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CASE REPORT

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A case of Behçet's disease with unusual cardiovascular complications

Rova Malala Fandresena Randrianarisoa[1] | **Lalao Nomenjanahary Rakotonirina**[2] | **Mirantosoa Fabiola Ravelonjatovo**[3] | **Nofihinina Joharisantatra Andrianjanaka**[3] | **Nouraly Habib**[4] | **Narindrarimanana Avisoa Randriamihangy**[3]

[1]Department of Internal Medicine, Joseph Raseta Befelatanana Hospital, Antananarivo, Madagascar

[2]Rheumatology Department, Joseph Raseta Befelatanana Hospital, Antananarivo, Madagascar

[3]Cardiology Department, Mahavoky Atsimo Hospital, Mahajanga, Madagascar

[4]Medical Imaging Department, Mahavoky Atsimo Hospital, Mahajanga, Madagascar

Correspondence
Rova Malala Fandresena Randrianarisoa, Department of Internal Medicine, Joseph Raseta Befelatanana Hospital, Antananarivo, Madagascar.
Email: rrmf7763@gmail.com

Key Clinical Message

Behçet's disease should be suspected in young adults who present with heart failure and/or vascular lesions. Cardiac involvement and mesenteric artery aneurysm are rare but should be recognized because of their severity.

KEYWORDS

abdominal aorta, aneurysm, behçet's disease, cardiac manifestations, superior mesenteric artery

1 | INTRODUCTION

Behçet's disease (BD) is a chronic inflammatory disorder of unknown etiology that progresses in relapses.[1] It is defined as a systemic variable-vessel vasculitis according to the Chapel Hill classification.[2] The disease is most common along the "old silk road", in Mediterranean countries, and in the Middle and Far East. Today, it has spread to Europe and the United States as a result of migratory flows.[3,4] In sub-Saharan Africa, it has rarely been described and the incidence appears to be low.[5]

The most common clinical presentations of BD are oral-genital aphthae, skin lesions, ocular involvement, and neurologic and vascular manifestations. Other less frequent presentations include joint involvement, gastrointestinal manifestations (6.3%), epididymitis (7.2%), pleuropulmonary involvement (1.8%), and cardiac involvement (1.8%).[6] Among the vascular manifestations, arterial involvement is less common than venous involvement. Arterial involvement often takes the form of aneurysms and rarely thrombosis.[7] Aneurysms of the superior mesenteric artery (SMA) are uncommon.[8]

We report a case of BD in a patient of Malagasy origin complicated by multiple cardiovascular diseases: cardiac involvement and aneurysms of the abdominal aorta and SMA.

2 | OBSERVATION

A 24-year-old man of Malagasy origin was admitted to the cardiology department of Mahavoky Atsimo

Mahajanga Hospital with a 3-week history of dyspnea. He reported a dry cough, night sweats, and asthenia. Four months prior to admission, he had been hospitalized for similar dyspnea and was diagnosed with dilated cardiomyopathy (DCM) associated with heart failure with reduced left ventricular ejection fraction. The etiology of DCM was not fully understood. A probable diagnosis of pulmonary embolism was made based on the elevated D-dimer level and an S1Q3 appearance on the electrocardiogram.

His other medical history included grade 2 hypertension discovered at age 22, recurrent oral aphthosis for 3 years (4 flares per year), recurrent episodes of angina treated in childhood, and an active smoking history of 1.5 pack-years for the past 2 years. He reported no joint pain or photosensitivity. He had no significant family history. His usual treatment consisted of enalapril 10 mg/d, atenolol 25 mg/d, dapagliflozin 10 mg/d, furosemide 20 mg/d, and rivaroxaban 20 mg/d.

On admission, hemodynamic parameters were normal. General condition was altered, with a performance status index of three. Physical examination revealed hepatojugular reflux, jugular venous distention, painless, white, soft edema of the lower extremities, galloping sounds on cardiac auscultation, and crackling rales at the bases of the lungs. Abdominal palpation revealed a painless pulsating periumbilical mass. Peripheral pulses were present and symmetric. He presented with dermographism, painful aphthae on the inner lips and cheeks, and very painful anal ulcerative lesions (Figure 1). The lymph nodes were clear, and the rest of the examination was unremarkable.

The blood count showed normocytic anemia at 11 g/dL. Neutrophil, lymphocyte, eosinophil, and platelet counts were normal. The erythrocyte sedimentation rate was 103 mm per hour. C-reactive protein was 48 mg/L. Serum glucose, blood ionogram, transaminase levels, coagulation, and lipid profiles were normal. Serologies for HIV, hepatitis B and C, and syphilis were negative. Sputum for tuberculosis and GenXpert were negative. Serum creatinine was 84 μmol/L (clearance: 129 mL/min). NT-proBNP level was 550 pg/mL.

The electrocardiogram showed sinus tachycardia, S1Q3, and left ventricular hypertrophy. The electrical axis of the heart was normal, and there were no conduction or repolarization abnormalities.

Transthoracic echocardiography revealed global hypokinesis, dilatation of all four chambers, functional mitral and tricuspid regurgitation, and a decrease in left ventricular ejection fraction to 37%.

A thoraco-abdomino-pelvic CT scan with contrast injection was performed. In the thorax, it showed a right peripheral triangular condensation with a pleural base suggestive of pulmonary infarction, minimal right pleurisy, and cardiomegaly (Figure 2). In the abdomen, two fusiform aneurysms were found without signs of rupture: an aneurysm of the subrenal abdominal aorta with maximum axial dimensions of 4.1 × 3.31 cm, vertically distributed over approximately 8.44 cm, and an aneurysm of the SMA with axial dimensions of 2.26 × 1.94 cm (Figure 3).

The dyspnea was related to congestive heart failure. The diagnosis of BD complicated by cardiac involvement and aneurysms of the subrenal abdominal aorta and SMA was maintained. Skin pathergy test was negative. Human leukocyte antigen (HLA) typing was not performed. Ophthalmologic examination (after 1 week of corticosteroid therapy) showed no significant pathology.

After a multidisciplinary discussion, we opted for medical management combining corticosteroids with an immunosuppressant and avoiding surgery. The patient received a bolus of methylprednisolone 400 mg (10 mg/kg) for 3 days, followed by prednisolone 40 mg (1 mg/kg/d). Treatment with cyclophosphamide was planned but not started due to financial constraints. Anticoagulation with rivaroxaban 20 mg/d was

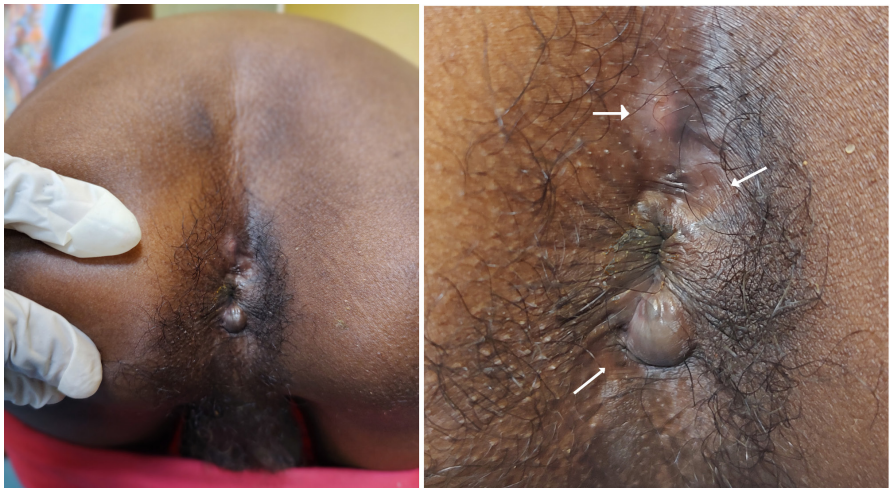

FIGURE 1 Patient's anal ulcerative lesions.

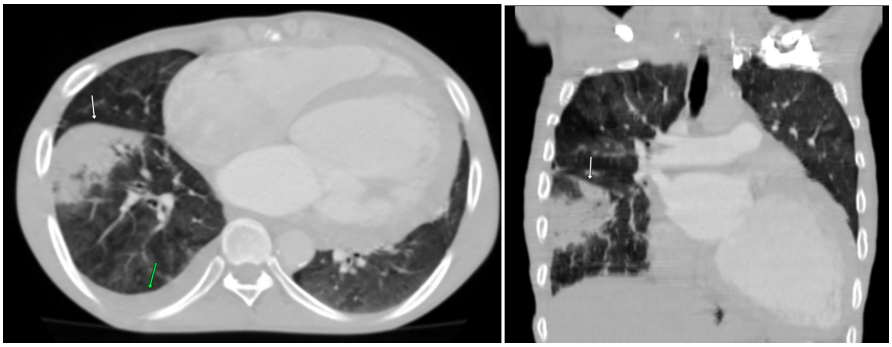

FIGURE 2 Axial (left) and coronal (right) thoracic CT scan. Right pulmonary infarction (white arrow) and minimal right pleurisy (green arrow).

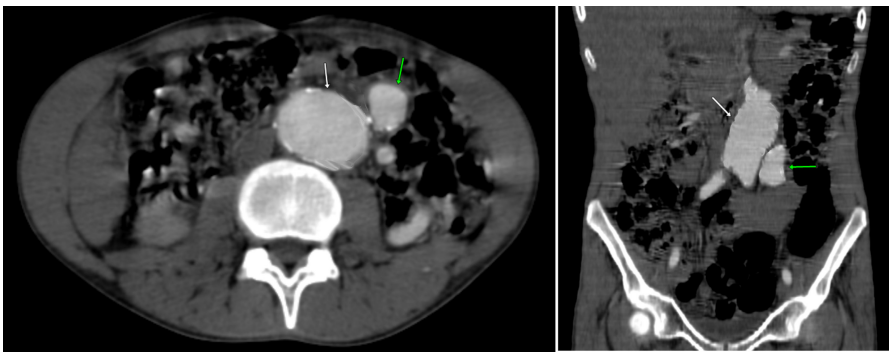

FIGURE 3 Axial (left) and coronal (right) abdominal CT scan. Fusiform aneurysms of the subrenal abdominal aorta (white arrow) and superior mesenteric artery (green arrow) without calcification.

continued. For the treatment of heart failure, enalapril and dapagliflozin were maintained at the same dose with the addition of spironolactone 25 mg/d. The doses of furosemide and atenolol were increased to 80 mg/d and 50 mg/d, respectively.

The initial course was characterized by improvement of dyspnea and regression of lower limb edema. After 1 month of corticosteroid therapy (1 mg/kg), he had no more respiratory symptoms and his general condition had improved. His erythrocyte sedimentation rate was 20 mm per hour and his C-reactive protein was 10 mg/L. Corticosteroid therapy was planned for at least 6 months with a tapered dose, but unfortunately the patient was lost to follow-up.

3 | DISCUSSION

BD was first described by Professor Hulusi Behçet in 1937.[9] Since then, many diagnostic criteria have been established. The International Study Group (ISG) criteria, published in 1990, have a sensitivity of 95% and a specificity of 98%. The main limitations of the ISG criteria are the exclusion of major organ damage, such as vascular manifestations, and the low positivity of the parthergic test.[4] In 2014, the revised version of the International Criteria for Behçet's Disease (ICBD) was proposed, and a score of ≥4 points corresponds to the diagnosis.[1] The ICBD criteria appear to be more sensitive, especially in the early stages of the disease, and are the most widely used today. Our patient had oral aphthosis (2 points), genito-anal ulcerations (2 points), and vascular manifestations (1 point), fulfilling the ICBD criteria.

The age of onset of BD is relatively early. It is usually diagnosed in the second and third decades of life. Diagnosis before the age of 15 and after the age of 50 is exceptional.[3,10] In our case, the diagnosis was made at the age of 24. Both sexes can be similarly affected, but the prognosis is worse in men. In addition, cardiovascular manifestations are more common in men.[3,7]

Cardiac involvement is a serious complication reported in less than 5% of cases.[10] It can manifest as pericarditis, myocarditis, endocarditis, intracardiac thrombosis, myocardial fibrosis, myocardial infarction, and DCM.[6] In our case, the left ventricular systolic dysfunction could be explained by coronary arteritis or DCM. The evaluation of the coronary arteries was insufficient. Coronary angiography and cardiac MRI were not performed. However, the electrocardiogram showed no abnormalities consistent with coronary artery disease. After a multidisciplinary discussion, we maintained the diagnosis of DCM. DCM is less common and can manifest as systolic or diastolic heart failure or be asymptomatic. In a study conducted in Iran, 4.3% of patients developed DCM.[11,12] In light of this observation, BD, although rare, should not be overlooked in young adults presenting with heart failure. Signs of BD should be sought on clinical examination. Special attention to bipolar aphthosis during the initial hospitalization may have led to an early diagnosis of BD.

Vascular manifestations of BD (or angio-Behçet) occur in 7%–32% of cases. They are dominated by

venous involvement, mainly deep vein thrombosis.[13] The majority of patients present with the first vascular manifestation within 5 years of disease onset. In 20% of cases, vascular involvement occurs simultaneously.[7] In our case, vascular lesions were discovered at the time of BD diagnosis.

Arterial involvement is less common, occurring in 5%–10% of cases. The frequency is probably underestimated, as autopsy data have reported a higher proportion.[10] Arterial lesions usually present as aneurysms and rarely as thrombosis.[7] Pseudoaneurysms are most common. Arterial aneurysms are often multiple, mainly affecting the abdominal aorta and femoral arteries.[10]

Involvement of the SMA is very rare, accounting for 5.5% of all visceral aneurysms.[8] A review of the literature by Kakehi et al. in 2019 found 15 cases.[14] Men were most affected. Abdominal pain was the main symptom (80%). Lesions consisted of aneurysm (11 cases), thrombosis (four cases), and dissection (one case) and were mostly associated with other arterial manifestations. In six patients, the discovery of SMA involvement was concomitant with the diagnosis of BD. In our case, it was an asymptomatic aneurysm of the SMA.

Pulmonary artery involvement is rare, with an overall incidence of less than 5%. It usually occurs 3–4 years after disease onset.[6,7,15] In our case, it is possible that the pulmonary infarction and minimal pleurisy seen on the CT scan were indirect signs of pulmonary embolism. In addition, the echocardiogram was typical. Lung scintigraphy is indicated but was not performed due to lack of technical resources. The diagnosis remained uncertain, but anticoagulation was started immediately.

The two aneurysms associated with cardiac involvement represent the originality of our observation. These are important complications of BD. Atherosclerosis, infection, and Takayasu's arteritis are the main differential diagnoses for aortic involvement. The young age, low tobacco exposure, and absence of diabetes or dyslipidemia did not suggest an atheromatous origin. Infectious tests (HIV, viral hepatitis, syphilis, and tuberculosis) were negative. Classification criteria for Takayasu's arteritis were not complete.

A multidisciplinary approach is essential for the management of cardiovascular complications. The 2018 European League Against Rheumatism (EULAR) recommendations suggest the combination of high-dose corticosteroids with cyclophosphamide in the acute phase of angio-Behçet. Surgical treatment should be performed at a later stage to reduce the risk of complications. Curative anticoagulation is recommended for venous thrombosis in the absence of pulmonary artery aneurysm.[16] There is no consensus on the treatment of cardiac involvement. The goal is to reduce inflammation to limit lesion progression and treat symptoms. The efficacy of anti-inflammatory and immunosuppressive drugs has been demonstrated.[12] However, treatment is individualized according to the patient's age and the type and severity of complications. We chose medical management alone because the aneurysms were asymptomatic and our local technical platform does not allow us to perform vascular surgery.

4 | CONCLUSION

BD should be suspected in young adults with heart failure and/or vascular lesions. Cardiac involvement and SMA aneurysm are rare but should be recognized because of their severity and complications. Management is multidisciplinary and varies according to the type and severity of complications.

AUTHOR CONTRIBUTIONS

Rova Malala Fandresena Randrianarisoa: Data curation; investigation; writing – original draft; writing – review and editing. **Lalao Nomenjanahary Rakotonirina:** Investigation. **Mirantosoa Fabiola Ravelonjatovo:** Data curation; investigation. **Nofihinina Joharisantatra Andrianjanaka:** Data curation; investigation. **Nouraly Habib:** Data curation; investigation. **Narindrarimanana Avisoa Randriamihangy:** Validation; visualization; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

Requests for additional clinical data can be addressed to the corresponding author.

CONSENT

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

ORCID

Rova Malala Fandresena Randrianarisoa https://orcid.org/0000-0003-1168-0200

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