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A Case of Behçet's Disease with Arterial Occlusion and Multiple Aneurysms

Vascular involvement in Behçet's disease is divided into venous and arterial thrombosis and arterial aneurismal formation. Multiple arterial aneurysms rarely occur in Behçet's disease; however, when they do occur, they cause so me complex signs and symptoms related to the location of arterial involvement. We descri be a 22-year-old male with Behçet's disease and multiple arterial aneurysms in the main arterial branches of the neck, such as left and right subclavian aneurysms, innominate and left caro tid bifurcation arterial aneurysms, together with right vertebral and left subclavian artery occlusions.

This case shows that multiple arterial involvem ents should be considered as one of the possible manifestations of Behçet's disease.

Keywords: Behçet's syndrome, aneurysm, angiography

Introduction

Behçet's disease is a chronic disease with multi-system involvement, characterized clinically by oral and genital aphthae, cutaneous lesions and ophthalmologic, neurologic and/or gastrointestinal manifestations. Although Behçet's disease is seen worldwide, the highest prevalence is reported from Japan, the Middle East and the United States. The age at onset of manifestation that fulfills the diagnostic criteria is typically in the range of mid 20s to 30s. In most countries, males are affected more frequently than females. Most patients present initially with mucocutaneous manifestations, and evidence of ocular and neurologic involvement may appear several years after diagnosis. Morbidity and mortality are the results of intracranial hemorrhage from aneurismal rupture.[1] Hearing and vestibular involvement as well as psychiatric problems may also occur. Behçet's disease can involve systemic arteries and veins, more commonly the latter, as well as the pulmonary circulation. About 8% of patients with Behçet's disease have severe vascular complications, such as arterial aneurysm and occlusion.[1] There are some reports of aneurismal arterial involvement of abdominal aorta, aortic arch[2], pulmonary, renal and subclavian artery in the literature.[2-6]

We present a known case of Behçet's disease in a young man, whose pulseless left upper limb was the chief vascular complaint, causing him to be referred for angiography. We are particularly focusing on multiple coincidental findings in digital subtraction angiography.

Case Report

A 22-year old man was admitted in our center for evaluation of his left upper limb pulselessness. He was a known case of Behçet's disease with a history of oral aphtosis from 7 years before, pseudofolliculitis from 5 months before, grandmal

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seizure, visual disturbances, speech problems, urinary incontinence and a new manifestation of left radial artery pulselessness from 4 months previously, which had been found in a routine physical examination. On our initial physical exam, a new finding was a small painful, pulsatile mass on the left clavicle medially.

On MRI, he had abnormal high T2 signal areas that were seen in pons, midbrain and basal aspect of cerebellar hemispheres that showed faint enhancement on post contrast images. In Color Coded Doppler assessment, there was an aneurismal dilation of the distal left common carotid artery, carotid bulb and internal carotid artery. Because of the high suspicion of vascular complications in this case, he had been referred to our center for angiography.

After evaluation of the coagulative conditions of the patient and a rheumatologic consultation, by using standard methods, digital subtraction angiography through right femoral artery was done by GE DSA Angiography system. Because of the high probability of vascular complication in left subclavian artery, initially an arch aortogram was obtained by placing a 5F pigtail catheter in the aortic arch. Multiple and complex findings of this image will be discussed later.

The left subclavian artery was obstructed proximally. There was a large aneurysm at the base of the left subclavian artery. The aneurysm had a fusiform, irregular, and deformed wall, which could be related to thrombosis (Figure 1, left black arrowhead). There was an aneurysm of similar size in the innominate artery and a large aneurysm in the proximal right subclavian artery (Figure 1, black curved arrowhead, and right black arrowhead respectively). Surprisingly, there was an obvious aneurysm at the bifurcation of the left common carotid artery (Figure 1, white arrowhead); the right carotid artery was normal.

By non-visualization of the right vertebral artery in contrast injection to the right subclavian artery, and not visualization of the left subclavian artery in the delayed image, occlusion of the right vertebral artery was demonstrated (Figure 2).

All these complex findings were related to the underlying disease. However, except for pulselessness of the left upper limb, there was no clinical symptom related to digital subtraction angiography findings. Angiography was terminated successfully, and the patient discharged without any complication after the diagnostic procedure.

Discussion

Vascular complications are a known feature of Behçet's disease, particularly affecting venous system. Arterial involvement has been reported in a few cases at various sites of vascular system, presenting with thrombosis or aneurysm. In our literature review, there was no report of carotid artery bifurcation aneurysm, which was present in our case; furthermore,

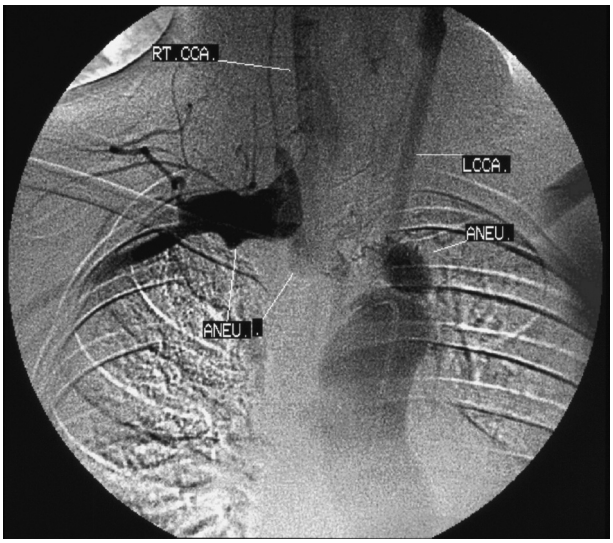

Fig 1. Illustration of the arch of aorta and the origins of neck vessels in a known case of Behçet's disease, with multiple aneurysms and right vertebral artery occlusion.

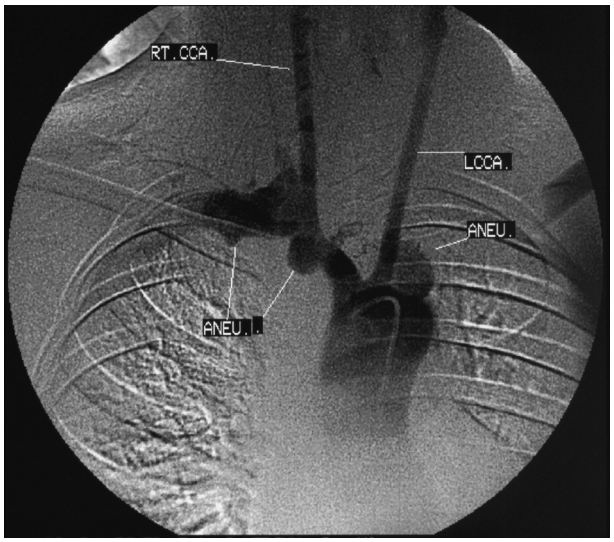

Fig 2. Illustration of right vertebral artery occlusion in delayed image of arch aortography.

this patient had also multiple aneurysms that is a rare finding in Behçet's disease. Another interesting point in this case was that occlusion and multiple aneurysms were present coincidentally. Bilateral subclavian artery aneurysm and innominate artery aneurysm were associated with right vertebral artery and left subclavian artery occlusion. Chronic inflammatory process had been detected in the adventitia that is compatible pathologically with diagnosis of Behçet's disease.[2]

This case had only the symptoms limited to his left upper limb vascular system, as could be expected with regard to his angiographic results. However, other findings in our patient were incidental findings, which can lead to clinical manifestations at future. Thus, it is possible that the low incidence of arterial involvement in Behçet's disease is related to our inability to find them, not actually for the disease itself. Because some of these findings are life threatening, and for the importance of early diagnosis[3], better assessment of vascular system by minimal invasive or non-invasive methods such as MRA is recommended.[2]

As previously noted, the pressure of subclavian artery aneurysm can lead to nerve compression and neurologic signs and symptoms of brachial plexopathy, and even an upper mediastinal mass.[4] This means that some vascular complications in Behçet's disease mayhaveindirectmanifestationsanditshouldbe kept in mind as a differential diagnosis of the noted problems.

For clinical decision, the patient was referred to a vascular surgeon for repair of his vascular aneurysm; control angiography has been planned to be done 6 months postoperatively.[3] Since either complication or recurrence is possible, prolonged monitoring is required.[5]

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