BEHÇET'S DISEASE AND ARTERIAL ANEURYSMS

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SUMMARY: Vascular lesions of the Behçet's Disease are known as Angio-Behçet Syndrome. Aneurysm formation is an important component of this syndrome. Seven such cases are presented and surgical intervention described. Two operated patients needed further intervention. We conclude that all patients with Behçet's Disease should be searched for arterial aneurysms and especially young patients with aneurysmatic formations should be investigated for Behçet's Disease and also patients with Behçet's Disease are not good candidates for surgery.

Key Words: Behçet's disease, aneurysm.

INTRODUCTION

Hulusi Behçet MD, described a triad which consisted of iridocyclitis, oral aphtous lesions and genital ulcerations back in 1937 which was named after him. Today it is known as a multi-system disease due to 'lymphocytic microvasculitis' (1,7,17).

Although vascular involvement was first recognized only 10 years after the disease was described it was not until 1965 that Urayama and his associates found out that the venous and arterial lesions were not a coincidence at all. They defined vascular lesions like venous thrombosis, arterial occlusion and aneurysms as Angio-Behçet Syndrome (24). According to 'International Study Group for Behçet's Disease' vascular lesions are considered as a cardinal symptom of the disease today (9,12).

The primary cause in the arterial lesions is a microvasculitis effecting the vasa-vasorums which induce necrosis in intimal and medial layers of the artery due to ischemia (7).

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Arterial aneurysms have been observed as frequent as 0.15 to 2.5% among patients with Behçet's Disease (7,24). These aneurysms could be seen in visceral and peripheral arteries as well as the aorta and its major branches. They are the leading mortality cause along with the Neuro-Behçet Syndrome (23). On the other hand this progressive systemic disease carries a risk on the vascular surgical intervention for prognosis.

The aim of this paper is to present the specific diagnostic and therapeutic experience on the arterial aneurysms seen in Behçet's Disease.

PATIENTS

Seven patients with aneurisms and Behçet's Disease were investigated and treated in the Çukurova University Medical Faculty Departments of Thoracic and Cardiovascular Surgery, Dermatology and Neurosurgery between 1986 and 1992. All of the patients were male with a mean age of 33 internal (24-43). Three aneurysms were found on the popliteal artery, one on the carotid artery, one on the splenic artery, one was intra-cranial on the anterior communication artery and one false aneurysm developed on the proximal

Table 1: Location of aneurysms.

Popliteal artery	3
Carotid Artery	1
Anterior Comninicating artery (intracranial)	1
Carotico-Subclavian by pass (false aneurysm)	1

portion of a carotico-subclavian bypass graft (Table 1).

Case 1: Y. Ç., 38, male (1986/4355)

Behçet's Disease had been diagnosed in Department of Dermatology 3 months ago. He has observed a mass on the right fossa poplitea for 1 month. He was suffering from pain on the right calf when he walks about 50 meters. A pulsatile mass was palpated and popliteal artery aneurism was diagnosed by right femoral angiography.

Aneurismectomy, distal embolectomy and end to end anastomosis were done. Postoperative angiography was obtained and the arterial tree was patent. He was being discharged without complication.

Four months later he admitted due to rest pain on the right leg. Distal pulses were absent and angiography displayed proximal occlusion of femoral artery. Bilateral lomber sympathectomy was performed. He was free of pain. On the 7th postoperative month 5th toe on right side was amputed because of gangrene.

Case 2: V. E., 37, male (1987/5923)

A Popliteal artery aneurism was shown by femoral angiography in this case with left popliteal pulsating mass. Aneurismectomy and end to end anastomosis were done.

Incomplete Behçet's Disease was diagnosed in postoperative period in the Department of Dermatology because of our observations of papulopustulous lesions on the shoulders and back in this relatively young patient the histopathological findings confirmed the diagnosis.

Case 3: S. B., 35, male (1987/8847)

Acute pain and pallor on left foot were the cause of his admission to our emergency room. A pulsating mass and findings of acute arterial obstruction were seen. Aneurismectomy, distal embolectomy and interposition of saphenous vein were done. Aphtous lesions on the mouth and serotum were encountered when his history was taken postoperatively, so Behçet's Disease was considered. The diagnosis was confirmed by Department of Dermatology.

Case 4: S.K., 40, male (1988/17001)

A growing mass on the left sub-mandibular region was noticed by this patient with Behçet's Disease. CT-scan revealed internal carotid artery aneurism and it was displayed by percutaneous carotid angiography. During the operation internal carotid artery was ligated because retrograde flow was not seen, probably due to preoperative distal embolization from the aneurismal sac. Although no complications were seen during the operation, aphasia and right hemiplegia was developed 24 hours later. Temporoparietal ischemia were seen by CT-Scan. He improved some days later and rehabilitated.

Case 5: M. K., 43, male (1991/186)

This right handed patient was admitted to the Dept. of Neurosurgery with history of sudden occipital headache, vomiting and confusion. Neurological examination revealed nuchal rigidity, right sided hemiparesis, somnolence and speech problem. L. P; revealed bloody CSF. CT-scan showed sub-arachnoidal hemorrhagia and a small hematoma localized in the preportine cistern. Angiographic investigation demonstrated an aneurysmatic, dilatation of the anterior communicating artery.

General physical examination revealed depigmented satirical areas on the scrotum. His post-history disclosed arthralgia, trombophlebitis and transient aphtous lesions in the mouth. Behçet's Disease was considered and the diagnosis have been confirmed by the Department of Dermatology.

Case 6: I. Y., 24, male (1991/14971)

This patient with five years history of Behçet's Disease was being treated for superficial trombophlebitis in The Department of Dermatology.

Routine abdominal ultrasonographic examination demonstrated a 4x5 cm cystic lesion near the tail of the pancreas. CT-scan and celiac angiography demonstrated a splenic artery aneuorizm. Splenectomy and aneurimectomy were performed.

Case 7: O. S., 43, male (1992/8393)

The patient had been admitted to our department 9 years ago for subclavian steal syndrome on the left.

Angiography had revealed occlusion of the left subclavian artery. He had been operated and a 8 mm synthetic graft had been used for carotico-subclavian artery by pass (1983/13759).

Following his discharge the patient had been by seen by an another physician in his home town and Behçet's Disease had been diagnosed and treated.

The patient admitted to our department with a 5x5 cm supraclavicular pulsatile mass Angiography demonstrated a fals aneurism at the site of the proximal anastomosis of the previous carotico subclavian by pass. The diagnosis of Behçet's Disease was confirmed by the Department of Dermatology.

The patient underwent operation in which both the aneurismal sac and synthetic graft have been respected. Carotid artery was repaired primarily and the subclavian artery was ligated. He was later discharged in good condition.

DISCUSSION

Aneurysms associated with Behçet's Disease are frequently seen before the fourth decade. Aneurysm at this age group is mostly due to mucopolysaccharidosis, Ehler's Danlos Syndrome, or syphilis (10). Excluding these in the presence of an aneurysm, especially in a patient who is an oriental young male, Behçet's Disease must be considered. In this series who patients had attended with popliteal aneurysms (Cases 2 and 3). Behçet's Disease was considered on both of them because they were young and histopathological findings at the aneurysm site were corresponding with lymphocytic vasculitis in vasa-vasorum and granulomatous changes in the arterial wall. The diagnosis was made by further investigations in the Department of Dermatology.

Extra-cranial and peripheric arterial aneurysms are recognized easily but those located in the thorax, abdomen and cranium usually recognized after a complication developed. Intra-cranial aneurysms usually appear with a lethal hemorrhage (16). The aneurysms on the aorta and its branches are rarely perceived on routine physical examination but can be noticed with systemic ultrasonographic analysis for aneurismal formation (1, 3, 8,11). With this in mind all patients with Behçet's Disease must be analyzed by means of non-invasive methods. In our series the aneurysm on the splenic artery was suspected on the routine ultrasonographic investigation of patients with Behçet's Disease and confirmed by both CT-scan and angiography (Case 6).

The primary lesion in the Behçet's Disease is lymphocytic vasculitis. It strikes vasa-vasorums and necrosis of intimal and medial layers of the artery develops due to ischemia (21). The disease proceeds with acute exacerbations (18,25). Surgical intervention is especially unsuccessful in the acute phase and surgery must be postponed until the disease can be controlled. Re-thrombosis and repeating aneurisms at the anastomosis regions are frequently reported (14,15,19). Ligation recommendations are gaining more attention for this reason (2, 5, 13, 22). The patient with aneurism on the carotico-subclavian bypass (Case 7) and the one with thrombosis formation in the femoral artery subsequent to popliteal aneurismectomy (Case 1) are good examples of poor surgical outcome in this disease. Treatment should be conservative where possible. If an anastomosis have to be performed; the location must be distant from the effected region and a synthetic graft must be preferred (2,10,14,19, 24).

CONCLUSION

All young patients with aneurysms have to be searched for Behçet's Disease and in the patients with Behçet's Disease a systemic investigation for an aneurysm should be accomplished.

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