

## Angio-Behçet: When Arterial Atypia Is at The Heart of The Disease: About Three Observations

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Received: 03 April 2024; Accepted: 13 May 2024

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### ABSTRACT

Behçet's disease is a systemic vasculitis, the etiology of which remains obscure. Ocular, cutaneous, articular, neurological, and vascular involvement remain the most significant. We report a series of 3 cases of Behçet's disease with atypical arterial vascular manifestations, collected in an internal medicine department. The majority of our patients, with an average age of 45 years at the onset of arterial involvement, were male, with only one female. The arterial manifestations observed were mainly characterized by atypical locations, including visceral arteries, and presented as arterial occlusions. Corticosteroid therapy was necessary in all cases, in combination with other treatments such as anticoagulants, colchicine, and immunosuppressants. Overall, the outcome was favorable for the majority of patients. In the literature, arterial involvement is much less frequent than venous involvement, mainly affecting the aorta and pulmonary arteries, and manifesting primarily as aneurysms or pseudoaneurysms, even less frequently as occlusions, and notably with rare or even exceptional locations found in very few studies, making our observations original. However, arterial involvement poses therapeutic challenges due to its recurrent nature, which can impact prognosis.

**Keywords:** *Angio-Behcet, Arterial Atypia, Systemic Vasculitis, Corticosteroid Therapy*

### Introduction

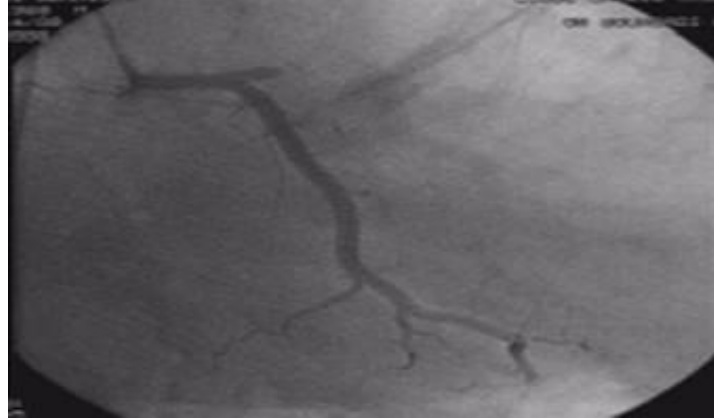
Behçet's disease is a multi-systemic inflammatory vasculitis of unknown etiology. Diagnosis is based on a clinical score including oral and genital aphthosis, neurological, ophthalmological and vascular involvement.

Vascular involvement is often severe and can be life-threatening. Arteries and veins of all calibres may be affected. Arterial involvement is rare, accounting for 5-10% of cases, and may involve occlusions, arterial aneurysms and/or thrombosis. The prognosis for arterial involvement remains severe, representing the main cause of death (30-40%).

We describe three cases of patients treated for Behçet's disease with atypical arterial vascular disease.

### **Observation 1:**

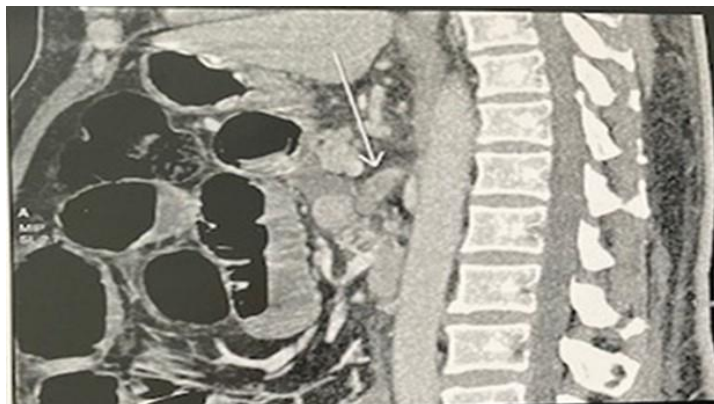
This concerns a 38-year-old patient known to be a chronic smoker with a 5-pack-year smoking history, with a BMI of 23 mg/m<sup>2</sup>, followed since the age of 30 for Behçet's disease diagnosed according to the international criteria of 2013, presenting with bipolar aphthosis, bilateral posterior uveitis, and pseudo-folliculitis. Initially, he received oral corticosteroid therapy, colchicine, and an immunosuppressant such as azathioprine for his ocular involvement, with poor treatment adherence. Admitted five years later to the emergency department, on day 2 of anginal chest pain, an immediate EKG revealed sinus rhythm at 60 BPM with ST-segment elevation in the anteroseptal and lateral leads with Q waves indicative of necrosis, and elevated troponin levels. The diagnosis of an ST-elevation myocardial infarction (STEMI) was made. A transthoracic echocardiogram showed features of dilated cardiomyopathy, with akinesia of the territory of the anterior interventricular artery (left ventricular ejection fraction at 35%). Diagnostic coronary angiography revealed acute occlusion of the proximal anterior interventricular artery ([Fig. 1](#)). Laboratory investigations for atherosclerosis risk factors were negative: fasting blood glucose: 0.96 g/L, lipid profile: normal (total cholesterol: 1.6 g/L, HDL: 0.5 g/L, LDL: 1 g/L, triglycerides: 0.66 g/L). Thrombophilia screening, including antiphospholipid antibodies, protein S, protein C, antithrombin III, hemostasis profile, and immunological panel, was normal. Infectious workup, including syphilitic serologies, HIV, viral hepatitis, and tuberculosis screening, was negative. Therefore, acute myocardial infarction treatment was promptly initiated, with conventional management of ischemic heart disease, including stent placement and oral BASIC therapy, along with intravenous corticosteroid therapy (methylprednisolone 1g/day) for three days. Azathioprine immunosuppressant at a dose of 150 mg/day, colchicine, and smoking cessation were also administered. The patient's clinical course was favorable, with vascularitis stability and no cardiac functional symptoms. The diagnosis of myocardial infarction associated with Behçet's disease was established.



**Figure 1:** Coronary angiography showing complete occlusion of the anterior interventricular artery.

### **Observation 2:**

A 50-year-old patient, diagnosed with type 2 diabetes and hypertension for 5 years under treatment, and followed up for Behçet's disease since the age of 33, presenting with bipolar aphthosis, right posterior uveitis, and thromboembolic venous involvement of the right lower limb, previously treated with anticoagulants and corticosteroids but discontinued treatment for years. Admitted to the emergency department for acute occlusive syndrome, characterized by diffuse abdominal pain and constipation, confirmed by abdominal angioscanner revealing occlusive thrombosis of the proximal portion of the superior mesenteric artery [Fig. 2](#). Laboratory tests showed an inflammatory syndrome with CRP levels at 74mg/l, negative thrombophilia screening, negative JAK 2 mutation, and negative tumor markers. The patient was immediately started on unfractionated heparin and urgently transferred to the operating room for revascularization surgery without intestinal resection. He received five days of low molecular weight heparin, antiplatelet therapy, oral corticosteroids at a dose of 1mg/kg/day, and immunosuppressive therapy with azathioprine 150mg/day, resulting in significant clinical and radiological improvement.



**Figure 2:** Total thrombosis of the proximal portion of the superior mesenteric artery.

**Observation 3:**

A 47-year-old female patient, diagnosed with type 2 diabetes and insulin-dependent, with a history of Behçet's disease since the age of 27, characterized by bipolar aphthosis and posterior uveitis. Initially treated with colchicine and a methylprednisolone bolus of 15mg/day, followed by oral corticosteroid therapy for ocular involvement with favorable outcomes. At the age of 45, she was admitted due to renal colic without arterial hypertension. Abdominal angioscanner revealed thrombosis of the right renal-pyelic artery and left interlobar renal artery, with no impact on renal function. Laboratory tests showed negative thrombophilia screening including protein S, protein C, antithrombin III, and negative lupus anticoagulant testing. There was no evidence of inflammatory syndrome, and other laboratory findings were unremarkable. Therapeutically, the patient was started on low molecular weight heparin followed by vitamin K antagonist, corticosteroid therapy at 1mg/kg/day, and six-monthly cyclophosphamide pulses followed by maintenance therapy with azathioprine 150mg/day. The patient showed clinical and radiological improvement with reperfusion of the renal arteries.

**Discussion:**

The vascular involvement known as 'Angio-Behçet' is the originality of this condition, as it occurs in young subjects, often male, with no associated vascular risk factors. The vascular tropism of MB was reported by Adamantiades in 1946. Vascular involvement affects both veins and arteries of all calibres. It is particularly serious, since it affects both vital and functional prognosis.

Arterial involvement occurs in only 2 to 8% of cases (Cormier *et al.*, 1993). It affects arteries of all calibres: the large trunks are more frequently affected than the distal arteries, which means that Behçet's disease can take on the appearance of Takayasu's disease (Wechsler *et al.*, 1989; Wechsler *et al.*, 1987). It takes three forms: occlusions, thromboses or, more often, arterial aneurysms. The latter classically produce true arterial aphthae which correspond to real perforations (Ajili *et al.*, 2014).

The pathophysiology of arterial vascular damage in MB, like that of the disease itself, remains poorly understood. Some authors believe that the anatomical basis of these arterial lesions is anomalies in the arterial wall, particularly involving parietal factors (Hamza, 2000; McNeil *et al.*, 1991). The mesenteric artery and the cerebral artery: rare cases have been described in the literature. Park, *et al.* (1984) reported the first case of thrombosis of the superior mesenteric artery. Hassan Khodja, *et al.* (1991) reported a second case of thrombosis of the superior and inferior mesenteric arteries. Two other cases were reported by Wechsler *et al* and Mercié, *et al.* (1996). Only one case was identified in our study.

**Coronary Involvement:**

This type of involvement remains exceptional, with only around thirty cases described in the literature (Kraiem *et al.*, 2004). Tohme, *et al.* (2003) reported a single case of occlusion of the anterior interventricular artery of the coronary artery and Lakhanpal, *et al.* (1985) described 3. It most often manifests itself as a myocardial infarction affecting young male subjects with no cardiovascular risk factors (Wechsler *et al.*, 1999), and is rarely the first event. It may involve occlusion of a coronary artery or, more rarely, a coronary aneurysm. This is consistent with our patient. Occlusion of the anterior interventricular artery is the cause, in 50% of cases, of a ventricular aneurysm, which has a poor prognosis. Mortality is 20%, and is linked to the direct complications of coronary insufficiency [Table 1](#).

**Table 1:** Location of arterial occlusions.

Seat	Our series	(Lakhanpal <i>et al.</i> , 1985)	Park <i>et al.</i> , 1984	Tohme <i>et al.</i> , 2003
Artère pulmonaire	0	5	0	1
Artère sous Clavière	0	1	0	1
Renal artery	1	3	1	1
Abdominal aorta		1		
Superior mesenteric artery	1	0	1	0
Coronary artery	1	3	0	1

Angio CT scan and MRI have become the reference examinations for the diagnosis and follow-up of these arterial lesions (Izzillo *et al.*, 2004). Two of our patients underwent abdominal angio CT scan.

In terms of the treatment of vascular damage, there are many therapeutic tools available but there is no precise therapeutic consensus. The treatment recommended by the European League Against Rheumatism is based, as in other vasculitides, on corticosteroid therapy, which remains the cornerstone of treatment, and/or immunosuppressants. In the case of occlusive or stenosing arterial disease, medical treatment is often sufficient, combining corticosteroids (1mg/kg/d), anticoagulants and/or anti-platelet agents. Immunosuppressive drugs (cyclophosphamide/ azathioprine) may be indicated in cases of severe and recurrent occlusive lesions (Hatemi *et al.*, 2008), as in the case of our three patients, all of whom received intravenous immunosuppressive drugs followed by oral treatment. Surgery is only indicated in the event of failure of medical treatment with threatening ischaemic lesions.

## Conclusion

Arterial involvement in Behçet's disease is both rare and serious, presenting therapeutic challenges due to its recurrent nature, which can jeopardize prognosis. Early diagnosis and prompt initiation of corticosteroids and immunosuppressants in severe cases, combined with surgery, can improve prognosis.

**Conflict of Interest:** No

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