Case Report **IJCR (2021) 5:195**

International Journal of Case Reports
(ISSN:2572-8776)

Isolated Aortitis with multiple Aortic aneurysms and widespread venous, arterial and intracardiac thrombosis: A challenging Vascular Behcet's Disease case management

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ABSTRACT

In 1937, Hulusi Behcet's brought to our attention the existence of a disease characterized by recurrent oral aphthae and any of several systemic manifestations including genital aphthae, ocular disease, skin lesions, gastrointestinal involvement, neurologic disease, vascular disease, or arthritis [1-3]. He became the first physician to describe this and the disease was henceforth named after his name. The aetiology of this disease remains unclear however it has been noticed that Behcet's syndrome is remarkable for its systemic vasculitides which include its ability to involve blood vessels of all sizes (small, medium, and large) on both the arterial and venous sides of the circulation with occlusive nature [4]. This makes Vasculitis the major cause of the clinical manifestations of the disease with the involvement of almost any organ.

We herein report an unusual case of a thirty-year-old Syrian man with no past medical illnesses. On presentation, he suffered from a sudden onset of shortness of breath and a history of hoarseness of voice for less than a week, and drastic weight loss of about thirty kgs within a month. Imaging confirmed the presence of pulmonary embolism, bilateral deep venous thrombosis, intracardiac thrombosis, popliteal artery thrombosis with thoracic, and abdominal aortic aneurysms. This case highlights the importance of having a high suspicion of the possibility of Behcet's disease when multiple thrombi and aneurysms manifest in a patient on the silk root country. Thereby, stressing the importance of familiarizing ourselves with such unusual presentations of Behcet's disease and the course of this unusual presentation with some learning points with a good literature review included below.

Keywords: BD Behcet's disease, CT Computed Tomography, MRI Magnetic Resonance Imaging, ICT Intra Cardiac Thrombus, AAA Aortic Artery Aneurysm, DVT Deep Venous Thrombosis

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How to cite this article:
Ahmed AR Mohamad Al Ani, Abdul-Wahab Al-Allaf, Ahmed Mansour Al Ani, Haajra Fatima, Haneen Ahmed Al Ani. Isolated Aortitis with multiple Aortic aneurysms and widespread venous, arterial and intracardiac thrombosis: A challenging Vascular Behcet's Disease case management. International Journal of Case Reports, 2021 5:195

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Website: http://escipub.com/

Introduction

Behcet's disease is a multisystem inflammatory disorder with a chronic course. Vasculitis is thought to be the major pathological feature of this disease with the involvement of almost all organs [5-6]. Intracardiac thrombus occurring as a thrombotic manifestation of Behcet's disease is an unusual presentation and represents a challenge in diagnosis and treatment [7].

Although it has a worldwide distribution, the prevalence of Behcet's disease is low in the western world. Behcet's disease is rare in America and Europe. The highest prevalence is in Asia, the Middle East, and Mediterranean areas which correspond to the high prevalence of the HLA-B51 allele. BD is difficult to diagnose, and in the absence of a definitive diagnostic laboratory test, reliance is mainly on characteristic clinical features. Oro-genital ulceration is such a constant feature that it is a cornerstone of the diagnosis. Diagnosis can be made with confidence in the presence of concomitant ocular involvement, arthritis, skin lesions, CNS, or cardiac disease [7].

This disease is seen mainly in young men aged 25-35 years. Behcet's disease has become well recognized all over the world and its clinical features and incidence vary according to the country and type of speciality clinic from which the report arises. Two mechanisms have been proposed for its clinical manifestation; vasculitis and hypercoagulable status [4]. However, several studies have failed to demonstrate the association between coagulation abnormalities and venous thrombosis in Behcet's disease.

Case Presentation

We present a thirty-year-old Syrian man with no past medical history and who presented on 21.01.2020 with two-days shortness of breath and lower back and left thigh pain on further history taking it was revealed that he had lost thirty kgs in the last one month. He also gave an acute history of dysphagia for liquids and hoarseness of voice for over a week. His family history for similar conditions and malignancy was unremarkable. Examination findings did not reveal lymphadenopathy or organomegaly and neurological findings were normal. As the patient was tachypneic and tachycardic urgent CT pulmonary angiogram was done and a diagnosis of pulmonary artery embolism associated with a saccular thoracic arch aneurysm was established (Fig 1 & 2 A, B). Therapeutic anti-coagulation was commenced. A transthoracic echo revealed reduced systolic LV function (EF 33 %), global hypokinesia of LV, and a fresh thrombus in the right ventricle cavity extending into the RVOT (Fig 1B & 3). Physical examination revealed the absence of left radial pulsation. The lower limb pulses were also absent on both sides raising the proximal arterial lesion. The abdominal US was done for that and to rule out retroperitoneal bleed as haemoglobin dropped by four grams in one day. A 10 cm long and 5.5 cm wide Abdominal aortic aneurysm just distal to the origin of the renal arteries down to the bifurcation pressing on lumbar vertebra causing anterior scalloping of L3 vertebra was found (Fig 4, 5A & B). This clearly explaining his lower back pain. A movable flap within the aneurysmal sac and turbulence of the blood at the site was spotted making it difficult to rule out dissection. A CT angiography was done, which revealed a large saccular aneurysmal dilatation of the abdominal aorta associated with a mural thrombus and surrounding hematoma suggestive of a leak. The leak was found to extend to the bifurcation of the aorta to the common iliac arteries and explaining his sudden drop in the Haemoglobin (Fig 5A). Below the diaphragm, there was another small saccular aneurysmal dilatation (Fig 6). The left popliteal artery was completely occluded and the right anterior and posterior tibial arteries appeared partially occluded (Fig 7). Thrombus was seen in the deep venous system of both lower limbs suggestive of DVT, despite that there was no sonographic evidence of deep venous thrombosis, which has been done a day before.

Accordingly, urgent infrarenal aortic aneurysm repair was done (Fig 8). Following the surgery,

the patient was hemodynamically stable. He was started on a therapeutic dose of heparin infusion with hourly monitoring of lower limb pulses.

The patient was assessed by the rheumatology team, who suggested that the most likely diagnosis was Behcet's Disease. This was based on the possibility of the inflammatory thrombotic vasculitis presenting with pulmonary embolism, right ventricle fresh thrombus, bilateral DVT, saccular thoracic arch aneurysm, left subclavian aneurysmal rupture with hematoma, infrarenal abdominal aortic aneurysm leak and left popliteal artery occlusion in a young patient from the Mediterranean origin. The less likely differential diagnosis was Takayasu's arteritis with possible Antiphospho-lipid syndrome.

His inflammatory markers were high with ESR 59 mm/hr and CRP 300 mg/L, CBC revealed hypochromic microcytic anaemia with the Hb 9.3 g/dL. They recommended starting prednisolone at 60 mg daily while awaiting the full immunology.

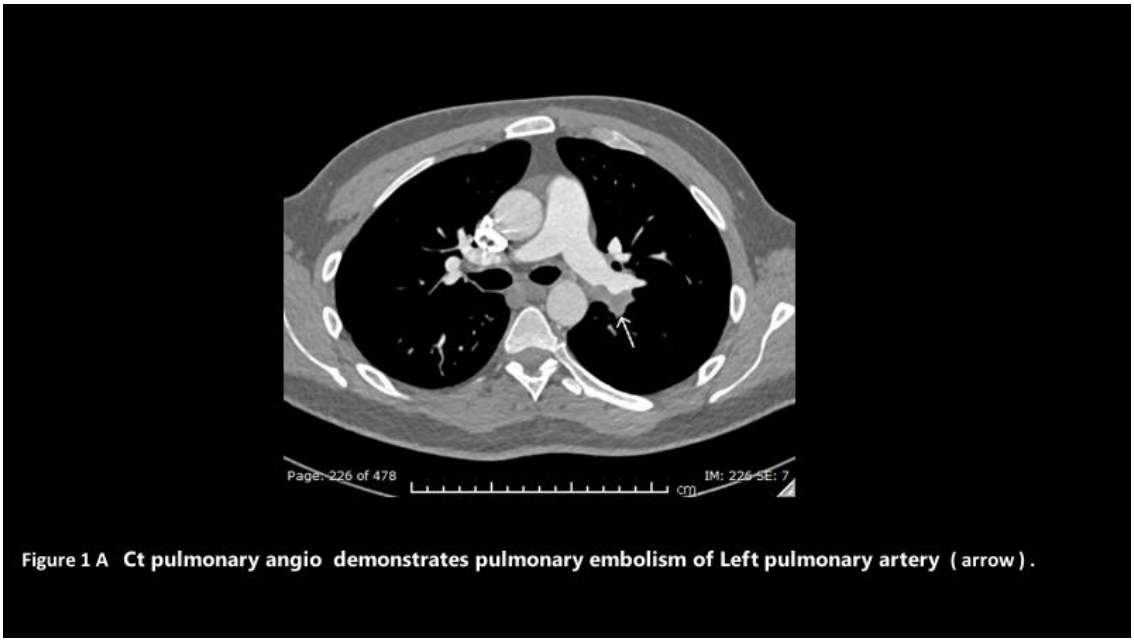


Figure 1 A Ct pulmonary angio demonstrates pulmonary embolism of Left pulmonary artery (arrow).

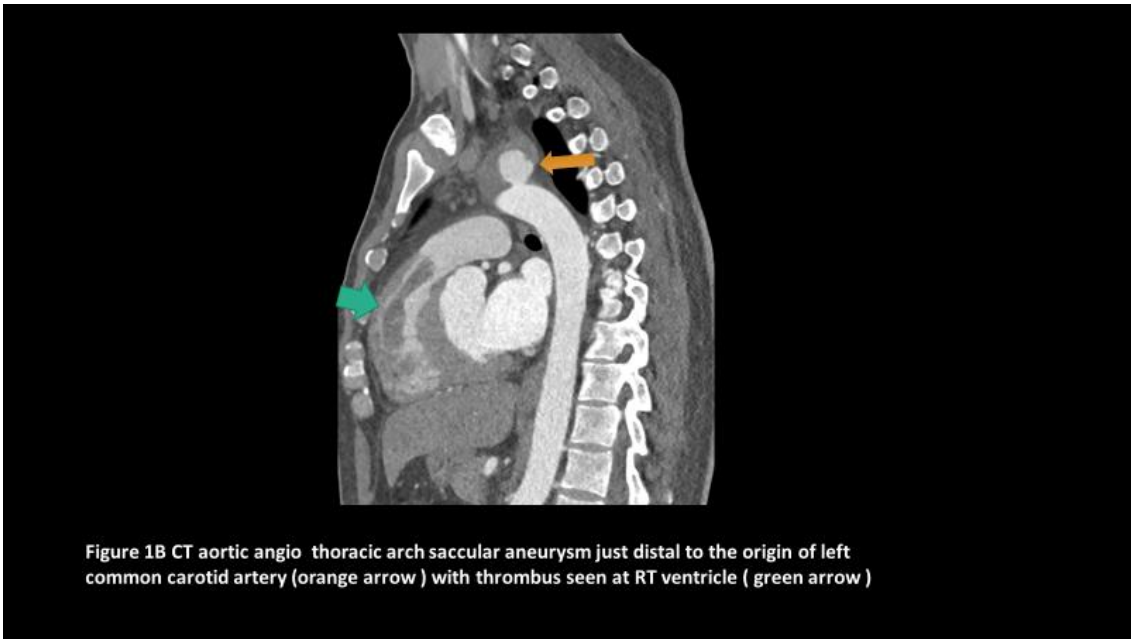


Figure 1B CT aortic angio thoracic arch saccular aneurysm just distal to the origin of left common carotid artery (orange arrow) with thrombus seen at RT ventricle (green arrow)

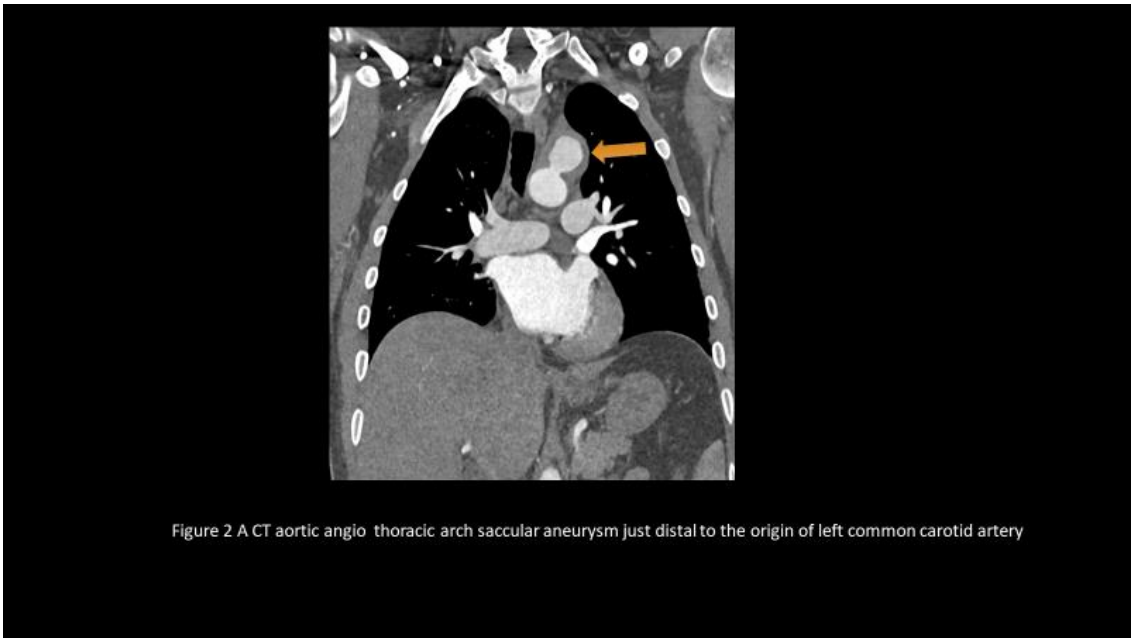
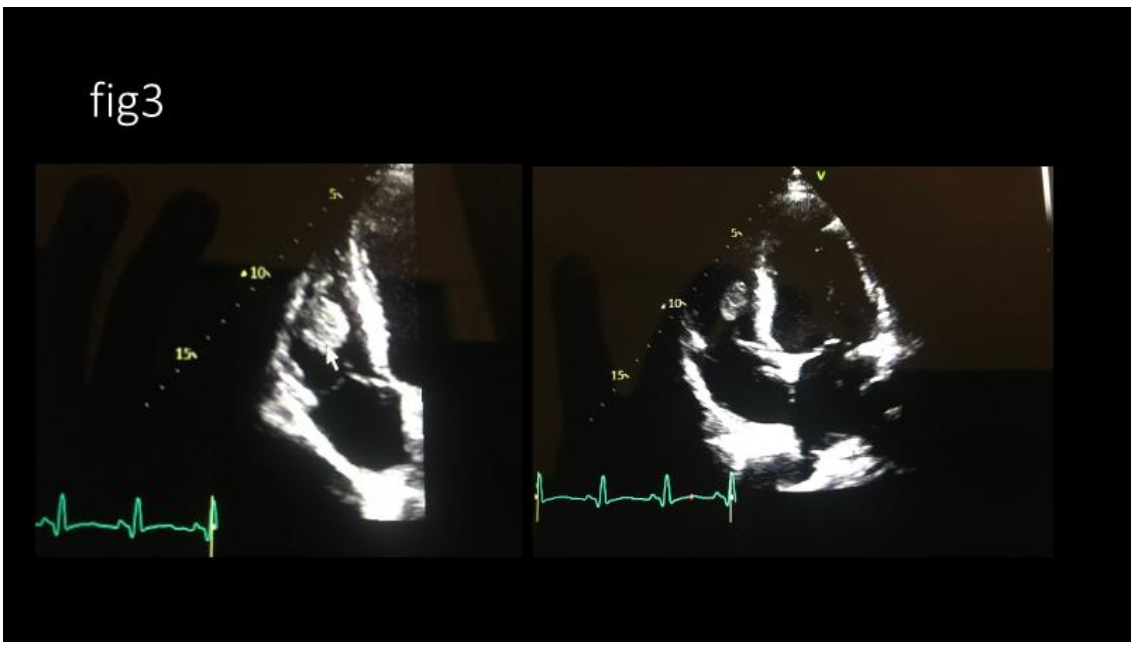


Figure 2 A CT aortic angio thoracic arch saccular aneurysm just distal to the origin of left common carotid artery

Figure 2 B CT aortic angio 3D volume rendering thoracic arch saccular aneurysm just distal to the origin of left common carotid artery



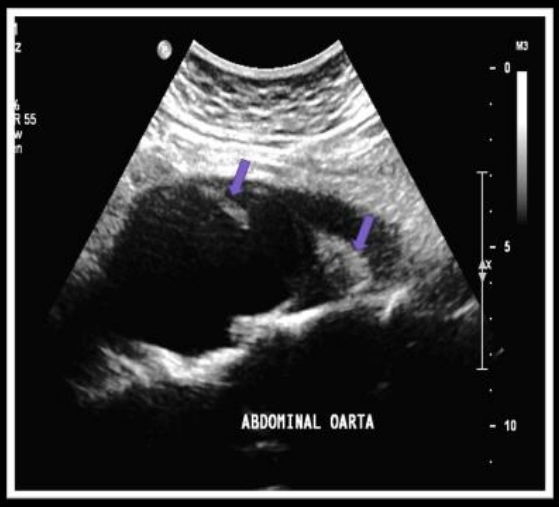


Figure 4 abdominal ultrasound fusiform abdominal aortic aneurysm below the origin of renal arteries with eccentric mural thrombus (arrows)

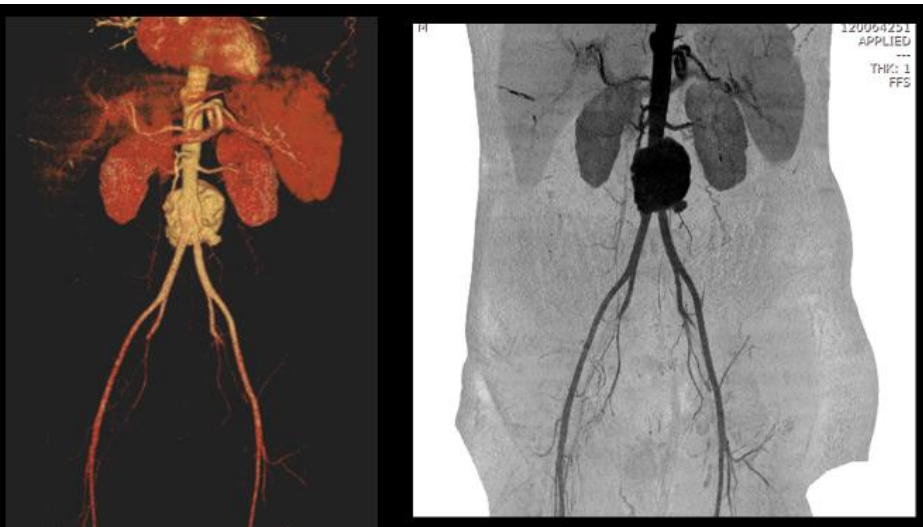


Figure 5 A CT aortic angio 3D rendering large saccular wide neck aneurysmal dilatation of the abdominal aorta originate distal to the origin of the renal arteries.

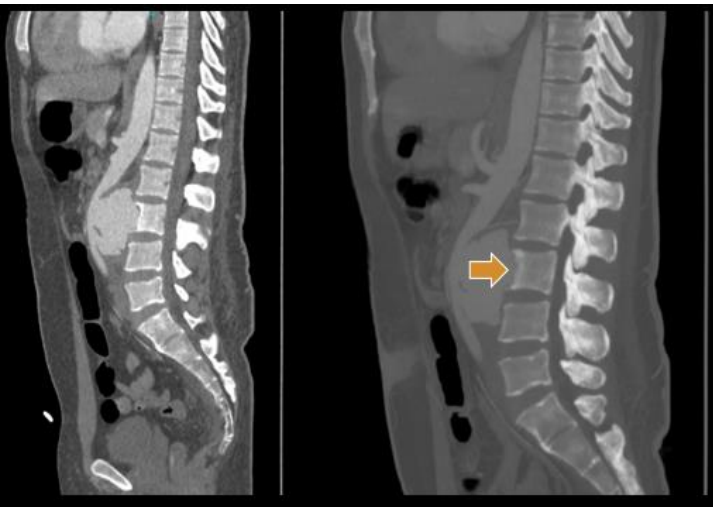


Figure 5 B CT aortic angio large saccular wide neck aneurysmal dilatation of the abdominal aorta associated with a mural thrombus and surrounding hematoma originate distal to the origin of the renal arteries. causing anterior scalloping of L3 vertebra at bone window figure (arrow)

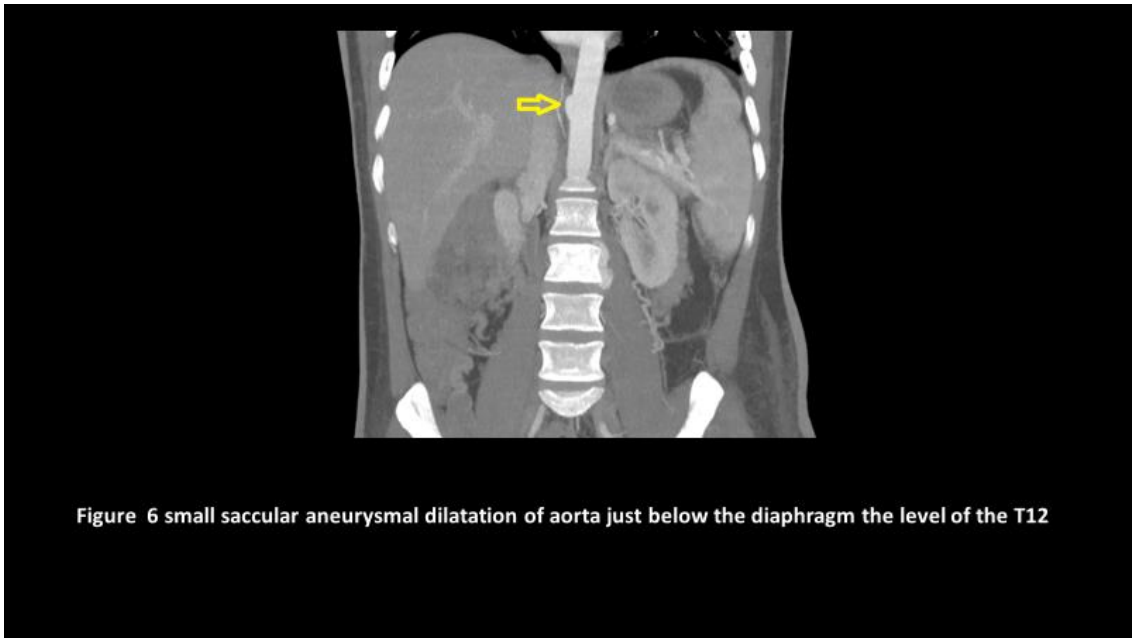


Figure 6 small saccular aneurysmal dilatation of aorta just below the diaphragm the level of the T12

Figure 7 CT lower limb angio complete occlusion of left popliteal artery with refilling of anterior and posterior tibial arteries by collaterals .

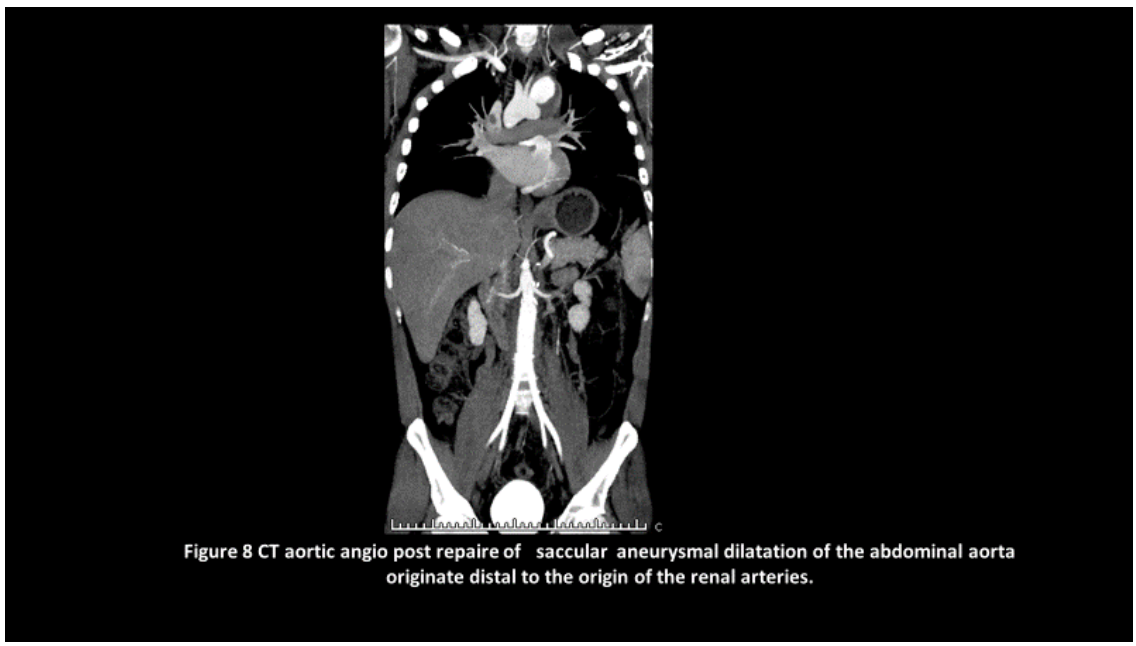


Figure 8 CT aortic angio post repaire of saccular aneurysmal dilatation of the abdominal aorta originate distal to the origin of the renal arteries.

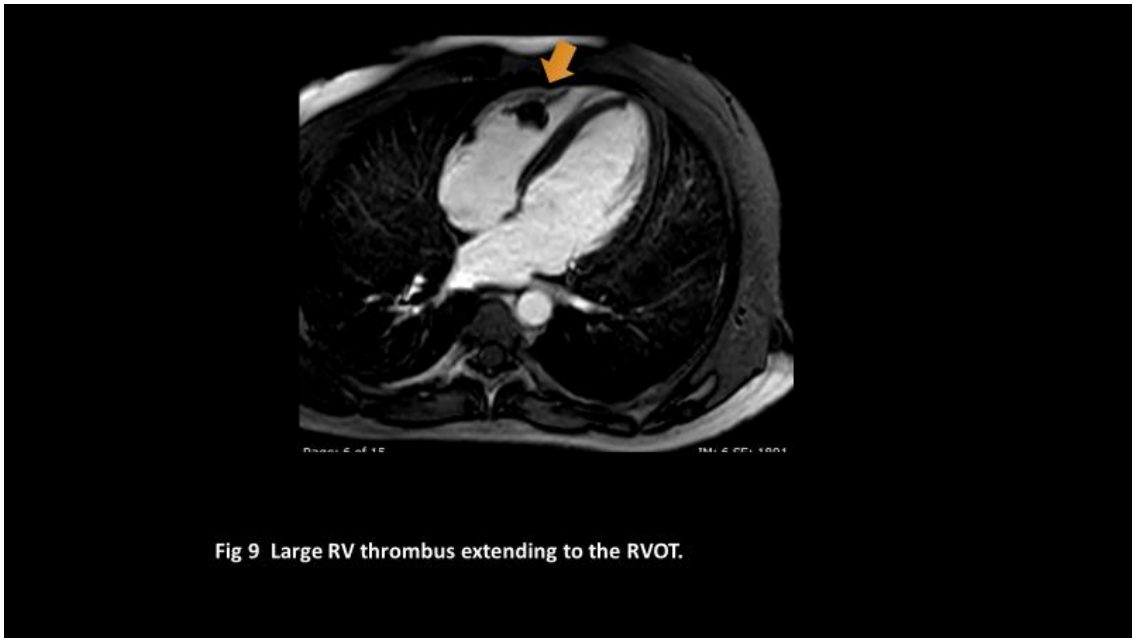


Fig 9 Large RV thrombus extending to the RVOT.

Laryngeal Fiberoptic examination revealed immobility of left vocal cord which was attributed to left subclavian artery aneurysm pressing on the left recurrent laryngeal nerve. A multi-disciplinary meeting was organized among cardiothoracic surgeon, vascular surgeon, rheumatologist, a cardiologist, and surgical intensivist. It was decided that CT whole aorta and its branches be done to assess the arch aneurysm anatomy & to assess the patency of other vessels. Transthoracic echo and CT coronary were planned to identify the cause of ejection fraction drop and to rule out any vasculitis that may include the coronaries. The rheumatologist decided to start the patient on high dose steroids, which was objected to by the vascular surgery team fairing infection at the site of the abdominal aortic aneurysm repair. However, the patient received 2 doses of 60 mg prednisone before it has been discontinued by the vascular surgical team. Heart failure medicines were started based on low EF and CT coronary Angio to rule out Coronary artery disease was planned followed by MRI heart (Fig 9).

In the meantime, ophthalmology opinion was sought to look for eye signs of Behcet's and it was negative. The CT coronary angiogram excluded coronary artery disease. CT aorta revealed saccular aneurysmal dilatation of the origin of the left subclavian artery with partial thrombus. A Pulmonary embolism, pulmonary vein thrombus, and deep vein thrombosis in common iliac veins on both sides were noted. A thrombus in the right ventricular cavity was seen. The Aortic wall biopsy from the resected abdominal aortic aneurysm revealed necrotic blood vessel wall and paraaortic lymph node was reactive with negative tissue for granulomas or malignancy.

Because of the aneurysmal aortic disease associated with widespread thrombosis and the age and origin of the patient, our final diagnosis was Behcet's disease. Five days post his AAA repair the prednisolone 60 mg daily was restarted with good symptomatic response and drop in his inflammatory markers. During this period patient developed a new complaint of weakness in the left hand and fingers along with left foot drop. Imaging of the brain and neck by MRI did not show any evidence of acute pathology. A nerve conduction study for the left leg revealed left peroneal axonal neuropathy only without a picture of mononeuropathy multiplex. We considered the vasculitis to be the aetiology for peroneal neuropathy and nerve biopsy was planned. To rule out secondary hypercoagulable state we did the Thrombophilia workup, which showed low Protein S at 31 (72-

126) and heterozygous for c.*97G>A (old name: G20210A).

Paraneoplastic coagulopathy was also in our differentials. However, clinical examination, other blood tests, and then the PET scan revealed no evidence for that. Urine analysis and serum complement levels (C3, C4) were all within normal limits. Antinuclear antibody, double stranded-DNA, anticardiolipin antibodies, beta-2 glycoprotein, and anti-neutrophile cytoplasmic antibodies (c-ANCA and p-ANCA) were all negative. Treponema pallidum antibody and QuantiFERON were negative. Interestingly, the HLA-B51 was absent. MRI heart was unremarkable for cardiomyopathy and the plan was to follow up with repeat echo. The patient was discharged on 18.02.2020 with the diagnosis of inflammatory thrombotic vasculitis in Behcet's disease. He continued warfarin with 30 mg of a tapering dose of prednisolone, Lisinopril 5mg daily and Metoprolol 25 mg bid, Esomeprazole 40 mg daily. He responded very well to that with normalization of his Hb, ESR, and CRP. He continued to have normal CRP till he follows up blood test on 20.05.2020.

He has been reviewed on 25.02.202 in the rheumatology outpatients' clinic patient when he has been started on methotrexate 15 mg a week with the folic acid 10 mg a week, two days after the MTX dose. He has been advised to continue on the same dose of the prednisolone at 30 mg daily for another 2 weeks, then to be dropped to 20 mg daily after that. A month later he has been reviewed again while he was on 20 mg prednisolone reducing the dose, his ESR risen from 17 to 32 and his CRP from 4 to 12 and found to have a low INR of 1.5 and the warfarin increased with the aim for INR target of 2.5 and Vit-D started (his Vit-D was 23ng/ml). His MTX increased to 20 mg weekly.

On 15.07.2020 he attended ED with SOB and conformed to have another PE with INR of 1.4 which is subtherapeutic and high CRP of 27 mg/L. Previous CRP on 07.06.2020 was 20 mg/L. Gated CT thorax on 16.07.2020 revealed the thoracic saccular aneurysm size was stable at 50*47 mm. His warfarin increased with target INR between 2.0 and 3.0.

Then he has been reviewed by the rheumatology team on 31.08.2020, clinically he feels reasonably well with no complaints. He was on MTX 20 mg weekly with the folic acid and on prednisolone 10mg daily for the last month. Unfortunately, he has no repeat blood test done before his clinic visit as has been previously planned. He has been disadvantaged by the COVID-19 pandemic situation in the way of interruptions of his follow-ups and the blood testing as he has difficulties in attending for routine blood testing either in the hospital or the primary care centre. He has had few telephone consultations, in which he said he feels fine, but unfortunately, no blood test has been done.

Then he attended the ED on 30.10.2020 with acute compression-like retrosternal central chest pain for the last 3 hours. BP 110/76mmHg. CRP 32, ESR 52, INR 3.0. He was on Methotrexate 20mg weekly, prednisolone 10mg daily, and warfarin. ECG normal, CT coronary revealed no evidence of coronary artery disease, ECHO revealed improved LVEF to 45%, RVEF to 53%, RV thrombus 2*3.4 cm. C-MRI scan confirmed the above. The rheumatologist increased his prednisolone to 30 mg on 01.11.2020 and his CRP dropped to 4 and ESR to 23 by 04.11.2020. He has had his last phone call consultation a week before this ED consultation when he said that he feels well, but unfortunately, has not done the blood as requested before this consultation, and he has been given another appointment in 2 weeks with the repeat blood before the clinic.

Gated CT thorax revealed that there is significant enlargement in the saccular thoracic aneurysm from 50*47 mm to 63*54 mm with surrounding thrombosed hematoma. The previously seen sessile outpouching of the thoracic aorta at the level of the diaphragmatic hiatus is noted with no significant interval change. The infrarenal aorta graft revealed associated tissue thickening with no significant

dilatation or new aneurysm. The RV thrombus is 6.7*3.3 cm.

Through a discussion with the vascular team, the patient is not suitable for stenting because of his active generalized vasculitis, previous AAA repair & short landing zones. Accordingly, the patient was stabilized with the normalization of his inflammatory markers which responded very well as before to increase prednisolone to 30 mg daily. Then he underwent repair of aortic arch aneurysm through sternotomy on 05.11.2020. During the aneurysm repair mediastinal lymph node, aortic arch aneurysm, and thymus biopsies were taken and the following results were obtained. Mediastinal lymph nodes: four reactive lymph nodes, no evidence of malignancy. Aortic arch aneurysm: degenerative changes with fibrosis and aggregates of hemosiderin-laden macrophages. Thymus: rare lymphoid thymic tissue and prominent adipose tissue in keeping with thymic involution.

He is followed closely by Rheumatology post aneurysm repair and Azathioprine 50 mg daily and Golimumab 50 mg subcutaneous monthly has been added to his medications including the prednisolone 30 mg daily, Lisinopril 5 mg daily, Metoprolol 25 mg bid and Ferrous Sulfate bid. Methotrexate has been discontinued as it was ineffective.

Then he has been reviewed 2 weeks later and he admitted that he is feeling much better with at least 90% improvement following the first injection of his Golimumab. His most recent blood on 29.11.2020 revealed: Hb 13 (was 8.8), ESR 23, CRP 8.9, normal renal and liver functions, BP right arm 118/74 and the left arm 99/66, PR 87. DXA scan: T-score LS -1.8 and Hip -1.7. Accordingly, he has been given one shot of IV Zoledronic acid 5mg.

He has been reviewed again in mid-January 2021 and as soon we reduced the prednisolone below 20 mg, his CRP started to raise again despite that he is feeling well. This is the same as it happens before his previous two flares; with the second PE and then the thoracic aortic aneurysm expansion, and despite the fact that his Azathioprine has been increased to 100 mg and he received 3 doses of Golimumab. His case has been discussed with few international Behcet's disease expert, who all agreed to give him IV Cyclophosphamide, which we are planning to give over next few days.

To summarize the case, this was thrombotic aneurysmal vasculitis due to Bechet's disease in a young patient from the Mediterranean origin with no other classical signs of Bechet's such as the mouth or genital ulceration or eye involvement. His main presentation is inflammatory aneurysmal aortitis with inflammatory widespread thrombosis including the intracardiac involvement. He needed an urgent repair for his leaking infrarenal saccular aortic aneurysm and then for his expanding thoracic saccular aneurysm which we think is related to his uncontrolled active disease. He also has another PE which we think is inflammatory and also due to his active uncontrolled disease. Then an expanding thoracic saccular aneurysmal dilation of origin of left subclavian artery led to his hoarseness of voice. Popliteal artery occlusion and bilateral DVTs and left peroneal axonal neuropathy were its other manifestations.

Discussion

Bechet's disease (BD) is a chronic relapsing systemic inflammatory disease involving mucous membranes, skin, eyes, the gastro-intestinal tract, joints, blood vessels, and the neurological system. It is more prevalent in the Middle East and Mediterranean regions. It is also distributed in the Central and Far Eastern Asian countries including Korea, Japan, and China [8,9,10,11]. The highest prevalence was reported in Turkey 20-420 per 100,000 people. The prevalence is now increasing in other parts of the world especially in the eastern Mediterranean and eastern Asia (30 per 100,000) and in North America (1 per 15,000) because of increased migration [12].

Diagnostic criteria published in 1990 by the International Study Group for Behcet's Disease [13] are still in use. These criteria are oral

ulcerations with at least two of the following: recurrent genital ulcerations, typical eye lesions, typical skin lesions, or a positive pathergy test (a sterile pustule developing after 24 to 48 hours at the site of a cutaneous needle prick).

The HLA-B51 is frequent in BD patients, with a range of 40-80% in ethnic groups including Turkish, European, and Asian populations from the Middle East to the Far East, whereas it can be as low as 13% among white patients in western countries [14]. Al Dalaan et al, found that 72% of the 85 Saudi patients with Behcet's disease had positive HLA-B5(51) compared with 26% general population [15]. It is interesting to note that HLA-B51 was not detected in our patient. The sporadic familial clustering that is typical of BD favours the use of B51 as a marker of disease severity [16].

BD is the only systemic vasculitis involving both arteries and veins of any size [10,11]. Vascular involvement is characterized by aneurysm formation and occlusion of large vessels. Most authors believed that venous manifestation is more common in BD [10]. 14 to 40% of BD patients are affected with venous complications making it the most frequent vascular complication. Superficial and deep lower limb thrombosis is the most common presentation of venous manifestations, however, one-third of venous thrombosis occurs in the large vessels (such as cerebral venous thrombosis, pulmonary embolism, and inferior or superior vena cava). Budd Chiari syndrome is known to be the worst prognostic factor which increases mortality by 9 times [16,11].

Although arterial involvement is rare, it can complicate the clinical picture and cause potentially fatal consequences [8-9]. The first description of arterial involvement in Behcet's disease was of an abdominal aortic aneurysm in a Japanese man. Since that time there have been sporadic reports of occlusions and aneurysms of several major arteries, as a result of Behcet's disease. Approximately 7% of patients with Behcet's disease will have serious vascular complications [16].

According to Hamza classification, there are five types of vascular involvements in BD, which include venous thrombosis, arterial thrombosis, arterial aneurysm, arterial thrombosis and aneurysm, and venous thrombosis and arterial lesion [10]. Our patient had venous thrombosis and arterial lesions which constitute class five according to this classification. The abdominal aorta is the most common site of aneurysm formation followed in decreasing order by the pulmonary, femoral, popliteal, brachial, and iliac arteries. Rare cases involving the carotid, vertebral, coronary, and visceral arteries have also been reported.

Aneurysms are usually saccular and show curvilinear enhancing, thickened walls, with partial thrombosis as occurred typically in our patient. Inflammatory cell infiltration leads to occlusion of the vasa vasorum which leads to transmural necrosis of the wall of large muscular arteries. Saccular aneurysms are probably produced by severe destruction of elastic fibres in the media and consequent perforation of the vessel wall. We think that endothelial damage leads to hypercoagulability and thrombosis, and abnormal flow and aneurysmal formation. The precise pathogenetic mechanism of the prothrombotic state in BD is unknown. It appears that thrombophilic factors could contribute to thrombosis in BD [10]. Thrombophilia workup in our patient showed low protein S and heterozygous for c*97G>A.

Few cases with intracardiac thrombus which often preceded other manifestations of BD have been reported [17]. These thrombi found mainly in the right ventricle are often associated with pulmonary artery aneurysm. In a study by Ben Ghrobe among 518 patients with BD, 8 were diagnosed as having intracardiac thrombus (ICT). All were male with the mean age at the time of ICT diagnosis as 30.8 years as in our case. All patients fulfilled the diagnostic criteria of the International Study Group of Behcet's Disease [12-18], Which is not the scenario in our case. The reason for predilection to involve the right ventricle is still unclear. However, both

ventricular involvements have also been reported [17]. Intracardiac thrombus can lead to superior vena cava syndrome and pulmonary embolism [10].

The diagnosis of BD should be considered if a patient presents with a mass in the right-sided cardiac chambers even in the absence of the characteristic clinical manifestations of the illness. This approach is particularly applicable if the patient is a young man from the Mediterranean or the Middle East. All Behcet's patients with ICT must be investigated with thoracic computed tomography for pulmonary and arterial involvements and lower extremity venous doppler ultrasonography for venous thrombosis regardless of whether they are symptomatic for these systems [7].

Hakan Emmungi studied 22 cases with ICT, out of which cardiac involvement was found to be the first clinical manifestation of BD in 9 out of 22 patients. All patients fulfilled three or more of the International Study Group Criteria for BD [18]. After treatment, the intracardiac thrombus disappeared in 13 cases and the size of the thrombus reduced in 7 cases [13-18]. Our Patient had repeat echocardiography after ten months of initial presentation and it did not show any significant change in the size of the thrombus. We think that this is due to continuing disease activity, as our patient failed to respond to the MTX on reducing his steroid dose. And as we know the thrombosis in such patients is inflammatory in nature and good control of the disease activity by intensive immune-suppression is essential and may contribute with the anticoagulation to dissolve the thrombus, if acted on quickly in the acute stage. We think that the inability to achieve good control of the disease was also the reason for the recent expansion of his thoracic aneurysm and before that contributed to the development of another PE.

Our patient was unfortunate as his disease occurred in the era of the COVID-19 pandemic. He missed some of his appointments and has not done the required blood tests on a few occasions as access to the hospital is limited during this period.

We suggest that such patients with Arterial, venous, cardiac involvement and neuro involvement should be treated aggressively with immunesuppressants to control their disease. Possibly the early use of iv cyclophosphamide or TNF-a blockers will help to prevent his complications.

Early recognition of vascular lesions in BD is important as it is the most common cause of death. The prognosis with arterial occlusions is better than that with aneurysmal manifestations in patients with BD [9] with a mortality of about 20% in severe cases. Delay in diagnosis is frequently cited as a major contributing factor leading to a fatal outcome [16].

BD can be manifested as hoarseness and vocal cord ulcers could be the first signs [19]. It was observed that our patient had hoarseness of voice upon presentation. Laryngeal Fiberoptic examination showed left vocal cord paralysis, which was attributed to a left subclavian artery aneurysm pressing on the left recurrent laryngeal nerve.

Central nervous system (CNS) involvement is common in BD. And about 10% of BD patients have CNS problems. Peripheral nervous system involvement, however, is very rare in this disease [20]. Our patient developed a left foot drop during hospitalization. Nerve conduction study for left leg diagnosed left peroneal axonal neuropathy. Vasculitis was considered to be the aetiology for peroneal neuropathy.

In Summary, we present this case of BD to raise the awareness that BD could have such a presentation of both venous and arterial lesions with the lacking of the typical clinical features of BD. However, the patient's typical venous and aneurysmal arterial disease with his age and origin raised our suspicion of BD. The other learning point is that we have to stress that such thrombotic events in BD were all inflammatory and should be treated with aggressive immunosuppression rather than relying on

anticoagulation which ideally should be of moderate intensity and for short period only. Intensive anticoagulation may lead to hemorrhagic events in the aneurysmal lesions. Close supervision is mandatory in such patients with thrombosis and arterial aneurysmal disease as quick and strict control of the disease activity is very essential in reducing morbidity and mortality in such patients.

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal. The authors declare that they have no competing interests.

Ethics statement: This work was approved by Medical Research Center (MRC) Qatar before submission.325742-- ABHATH - MRC (HMC)

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