CASE REPORT

Ruptured innominate artery pseudoaneurysm presenting as hoarseness in Behçet’s syndrome: a case report

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Abstract

Background: Vascular involvement is an infrequent clinical manifestation of Behçet’s syndrome. Owing to the rarity of arterial involvement in Behçet’s syndrome, there is limited experience in managing this phenomenon.

Case presentation: Here, we report a 28-year-old Iranian man with a Behçet’s syndrome background, who presented with shoulder pain and hoarseness. Chest computed tomography angiography was conducted with a suspicion of a vascular pathology causing pressure on the recurrent laryngeal nerves. The patient was diagnosed with a ruptured innominate artery pseudoaneurysm. An innominate artery to the right common carotid artery bypass was performed, and the pseudoaneurysm was excised and replaced with an expandable polytetrafluoroethylene graft. Eventually, the patient was discharged after an uneventful hospital course.

Conclusion: It appears that we are still a long way from finding the optimal treatment for Behçet’s syndrome vascular involvement, and a combination of surgical and medicinal treatments is required.

Keywords: Innominate artery, Behçet’s syndrome, Pseudoaneurysm

Background

Behçet’s syndrome (BS), as a variable vessel vasculitis, is more common in the Silk Road countries than in other parts of the world. Clinical presentations vary diversely depending on which organs are involved. In contrast to cutaneous and mucosal involvement, vasculitis is relatively rare. The primary form of vasculitis is venous involvement, particularly deep vein thrombosis [1]. On the other hand, in arterial vasculitis of BS, pulmonary artery aneurysms, thrombosis, and extrapulmonary arterial aneurysms/pseudoaneurysms have been reported [2]. Overall, innominate artery aneurysm is a rare condition comprising about 3% of all supra-aortic aneurysms [3].

In this article, we describe an extremely rare presentation of BS and further share our experience in the surgical management of this case.

Case presentation

The patient was a 28-year-old Iranian man with a history of BS, who presented with right shoulder pain and hoarseness 1 week prior to admission. Going through his medical records, we realized that he had undergone an open right subclavian artery bypass for acute right upper extremity ischemia caused by thrombosis in the aneurysmal right subclavian artery to the distal arterial branches around 1.5 years ago. However, follow-up imaging suggested occlusion in the bypass area. Given the history of recurrent oral aphthous lesions and extensive body folliculitis lesions, BS was suspected. The BS diagnosis was

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confirmed by a positive human leukocyte antigen (HLA)-
B*51 test, an erythrocyte sedimentation rate (ESR) of
79, and a C-reactive protein (CRP) of 5.2 (normal: 0.5),
as well as pathological findings from the open right sub-
clavian artery bypass surgery. Cyclophosphamide therapy
was the treatment of choice for the patient, but due to
infertility concerns, he refused. Therefore, he was treated
with azathioprine 100 mg daily, prednisolone 30 mg
daily, colchicine 1 mg daily, and rivaroxaban 20 mg daily
postoperatively.

The patient’s compliance was good, and according to
the latest rheumatological reports prior to his admission
to our facility, his BS was under control and no hyperco-
agulable symptoms were observed.

After admitting the patient, with all his past history
in mind, we requested a contrast-enhanced chest com-
puted tomography angiography (CTA) with suspicion
of a vascular pathology causing pressure effect on the
recurrent laryngeal nerves. CTA revealed a $32 \times 27$ mm
ruptured pseudoaneurysm originating from innominate
artery bifurcation with a $90 \times 61 \times 65$ mm hema-
toma extending to the right shoulder and forming an
extrapleural hematoma at the apex of the right thoracic
cavity (Fig. 1). The trachea was deviated to the left side
due to the mass effect of the pseudoaneurysm. Despite
total occlusion of the right subclavian and axillary
arteries by thrombosis, the patient had no complaint
regarding his right upper extremity, which shows that
there was probably sufficient collateral blood perfusion.
Even though we retrospectively reexamined his previ-
ous CTA from the former admission, no anomaly was
found in the innominate artery. We decided to repair
the ruptured pseudoaneurysm by an endovascular
approach. Preoperatively, the patient received methyl-
prednisolone to decrease postoperative complications.
In a hybrid operation room, we performed a digital
subtraction angiography. Unfortunately, owing to the
lack of proper landing zone, we could not perform

![Fig. 1 Preoperative contrast-enhanced computed tomography angiogram (CTA).](image-url)

a Axial image: the pseudoaneurysm of the innominate artery causing pressure effect on the trachea (black arrow).
b Axial image: the ruptured pseudoaneurysm hematoma (white arrow).
c Coronal reconstruction: a large hematoma is observed in the right shoulder extending to the extrapleural area in the apex of the right thoracic cavity (white arrow).
d Coronal reconstruction: innominate artery pseudoaneurysm is observed (white arrow). It must be noted that the right subclavian artery is occulted.
an endovascular repair by stent graft. Inevitably, we approached the pseudoaneurysm by a median sternotomy with supraclavicular extension. After excising the ruptured pseudoaneurysm and evacuating the surrounding hematoma, we performed an innominate artery to the right common carotid artery bypass with a 6 mm expandable polytetrafluoroethylene (ePTFE) graft, which passed right behind the innominate vein (Fig. 2). The pressure effect of the pseudoaneurysm was removed from the right recurrent laryngeal, phrenic, and vagus nerves with no neural damage. We decided not to perform a right subclavian artery bypass since it was previously thrombosed, and the patient had no complaint concerning the right upper extremity. Postoperatively, the patient developed a deep vein thrombosis at the central venous catheter site in the left internal jugular vein. Later, the patient was transferred to the rheumatology ward. The hoarseness of the patient resolved through the hospital course. Monthly infusion of cyclophosphamide 750 mg was started, and the azathioprine was discontinued. Follow-up during the next year showed no sign of recurrence.

**Discussion and conclusions**

BS is a rare condition without a known etiology, which includes multisystem inflammatory presentations. In BS, vascular pathology is assumed to be induced by immunological and inflammatory mechanisms that generate a hypercoagulable state and endothelial damage. The vasculitis causes thrombosis by increasing platelet aggregation and inhibiting fibrinolysis [4]. In addition, vasculitis causes deterioration of the media layer, arterial dilatation, and fibrosis, which destroys the vasa vasorum and results in transmural necrosis and subsequent pseudo aneurysm formation or artery wall rupture [5].

Vascular involvement of BS, particularly arterial vasculitis, consists of aneurysm or pseudoaneurysm formation and thrombosis. Contrary to venous vasculitis, arterial involvement of BS usually requires surgical intervention besides high dose prednisolone and cyclophosphamide or anti-tumor necrosis factor(TNF)-α agents [6, 7]. Depending on the location of the arterial involvement, various interventions may be performed. However, the main problem with arterial vasculitis in BS is the recurrence of the condition after proper treatment. In a retrospective study of 25 cases of definite BS with arterial involvement by Tuzun et al., despite medical and surgical therapy, two cases developed aneurysms at new arterial sites after an average of 2 years, and three patients experienced aneurysmal recurrence in the surgical site 6 months postoperation [8]. Although our patient was medically managed to prevent new vascular pathology, he had developed a new pseudoaneurysm in the innominate artery.

There is still no consensus on whether the endovascular or open surgical approach is best in the arterial vasculitis of BS. Endovascular surgery offers a less invasive procedure with probably lower risks of procedure-related mortality. However, in a study by Kwon et al. including 12 BS patients with abdominal aortic aneurysm, it was concluded that treating the patients with graft interposition has a lower postoperative aneurysmal recurrence rate than the endovascular approach (14.3% versus 40%) [9].

Various surgical approaches have been practiced to manage innominate artery aneurysms, such as endovascular surgery and partial or full median sternotomy with or without anterior neck and supraclavicular fossa dissection [10].

There is limited experience in managing innominate artery aneurysms in BS patients. In a review of the literature, we only found two similar cases. The first description of this rare presentation of BS was in 1987 by Hamza et al. [11]. Furthermore, in 2016, Tsuda et al. presented a BS patient with an innominate artery aneurysm that underwent axillo-axillary bypass by PTFE graft
interposition. Later, an innominate artery stent graft was deployed through the right common carotid artery [12].

In our case, the subclavian artery was occluded, and the patient had no complaints regarding his right upper extremity. Hence, we avoided any attempt to reconstruct the right upper extremity circulation. Considering the rarity of this condition and the limited experience in managing this presentation, it is not clear whether endovascular or open surgery would be superior. However, the endovascular approach is preferred due to minimal invasiveness. In our case, proper stent graft was unavailable owing to the anatomy of the pseudoaneurysm. Short-interval follow-ups are strongly recommended looking for any signs of recurrence of the aneurysm. New signs and symptoms must be carefully examined for possible further vascular involvement. Hoarseness should be considered as a possible sign of vascular pathology in the mediastinum. Furthermore, the role of proper immunosuppressive medical therapy must not be underestimated. Recent studies show promising outcomes in patients who received immunosuppressive medical therapy before and after the surgical intervention [13].

In addition, there is still controversy regarding the use of anticoagulants in the treatment of BS. There is a concern that anticoagulants may increase the risk of aneurysmal bleeding and hematoma formation in BS patients [14]. In our case, it seems that using anticoagulants led to large hematoma formation after the rupture of the pseudoaneurysm. In BS, thrombosis usually occurs secondary to local endothelial activation due to vasculitis rather than a primitive coagulation disorder. These medications may help prevent thromboembolism formation at the aneurysmal site and prevent end arterial microembolization and gangrene [15, 16]. Therefore, while warfarin and direct oral anticoagulants are used to manage hypercoagulability in patients with BS, direct oral anticoagulants also appear to reduce the risk of major bleeding in routine practice[17, 18]. On the other hand, Vautier et al. suggested the use of direct oral anticoagulants in case of deep vein thrombosis (DVT) in BS due to the lower risk of bleeding, as it may also synergize with the rest of the medication, preventing further vascular pathologies [19]. Therefore, by considering all aspects, we chose to add rivaroxaban to the immunosuppressive medication.

Finally, we believe that effective therapy for vascular involvement in BS is still a long way off, and a combination of medicinal and surgical techniques is required. Moreover, additional research is required before the appropriate management of arterial vasculitis in BS can be determined.

The patient consented to the use of related medical history and imaging for educational and scientific purposes.

Abbreviations
BS: Behçet’s syndrome; HLA: Human leukocyte antigens; ESR: Erythrocyte sedimentation rate; CRP: C-reactive protein; CTA: Computed tomography angiography; DOA: Direct oral agents; DVT: Deep vein thrombosis; ePTFE: Expandable polytetrafluoroethylene.

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Author contributions
SAM and SSN contributed to the literature review, review of the patient’s information, and drafting of the manuscript. HGJ contributed to the patient’s surgery, review of the manuscript, and provision of guidance on the approach to the topic. AA contributed to the patient’s surgery and review of the manuscript. SAM, SSN, and SS contributed to the clinical evaluation of the patient and search of the databases. All authors have reviewed the manuscript, read and approved the final manuscript.

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Availability of data and materials
All data generated or analyzed during this experimental study are included in this published article.

Declarations

Ethics approval and consent to participate
The present study was approved by the Medical Ethics Committee of Shiraz University of Medical Sciences. The purpose of this report was completely explained to the patient and written informed consent was obtained from the patient.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

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