

Behcet's Disease with Vascular Complications

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Summary

A young gentleman presented with a history of multiple pseudoaneurysms of the right carotid artery, left radial artery, right femoral artery as well as deep vein thrombosis involving the right femoral vein. A diagnosis of Behcet's disease was made on the basis of his history of recurrent oral and genital ulceration with characteristic eye lesion.

Key Words: *Behcet's disease, Pseudoaneurysms, Deep vein thrombosis*

Introduction

Behcet's syndrome is a recurring illness characterized by the triple symptom complex of aphthous stomatitis, genital ulcerations and uveitis. This entity was first reported by Hulusi Behcet, in 1937 and has been most commonly encountered around the Mediterranean, Middle East and Japan. In 1990 the International Study Group (ISG) for Behcet's disease¹ recommended a new set of diagnostic criteria, which requires the presence of oral ulceration plus any two of the following; genital ulceration, typical defined eye lesions, typical defined skin lesions or a positive pathergy test.

Behcet's disease can be divided into three subtypes: neuro-, entero-, and vasculo- Behcet's disease. The major manifestations of vasculo-Behcet's disease are aneurysm formation, arterial occlusion and venous occlusion.

Case History

This patient first presented to us with a right carotid artery pseudoaneurysm for which a repair was done with an autologous vein patch (Figure 1). There was no prior history of trauma. Two months later he developed left radial artery pseudoaneurysm from his arterial line site, which was ligated. Subsequently he presented

again with bleeding from a recurrent pseudoaneurysm over the earlier patch repair of his right carotid artery. A decision was made to ligate the right common carotid, internal carotid and external carotid artery. There was no neurological deficit postoperatively.

During that admission, he was also noted to have multiple ulcers over the penis as well as oral cavity. He admitted to have sexual exposure with multiple partners. The biochemical investigations revealed a high ESR and C-reactive protein but screening for syphilis, HIV, hepatitis virus and connective tissue disease were negative. He was treated for his genital and oral ulcers and discharged home with antibiotics.

Two weeks later he complaint of pulsatile right groin swelling at the site of punctured wound from a previous angiogram and associated swelling of the right lower limb. An ultrasound colour doppler and angiogram examination revealed a pseudoaneurysm of the right common femoral artery and thrombosis of the right common femoral vein. At this time, referral to the ophthalmology unit confirmed the presence of uveitis.

Ligation and debridement of the right common femoral artery was done to manage the recent pseudoaneurysm. The histopathological report of the diseased artery revealed that clusters of lymphocytes,

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neutrophils as well as plasma cells infiltrated the adventitial layer. The intima and medial layer showed destruction and some areas covered by fibrin, fibroblastic proliferation and thick collagen bundles. These features are consistent with vasculitis and compatible with Behcet's disease.

Oral prednisolone was started at 25mg twice daily. He responded well to immunosuppressive therapy and C-reactive protein levels was back to normal. His usual stigmata of Behcet's disease of oral ulcer, genital ulcer and uveitis were in remission and he was discharged to a peripheral hospital for follow-up.

Discussion

There have been several report of multiple aneurysms appearing in single patient with Behcet's diseases^{2,3}. Arterial complications occur in approximately 7% of cases, with true/false aneurysm of the abdominal aorta being most common, followed by femoral and pulmonary arteries. Venous thrombosis occurs in approximately one third of patients with Behcet's disease.

The course of Behcet's disease is still unclear. Inherited and environmental factors are thought to trigger an immune reaction and cause a systemic vasculitis. HLA B51 positivity has been reported to be significantly higher in patients with Behcet's disease. Among the acquired causes, streptococcal or viral infection and some insecticidal chemicals have been implicated.

Pathogenesis of aneurysms in Behcet's disease is thought to be caused by obliteration of vasa vasorum by inflammatory process, resulting in disruption of the nutrient flow to the aortic wall⁵. The intimal layer is thickened by fibroblasts and smooth muscle cells, whereas the media and adventitia are disrupted. Destruction of the media seems to be responsible for the development of the saccular aneurysmal dilatation.

Pseudoaneurysm formation at sites of arterial puncture has been recognized in patients with Behcet's disease⁴. Arterial reconstructions are also prone to thrombosis and pseudoaneurysm formation², as was in our patient.

Most deaths from Behcet's disease stem from arterial complications, resulting in ischemic bowel perforations, exsanguinating hemorrhage after aneurysm rupture, and cerebrovascular accidents. To prevent this outcome, such patients should receive immunosuppressive therapy. There is a wide range of different treatment options, which include oral prednisolone, intravenous methylprednisolone, cyclosporin A, azathioprine, chlorambucil, etc. It is likely that the cascade of arterial lesions is similar and related to the flares and resolution of the oral and genital lesions. Appearance of new oral and genital lesions may be predictive of new arterial complication in the future and may indicate the need for increased immunosuppressive therapy. In retrospect, the failure of the first repair could be attributed to the active phase of the disease and the absent of immunosuppressive therapy



Fig. 1: Angiogram showed right Common Carotid Artery Pseudoaneurysm

Conclusion

Behcet's disease is an uncommon disease in our country. Vascular surgeons dealing with young adults with peripheral aneurysms/pseudoaneurysms must be aware of this challenging clinical entity. Careful follow-

up with noninvasive testing combined with aggressive immunosuppressive therapy may allow early intervention and prevent recurrent anastomotic, remote or puncture site pseudoaneurysm formation and their attending complications.

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