticas, especialmente nas apresentações atípicas, enfatizando a relevância do tema exemplificado neste relato de caso.

Palavras-chave: Doença de Behçet; Aneurisma de artéria pulmonar; Trombose artéria pulmonar.

INTRODUCTION

Behçet's disease is a rare, chronic and multisystem inflammatory syndrome involving small, medium and large vessels of the arterial and venous circulation. It is characterized by recurrent oral and/or genital aphthous ulcers accompanied by skin, eye, joint, gastrointestinal and nervous system lesions, and thromboangiitis, thrombosis, arteritis and arterial aneurysms may also occur[1].

Vascular impairment in Behçet's disease is present in 25 to 30% of cases, and pulmonary artery aneurysm, although rare, is the most lethal complication of the disease, with greater severity in cases associated with pulmonary artery thrombosis[2].

There are rare cases reported in the literature of pulmonary aneurysms associated with pulmonary artery thrombosis as the first clinical manifestations of Behçet's disease. Our objective is to describe a case of a young woman with a rare and potentially serious initial form of the disease.

CASE REPORT

Female, Brazilian, 36 years old, presented with an unintentional weight loss of 26,45 lbs in six months, with fever for one week, dyspnea on moderate exercises and minor hemoptysis. She sought medical attention, was hospitalized, and during hospitalization developed oral and genital ulcers, in addition to pseudofolliculitis-type lesions on the trunk.

A computed tomography angiography of the chest showed occlusive filling defects involving the branches of the lower lobes of the pulmonary arteries, subsegmental branches from the posterior segment of the right upper lobe and the lingular segments of the left upper lobe, compatible with pulmonary artery thrombosis. Dilation of the respective arterial segments is associated upstream of the areas of occlusion, constituting aneurysms measuring approximately 1.5 x 1.5 cm (branch of the posterior segment of the right upper lobe), 1.7 x 1.9 cm (proximal portion right lower lobar artery), 1.2 x 1.4 cm (proximal lingular branch) and 1.7 x 1.6 cm (left lower lobar artery) (Figures 1, 2 and 3).

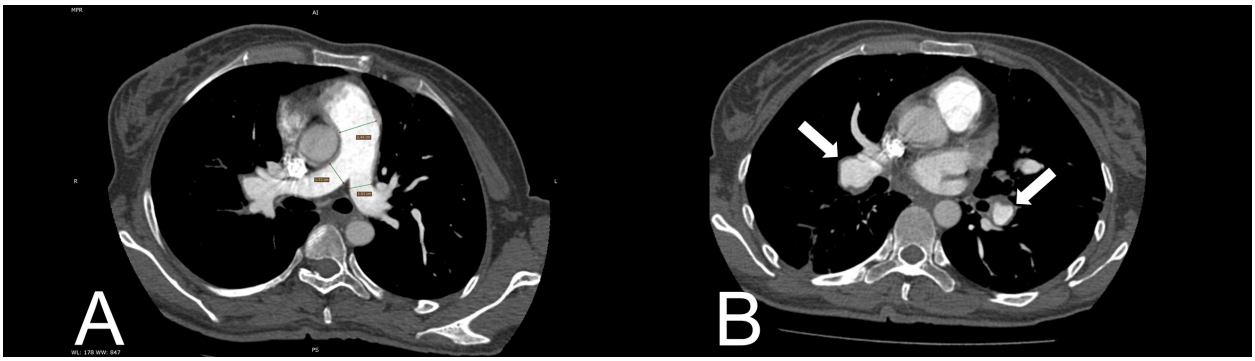
Considering the International Study Group (ISG) criteria and pulmonary artery aneurysm, the diagnosis of Behçet's disease was made, after excluding other inflammatory and infectious systemic conditions.

Pulse therapy with methylprednisolone and cyclophosphamide was promptly instituted due to the potential severity related to the case, and after the initial treatment, the patient evolved without further episodes of hemoptysis and with improvement in the general condition.

In a multidisciplinary decision, considering areas of thrombosis involvement and right heart overload, the use of rivaroxaban was associated with immunosuppressive treatment, with no adverse events.

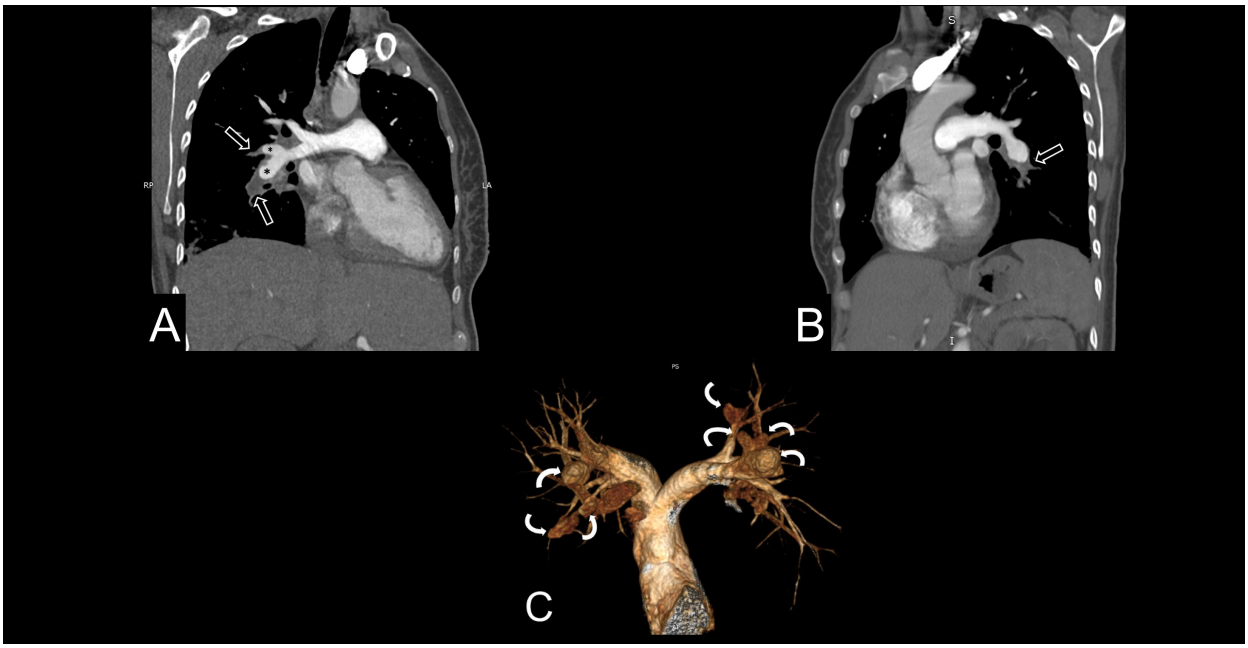
After 6 months, with complete resolution of symptoms and without clinical intercurrences, immunosuppression with azathioprine was maintained and rivaroxaban was discontinued, and new imaging tests and clinical control were scheduled.

FIGURE 1. CT Angiography



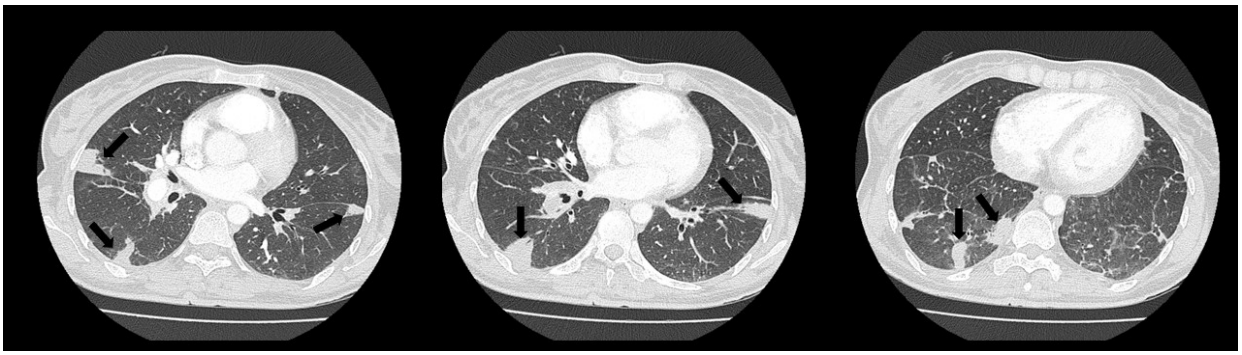
A. CT Angiography (oblique multiplanar reconstruction at the truncus pulmonalis): normal caliber of the trunk, right and left pulmonary arteries. B. CT Angiography (axial image): focal aneurysms of the right (19mm) and left (15mm) inferior lobe arteries (arrows). Source: Authors.

FIGURE 2. CT Angiography



A. CT Angiography (coronal oblique at the level of the right pulmonary artery): massive thrombi with complete filling defects of the medial lobe and inferior lobe arteries (open arrows) with proximal aneurysms (*). B. CT Angiography (coronal oblique at the level of the left pulmonary artery): massive thrombi with complete filling defect of the left inferior lobe artery (open arrow with proximal aneurysm (*). C. CT Angiography (volume rendering reconstruction of the pulmonary tree - posterior view): multiple filling defects and associated with proximal aneurysms (curved arrows). Source: Authors.

FIGURE 3. CT Angiography (lung window): peripheral cuneiform opacities consistent with pulmonary infarcts (black arrows)



Source: Authors.

DISCUSSION

The diagnosis of Behcet's disease is challenging, especially in rare and atypical manifestations. The case described started with symptoms of hemoptysis resulting from pulmonary aneurysms and pulmonary artery thrombosis, which forces us to carry out a careful diagnostic exercise, since there are no specific markers and this has therapeutic implications that can define the prognosis[4].

In one study, most patients (87%) with initial vascular compromise had deep vein thrombosis, while only 1 in 882 patients had pulmonary artery involvement[6].

Pulmonary manifestations occur rarely, but pulmonary artery aneurysm (PAA) is the most typical pulmonary complication of Behçet's disease, with hemoptysis being the most common symptom, followed by dyspnea. The most common location of the PAA is in the right lower lobe branch, as was found in our patient[3].

The presence of pulmonary artery aneurysms correlates with a worse prognosis, with an estimated 30% mortality within two years[5].

Our patient also had thrombosis as a concomitant clinical manifestation of the AAP. In Behçet's disease, thrombus development in the pulmonary artery occurs in the form of in situ thrombosis associated with endothelial injury[13]. Neutrophils and the inflammatory process are essential in the pathogenesis of thrombotic phenomena. Activation of neutrophils causes the formation of thrombi by oxidizing fibrinogen[8], and the cytokines IL-1, IL-6, IL-17, CXCL8 and TNF-alpha are also important in pathogenesis[9].

Regarding treatment, it is still a challenge, and there is no consensus regarding anticoagulation, since thrombosis in Behçet's disease presents, in its pathophysiology, a basic immune-mediated inflammatory process and, moreover, anticoagulation presents risks for hemorrhage, especially when there is concomitant pulmonary artery aneurysm[11]. In the therapeutic choice, we must take into account the severity of the hemoptysis, the sites and the number of AAP. Immunosuppressants should be the first-line treatment, causing regression of aneurysms and thrombi.

The etiopathogenesis in the formation of pulmonary artery aneurysms is still uncertain[8]. Several immunological alterations have been described in BD, such as involvement of free radicals, action of lysosomal enzymes, autoantibodies against endothelial cells and necrotizing vasculitis with lymphocytic infiltrate, mainly in the media and adventitia layers of vessels[8]. Histologically, the formation of aneurysms is characterized by rupture of the internal and external elastic lamina, with thinning of the intima, degeneration of the media layer and vasculitis of the vasa vasorum with lymphocytic perivascular infiltrate[9 10].

In the recommendations for the management of Behçet's disease by the European League Against Rheumatism (EULAR), in the case of pulmonary artery involvement, the primary management should be with immunosuppressants, in particular corticosteroids and cyclophosphamide as the first line, or with anti-TNF in refractory cases. For patients at or high risk of major bleeding, prefer embolization over open surgery[7].

For pulmonary and peripheral artery aneurysms, the choice of surgical intervention between graft insertion, ligation and revascularization surgery can be made according to the size and location of the aneurysm and the experience of the surgeon[7].

Treatment with anticoagulants for pulmonary thrombosis in Behcet's disease is still controversial due to the risk of worsening symptoms and side effects[12].

In addition to immunosuppressive treatment, we chose to associate rivaroxaban in our patient, due to thrombosis extension and possible hemodynamic repercussions on the right ventricle. Clinical follow-up monitoring adverse events was performed. There were no clinical complications during the period.

CONCLUSION

Anticoagulation in thrombosis, without the use of immunosuppressants in patients with Behçet's Disease, may lead to a poor outcome, as the underlying vasculitis will not be treated. The clinical suspicion of this condition has prognostic implications, especially in atypical presentations, emphasizing the relevance of the theme exemplified in this case report.

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DECLARATIONS

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Conflict of Interest

The authors declare no conflicts of interest.

Ethics Committee Approval

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Availability of Research Data and Other Materials

Research data and other materials can be obtained by contacting the authors.

Responsible Editors

Carolina Fiorin Anhoque, Blima Fux.

Correspondence Address

Ambulatório seis, Serviço de Reumatologia, HUCAM, Universidade Federal do Espírito Santo, Av. Marechal Campos, 1468, Maruípe, Vitória/ES, Brasil, CEP: 29043-900.