



## Ruptured popliteal artery aneurysm: An initial presentation of Behçet's disease

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### Abstract

Behçet's Disease (BD) is an uncommon systemic vasculitic disorder, that can affect multiple organ systems. Arterial and venous aneurysmal and occlusive manifestations can occur. We report a case of a 25-year-old male presenting with a 4-week history of left leg pain and swelling, with acute deterioration due to ruptured popliteal aneurysm, confirmed on imaging. He underwent emergency popliteal bypass with ipsilateral reversed great saphenous vein. Subsequent workup revealed pulmonary aneurysms and features suggestive of BD. He received aggressive immunosuppressive therapy to good effect. The management of the patient's popliteal aneurysm, diagnostic procedure and subsequent management are discussed.

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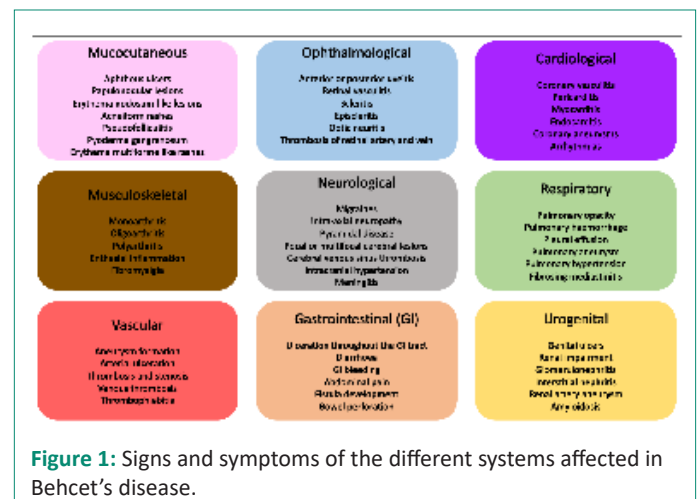
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### Background

Behçet's disease is an uncommon systemic inflammatory vasculitis with an unidentified aetiology affecting both arteries and veins [1]. BD can involve the mucocutaneous, musculoskeletal, vascular, ophthalmological, neurological, gastrointestinal, cardio-respiratory, and urogenital systems with a relapsing-remitting course [1]. BD is a complex chronic condition which can present with a varying number of symptoms and signs, mimicking other conditions and thus making it a challenging diagnosis [1] (Figure 1). BD is most commonly seen in young males (20-40 years-old) with a high prevalence in the Middle East and Turkey [1]. The International Study Group (ISG) criteria state that, for a formal diagnosis there must be 'presence of oral aphthosis together with any two of; genital ulcers, ocular lesions, skin lesions and positive pathergy test'. More recently, the new International Criteria for Behçet's (ICBD) includes the presence of neurological and vascular manifestations [2]. Vascular manifestations such as arterial aneurysms, stenoses, occlusions, and thromboses can affect up to 40% of patients with BD and are more commonly seen in males [3]. Vascular manifestations of Behçet's present early on in disease progression and are associated with higher morbidity and long-term mortality. This case highlights the importance of history taking and examination, especially for an atypical vascular presentation.



**Figure 1:** Signs and symptoms of the different systems affected in Behçet's disease.



## Case presentation

A 25-year-old Moroccan gentleman presented to a London district general hospital with a 4-week history of progressive left leg pain, unable to weight-bear and requiring a stick for mobilisation. 48 hours prior to his admission his pain became unbearable even at rest with associated tingling sensation on his toes. He was provided by his general practitioner paracetamol, ibuprofen, dihydrocodeine and oral morphine sulphate with no significant improvement. He denied any additional symptoms such as cough, breathlessness, headaches, nausea, vomiting, chest or abdominal pain and visual changes. Examination demonstrated left proximal calf tenderness with associated localised swelling, intermittent paraesthesia at L4/L5 dermatomes and preserved pedal pulses with no other signs of compartment syndrome. A neurovascular examination of the right leg was normal. Medical history consisted of bilateral pulmonary embolisms (4-months prior), right leg Deep Vein Thrombosis (DVT) (1-month prior) and sport trauma with internal fixation to right tibia and fibula (2014). He took regular paracetamol, morphine, amitriptyline and therapeutic dose low molecular weight heparin (tinzaparin sodium) injections, recently having switched from rivaroxaban due to the development of DVT. He was allergic to sulfonamide and aspirin. He had recently immigrated to the UK from Morocco with his wife and worked in a delivery warehouse. He was a current smoker (7-pack years), with no alcohol or illicit drug use history.

Initial Computed Tomographic Angiogram (CTA) of the lower limbs demonstrated an irregular 3.7x1.5x3.1 cm (MLxAPxCC) hypervascular lesion of the deep proximal left calf consistent with a pseudoaneurysm with evidence of active extravasation. He was transferred to a tertiary university hospital under the vascular surgical team, where he underwent a left popliteal - tibioperoneal trunk bypass with ipsilateral reversed great saphenous vein via a posterior approach, and ligation of the anterior tibial artery.

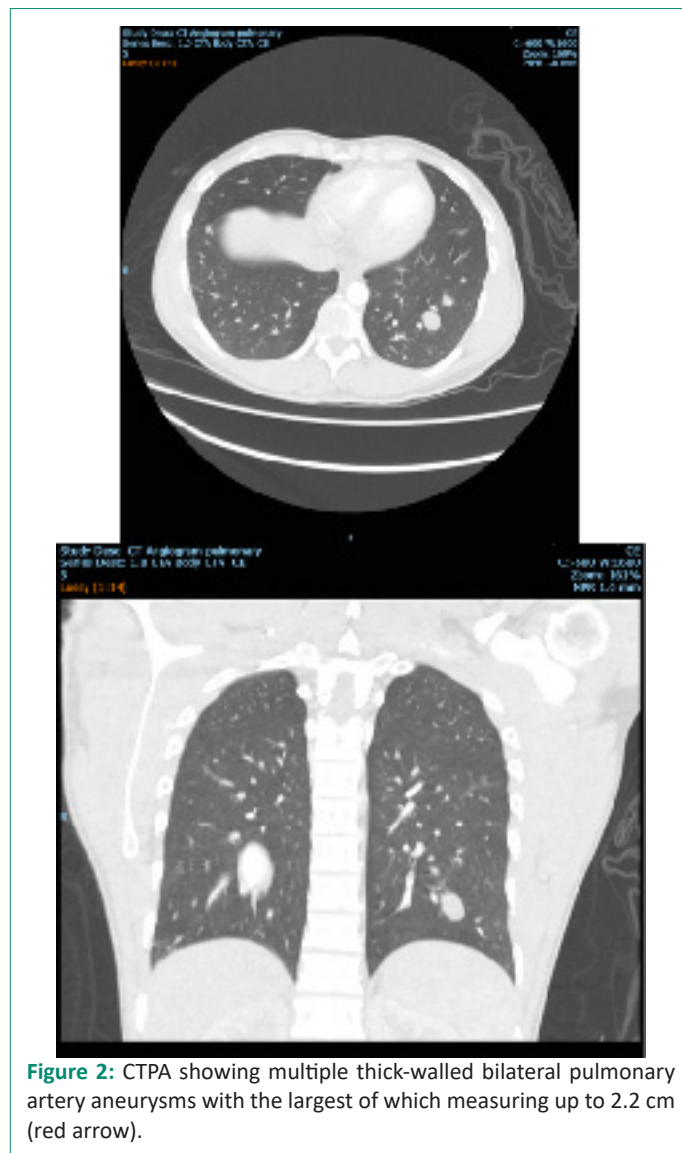
## Investigations

Two days postoperatively, the patient reported sudden onset left calf and popliteal fossa pain with a cold foot and monophasic handheld doppler signal at the posterior tibial artery, which was multiphasic the day before. He also developed a new oxygen requirement with shortness of breath and tachycardia. Given his past medical history he was investigated further with a CTA of the lower limbs and a CT Pulmonary Angiogram (CTPA) (See figures below). The former showed a large peripheral enhancing low attenuation collection surrounding the bypass graft with an appropriately sited postoperative drain, the graft was patent, with no evidence of extravasation. The latter showed multiple thick-walled bilateral pulmonary artery aneurysms, the largest of which measuring up to 2.2 cm, with no right heart strain or evidence of infarction. Further questioning revealed a history of recurrent painful mouth ulceration as well as genital ulcers for the last 4-5 months. On re-assessment, examination demonstrated pathergy at the cannula insertion site and findings consistent with anterior uveitis as well as oral and genital aphthosis. Blood tests revealed the following:

A vasculitis screen was sent, this revealed a high ESR, WCC and CRP in the context of recent major surgery. Immunoglobulins were all within normal limits and subsequent retroviral, tuberculosis and syphilis screening were negative. Histology results from the patient's pseudoaneurysm reported findings favouring an arteritis but were not discriminatory.

Blood test	Admission day	2-days postoperatively	Discharge day
Hb (g/L)	125	114	116
WCC (x10 <sup>9</sup> /L)	10.34	14.92	20.75
PLT (x10 <sup>9</sup> /L)	396	446	794
CRP (mg/L)	160	375	6
ESR (mm/hr)	-	100	-
Urea (mmol/L)	3.7	4.6	6.1
Creatinine (μmol/L)	71	60	61

HB: Haemoglobin; WCC: White Cell Count; PLT: Platelets; CRP: C-Reactive Protein; ESR: Erythrocyte Sedimentation Rate.



**Figure 2:** CTPA showing multiple thick-walled bilateral pulmonary artery aneurysms with the largest of which measuring up to 2.2 cm (red arrow).

## Differential diagnosis

The differential diagnoses for pulmonary aneurysms are broad. Vascular causes include pulmonary artery hypertension, tuberous sclerosis and cystic medial degeneration, all of which could be excluded given the age of the patient and the absence of characteristic systemic signs. Infective causes include mycotic aneurysms, tuberculosis or syphilis, all of which were felt less likely as screening tests were negative. Therefore, the two differentials left are idiopathic or autoimmune, such as Behçet's disease [4,5]. Given the patient's additional signs and symptoms discovered on further history taking and examination the most likely diagnosis is Behçet's disease.

## Treatment

The patient was reviewed by the rheumatologists who initiated treatment with 1g IV methylprednisolone once daily for a total of 3 days. CT-Positron Emission Tomography (PET) excluded other sites of vasculitis and showed no evidence of hypermetabolic activity in the pulmonary aneurysms. Upon completion of the methylprednisolone, the patient was started on an oral high-dose steroid therapy (60 mg prednisolone once daily), after which his aphthosis, anterior uveitis and mood improved, and was discharged on a weaning schedule of his steroid therapy. The patient was also reviewed by the haematology team, due to his past medical history of deep vein thrombosis and pulmonary aneurysm, who suggested the patient to continue on treatment-dose low molecular weight heparin (tinzaparin) injections with a plan to switch to rivaroxaban after review in clinic.

## Outcome and follow-up

The patient was discharged on a weaning dose of oral prednisolone with adjunctive monthly infliximab infusions. 3-month follow-up by the rheumatology and haematology teams showed good response to steroid and infliximab therapy with complete resolution of his presentation symptoms, ESR reduced to 2 mm/hr and no further episodes of venous thromboembolisms on rivaroxaban. A repeat CTPA after 4 months showed resolution of most pulmonary aneurysms with only a solitary aneurysm of 8 mm remaining, possibly a remnant of the largest previous one. He continued to improve with his mobility post operatively, being able to mobilise independently now and therefore he was discharged from the vascular service with advice to continue lifelong anti-thrombotic therapy. Lifelong surveillance will continue under his BD specialists.

## Discussion

BD is a complex chronic inflammatory condition with multi-organ involvement for which there is currently no formal diagnostic histopathological or laboratory confirmatory test [6]. Vascular manifestations of BD are commonly reported in central and peripheral large arteries in the form of thromboses, aneurysms and pseudoaneurysms [1,3]. We present here a case of young man with previous history bilateral pulmonary embolisms and lower extremity DVT, who presented to hospital with limb ischaemia secondary to a popliteal pseudoaneurysm and subsequently diagnosed with multiple high rupture risk pulmonary arterial aneurysms on a background of undiagnosed BD. This is the first case in the UK to our knowledge that a popliteal pseudo-aneurysm and subsequently pulmonary aneurysms lead to the diagnosis of BD. Similarly, a case report from India described a 60-year-old man that had to undergo 4 vascular surgeries for lower limb occlusion, thrombosis and aneurysms prior to the diagnosis of Behcet's disease [7].

Given the complexity of disease and varied presentation, patients with BD have poor prognosis if aneurysmal lesions are identified, with pulmonary aneurysm being the most common fatal complication in men [8]. A retrospective study from China in 2019 showed that development of aneurysms happens in the early stages of treatment naïve male BD patients, and new oral/genital lesions or pathergy reactions can be independent risk factors for aneurysm development [9]. Interestingly it has been reported that BD patients with pulmonary artery aneurysms usually present with haemoptysis, which was not observed in

our patient [9,10].

Since 2018 the European League Against Rheumatism (EULAR) suggested that glucocorticoids and anti-TNF- $\alpha$  therapy should be the mainstay of treatment in patients with aneurysmal lesions such as with our case. Immunosuppressive therapy is believed to regress both thromboses and aneurysms [11,12]. In a retrospective study in Turkey, 70% of patients showed aneurysmal regression with aggressive treatment, nonetheless 20% of those recurred with a significant increase in the mortality rate [13]. It is important to note that TNF- $\alpha$  inhibitors can be limited by intolerance, patient contraindications, inadequate response or loss of efficacy and there is still increased need for more evidence [11].

Acute arterial involvement must be managed as a medical emergency with intravenous methylprednisolone therapy, appropriate rheumatology review and urgent initiation of immunosuppressive therapy [7,14]. BD patients with vascular manifestations might also require surgical interventions such as endovascular stenting, embolisation or open aneurysmectomy and/or bypass grafting. Management of a peripheral artery aneurysm is primarily determined by the clinical presentation, location of the aneurysm, risk of rupture and whether the BD patient has active or controlled disease [14,15]. These patients remain a challenge for vascular surgeons as the vascular inflammation increases the intra and post-operative risks and complications. BD patients are also prone to recurrence of the aneurysmal lesions requiring subsequent surgical interventions [9]. Studies have shown the important contribution of medical intervention with immunosuppressant medications either to avoid surgical intervention or reduce the risk of postoperative complications, thereby highlighting the need for appropriate evaluation of medical and surgical intervention of aneurysmal lesions in patients with BD [8,9,16]. A recent literature review showed that endovascular treatment of vascular BD is increasingly preferred due to higher success rates and low recurrence. Endovascular repair remains a safe alternative to traditional open surgical intervention as it minimises endothelial injury and reducing post-operative complications [17].

## Conclusion

In conclusion, vascular manifestations have the leading cause of death in BD patients and therefore early identification and management is key. CTA remains a valuable imaging modality in the diagnostic work-up of BD [18]. Open and endovascular surgical repair are available options for peripheral artery aneurysms with high recurrence rates thus highlighting the importance of early immunosuppressive therapy in BD.

## Learning points

- When presenting with atypical vascular malformations such as pseudo-aneurysms in young men, Behcet's should be considered in your differential diagnosis.
- Vascular thrombosis or occlusion can be a predictive factors for aneurysmal lesions in Behcet's disease.
- Vascular manifestations of Behcet's often present early, and with concomitant vascular features
- Pulmonary aneurysms are life-threatening complications of Behcet's and clinicians should have a low-threshold for investigating if any clinical suspicion.

**Patient's perspective**

"Hello everyone,

Today I am here to tell you a short story about a medical condition that I am currently going through and the meanings and pain that I have experienced. First, it started with a sharp pain in the right side of my chest, difficulty breathing and a high temperature. I therefore, tried to call the ambulance and from there the story became clear that there was something strange going on in my body. For a few weeks I was also experiencing some infections in my mouth, genitals, swelling of my legs and difficulty sleeping. I decided to take this matter to my local hospital and after a lot of effort, patience and pain, I was told that I have a chest infection. I was not sure where the problem came from and after months of having lots of different tests I was diagnosed with multiple clots in my lungs and legs. Few weeks ago, I started suffering from leg pain and I tried to return to the hospital because the strong painkillers (morphine) that the GP gave me did not help me. There I had a scan of my leg and suddenly I was transferred to another hospital for surgery. After the surgery on my left leg, I had a scan of my chest which showed aneurysms on my lungs. During that period in the new hospital and by putting all of this together it was discovered what was happening in my body as I was diagnosed with Behcet's disease. Here, I learned what is happening in my body thanks to the specialised medical team, so I learned about all the causes. I feel better day by day and the condition will be monitored until the end.

Well, it seems appropriate that my life story fits on one page."

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