Percutaneous Treatment of a Behçet Patient with Subclavian Vein Thrombosis: Case Report

Bir Behçet Hastasında Subklavian Ven Trombozunun Perkütan Tedavisi

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Geliş Tarihi/*Received:* 08.01.2016
Kabul Tarihi/*Accepted:* 06.09.2016

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ÖZET Behçet hastalığı kronik tekrarlayıcı multisistemik bir hastalıktır. Sistemik tutulum nedeniyle oluşan vasküler tutulum %5-10 sıklığında gözlenir ve sıklıkla santral venleri tutarak alt ve üst ekstremite venöz trombozuna yol açar. Behçet hastalığındaki vaskülitik (geniş venlerin vasküliti) ve trombotik (koagülasyon ve fibrinolitik aktivitedeki anormallikler) bozukluklar venöz lezyonlar için iyi tanımlanmış predispozan faktörlerdir. Biz bu olgu raporunda daha önce Behçet tanısı bulunmayan ilk başvuru olarak sol subklaviyen ven trombozu nedeniyle endovasküler girişim uygulanan genç bir kadın hastadan bahsettik. Hasta öncelikle balon anjioplasti ile tedavi edildi. Bir ay sonra hasta total re-oklüzyon ile tekrar başvurdu. Altta yatan patoloji olarak Behçet hastalığı tanısı konuldu ve hasta stent ile tedavi edildi.

Anahtar Kelimeler: Behçet sendromu; endovasküler prosedürler

ABSTRACT Behçet disease is a chronic inflammatory disease which can affect multisystem and had recurrent course. Vascular involvement due to systemic vasculitis occurs in 5% to 10% of these patients and frequently affects central veins and most often results in venous thrombosis of the upper and lower extremity. The vasculitic (large vein vasculitis) and thrombotic (abnormalities of coagulation or fibrinolytic activity) disorders in Behçet disease are both well known as predispositions for venous lesions. We report a young female patient with previously undiagnosed of Behçet disease was treated with endovascular therapy because of left subclavian vein (SCV) thrombosis as first presentation. The patient was primarily treated with balloon angioplasty. One month later, the patient was re-admitted due to total re-occlusion of SCV. A self-expanding stent was applied and Behçet's disease was diagnosed as underlying pathology.

Key Words: Behcet syndrome; endovascular procedures

Turkiye Klinikleri J Cardiovasc Sci 2016;28(2):69-72

Behçet disease (BD) is a chronic relapsing systemic vasculitis.[1] BD can contributes to the development of central vascular occlusions particularly affecting venous system and leading to venous thrombosis and stenosis in central (superior vena cava, brachiocephalic, or subclavian) venous system.[2] The vasculitis (large vein) in addition to thrombosis is main factor in BD to cause vascular lesions.

Open surgery is considered the primary method of treatment for symptomatic systemic venous occlusions of nonmalignant etiology.[3] On the other hand, surgery has the disadvantage of requiring general anesthesia and car-

doi: 10.5336/cardiosci.2016-50082

ries the risk of pneumothorax or arterial and brachial plexus damage with the possibility of disrupting venous collaterals.[4] As an alternative treatment, endovascular approach can be used in central venous occlusions to maintain patency of veins.[5]

CASE REPORT

A 28-year old female was admitted to our clinic for evaluation of swelling in her facial and left upper extremity. Her laboratory tests were normal limits. Inflammatory markers including sedimentation and Hs-CRP were found normal range at initial admission. In her physical examination, she had acneiform nodules and swelling on her left arm and neck. Chest radiography was normal. Computed tomographic (CT) angiography was related with subtotal occlusions of proximal site of superior vena cava with thrombus near the origins of subclavian veins (Figure 1, left sided image). We started systemic anticoagulant therapy with enoxoparine and compressing the swelling with elastic bandages. We thought that all these complex findings were related to an underlying disease. After evaluation of the coagulative conditions of the patient and a rheumatologic consultation, patient was diagnosed with Behçet disease according to the International Study Group (ISG) criterias of Behçet's Disease.[1] Despite 5 days of treatment with systemic steroid and anticoagulant therapy, the swelling did not improve, and therefore we decided to diagnostic venous angiography and add interventional procedure if it is needed. After patient informed consent, A 6 F introducer was inserted to the right femoral vein and retrograde 6 F right judkins diagnostic catheter was inserted via left radial vein to identify the anatomy of the occlusion. The venous angiography showed subtotal occlusion of left subclavian vein (SCV) with costocervical collateral vessels to the superior vena cava (Figure 1, middle image). We decided to perform a balloon inflation to provide distal flow. A 6 F right judkins guiding catheter was changed with the diagnostic catheter and inserted via the left radial vein. A 4.0 x 60 mm and 6.0 x 40 mm (ATB advance, Cook Medical, USA) balloons were inflated to the lesion at 12 and 14 atm respectively. The control angiogram showed normal flow pattern to the superior vena cava (Figure 1, right sided image). Patient discharged with a systemic steroid and acetylsalicylic acid 100 mg.

After one month, the patient was readmitted with left arm swelling. The control venous angiography showed that left SCV was totally occluded with thrombus again (Figure 2, left sided image). We decided to perform repeated revascularization with stenting. After performed predilatation with a 4.5x20 mm balloon (invader, Alvimedica, Turkey) at 10 atm, a 8.0x135 mm peripheric (Misago, Terumo medical, Japan) self-expandable stent was implanted at 12 atm to the proximal site of left SCV (Figure 2, right sided image). Patient was discharged with coumadin 5 mg in addition to anti-

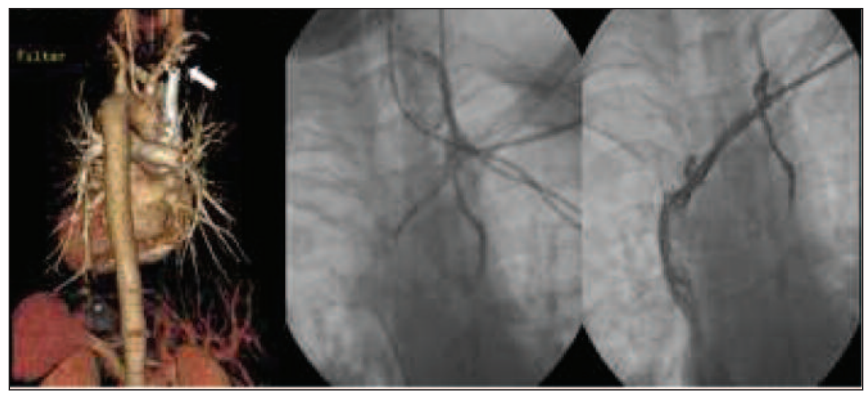

FIGURE 1: Subtotal occlusion of superior vena cava with thrombus (White arrow, left sided image), the contrast flow from axillary vein to the superior vena cava with collateral vessels and totally occluded and obliterated left subclavian vein (SCV) (middle image), and the control venous angiography of left SCV after balloon inflation (right sided image).

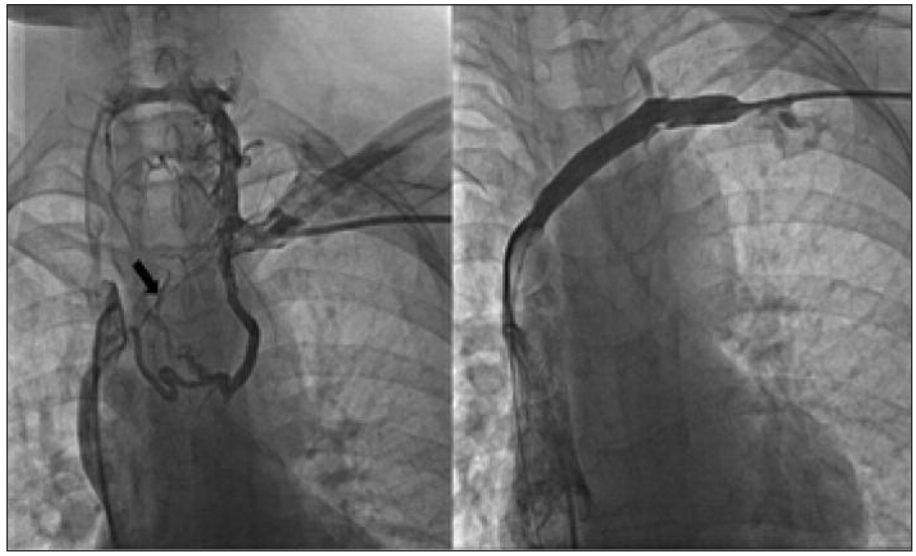

FIGURE 2: Left sided image show totally occluded left SCV(arrow) and right sided image show control imaging of SCV after stenting.

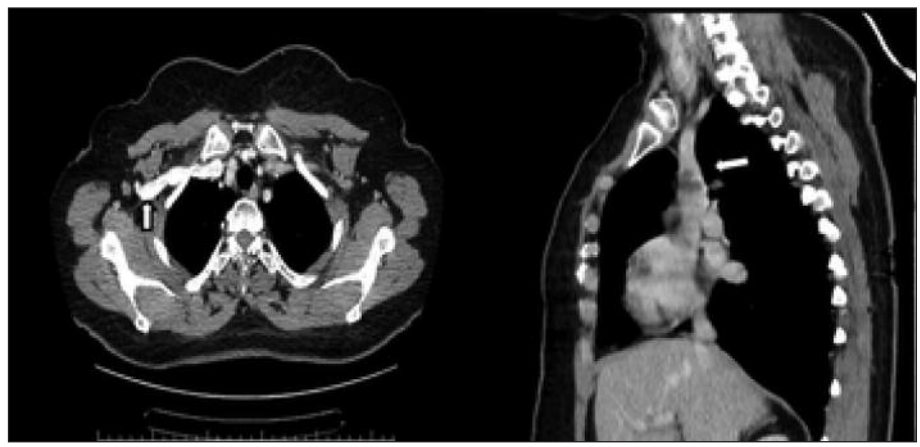

FIGURE 3: The control CT-A showed normal patency of left SCV (left arrow) and superior vena cava (right arrow).

inflammatory therapy. After six months, the patient was still asymptomatic and control CT angiography showed normal patency of superior vena cava and left SCV (Figure 3).

DISCUSSION

Behçet disease is a chronic inflammatory disease which can affect multisystem and had recurrent course.[1] Although most Behçet patients presents initially with mucocutaneous and ocular manifestations, vascular involvement may occurs approximately in 5% to 10%. For example, pulmonary artery involvement especially right pulmonary artery aneurysm can be seen in Behçet disease and the reported prevalence of pulmonary involvement has ranged from 1% to 7.7%.[6] On the other hand, our patient didn't have any symptom associated with pulmonary involvement and her contrast computed tomography findings were normal limits. Also Behçet is frequently leading venous thrombosis, when affects venous system of upper and lower extremities.[2] The characteristics of subclavian vein occlusions are swelling of the affected arm, venous engorgement, mild cyanosis and pain or discomfort.[4] Venography is ordinarily standard diagnostic method in venous occlusions and is sufficient diagnostic tool in most cases.[7] In our case, we performed venous angiography due to unsuc-

cessful result of anticoagulant therapy. Because this modality also allow endovascular intervention if needed.

There are no large trials comparing patients who have received surgery and those treated by endovascular approach in Behçet disease. Endovascular therapy with balloon or stenting for upper-extremity central venous occlusion has been found a safe approach and can be used as an alternative treatment method with immediate relief of the symptoms and prolonged duration of the response in venous thrombosis of Behçet disease.[8-10] Surgery is considered the primary method of treatment for symptomatic systemic venous occlusions. Although it has the disadvantage of requiring general anesthesia and carries the risk of pneumothorax or arterial and brachial plexus damage with the possibility of disrupting venous collaterals.[3] Also aneurismal reformation can be seen after open surgery in Behçet patients.

When intended endovascular intervention to the venous occlusions, it is important that the primary stenting is not preferred due to poor longterm outcomes.[11] On the other hand, residual stenosis may be developed more commonly with angioplasty than with stenting in such procedures especially when there is a underlying disease which leads to thrombo-embolism as in our patient.[8,9] Although anticoagulant therapy is mandatory at least first month after percutaneous treatment of venous occlusions, if the stenting was needed it may be prolonged at least 6 months, or longer if hypercoagulability is present.

In this case report, we try to imply another important point that our case is very rare in terms of clinical presentation. Because she was admitted to our clinic only the symptoms limited to vascular system. So when a patient presented with systemic venous thrombosis, physicians must be aware for coexisting pathophysiologies and take into account the differential diagnosis of all causes such as systemic vasculitis. It should be especially thought in patients under 40 years old and during differential diagnosis, the laboratory parameters associated with thrombotic disorders should be initially studied including antithrombin III, protein C and S deficiencies.

In conclusion, percutaneous transluminal angioplasty and stent procedure can be performed to maintain patency of central veins in Behçet vasculitis. Also the vascular pathologies may have indirect manifestations of Behçet disease and it should be kept in mind as a differential diagnosis of the noted problems.

REFERENCES

1. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet 1990;335(8697):1078-80.
2. Kuzu MA, Ozaslan C, Köksoy C, Gürler A, Tüzüner A. Vascular involvement in Behcet's disease: 8-year audit. World J Surg 1994; 18(6):948-53.
3. Kalra M, Gloviczki P, Andrews JC, Cherry KJ Jr, Bower TC, Panneton JM, et al. Open surgical and endovascular treatment of superior vena cava syndrome caused by non-malignant disease. J Vasc Surg 2003;38(2):215-23.
4. Rochester JR, Beard JD. Acute management of subclavian vein thrombosis. Br J Surg 1995;82(4):433-4.
5. Tekbas G, Oguzkurt L, Gur S, Onder H, Andic C. Endovascular treatment of veno-occlusive Behcet's disease. Cardiovasc Intervent Radiol 2012;35(4):826-31.
6. Erkan F. Pulmonary involvement in Behçet disease. Curr Opin Pulm Med 1999;5(5):314-8.
7. Chengelis DL, Glover JL, Bendick P, Ellwood R, Kirsch M, Fornatoro D. The use of intravascular ultrasound in the management of thoracic outlet syndrome. Am Surg 1994; 60(8):592-6.
8. Glanz S, Gordon DH, Lipkowitz GS, Butt KM, Hong J, Sclafani SJ. Axillary and subclavian vein stenosis: percutaneous angioplasty. Radiology 1988;168(2):371-3.
9. Lumsden AB, MacDonald MJ, Isiklar H, Martin LG, Kikeri D, Harker LA, et al. Central venous stenosis in the hemodialysis patient: incidence and efficacy of endovascular treatment. Cardiovasc Surg 1997;5(5):504-9.
10. Bakken AM, Protack CD, Saad WE, Lee DE, Waldman DL, Davies MG. Long-term outcomes of primary angioplasty and primary stenting of central venous stenosis in hemodialysis patients. J Vasc Surg 2007;45(4): 776-83.
11. García Mónaco R, Bertoni H, Pallota G, Lastiri R, Varela M, Beveraggi EM, et al. Use of self-expanding vascular endoprostheses in superior vena cava syndrome. Eur J Cardiothorac Surg 2003;24(2):208-11.