Cerebral Aneurysms Associated with the Behçet’s Disease

Department of Neurosurgery, Brain Research Institute, Yonsei University College of Medicine, Seoul
Ilsan Hospital, National Health Insurance Corporation, GyeongGi-Do

Tae Yup Kim, MD · Jae Whan Lee, MD · Seung Kon Huh, MD · Kyu Chang Lee, MD

ABSTRACT

We report two cases of ruptured cerebral aneurysms associated with Behçet’s disease. One was from right superior cerebellar artery and treated by intra-aneurysmal coil embolization. The other was from the bifurcation of right middle cerebral artery and treated by clipping. Both patients showed good result. (Kor J Cerebrovascular Surgery 8:135-7, 2006)

KEY WORDS: Intracranial aneurysm · Behçet’s disease

Introduction

Behçet’s disease is a multi-system disorder presenting with recurrent oral and genital ulcerations as well as ocular involvement. The etiology and pathogenesis of this disease remain obscure; it is considered an autoimmune disease since vasculitis is the main pathologic lesion and circulating auto-antibodies to human oral mucous membrane are found in approximately 50 percent of the cases.

There are four types of well-known vascular lesions in the Behçet’s disease; arterial occlusion, arterial aneurysm, venous occlusion, and venous varices in the body. But, the intracranial aneurysms associated with Behçet’s disease are not common.

We report two cases of ruptured cerebral aneurysms associated with Behçet’s disease.

Case Reports

Case 1

A 34-year-old man admitted to our hospital with sudden severe headache, nausea, vomiting, and, loss of consciousness. He had suffered from Behçet’s disease since he was 24 years old. At this time, he exhibited recurrent aphthous oral ulcer, many pustules on the trunk, and blindness due to uveitis and cataract of right eye. And when he was 30 years old, he lost the remained vision of the left eye due to uveitis, and cataract, so became completely blind. On admission, he was drowsy and confused without focal neurological deficit and clinically appropriate to Hunt-Hess grade III. Brain CT scan revealed a dense subarachnoid hemorrhage in the basal cisterns and both sylvian fissures (Fig. 1A.) and digital subtraction angiography showed a 2.4 × 1.8 mm-sized right superior cerebellar artery aneurysm projecting anteroinferiorly (Fig. 1B). Since the surgical approach was difficult due to the location of the aneurysm, we tried endovascular treatment. Using ultra soft Gugliemi detachable coil (2 mm × 4 cm × 1ea), the embolization was performed and final angiography showed small remnant aneurysmal neck without parent artery compromise (Fig. 1C). After coil embolization, tracheostomy was done for care of pneumonia. During the recovery period, the orbital cellulitis associated with Behçet’s disease was developed and treated with IV antibiotics, Diamox (for intra-ocular pressure care), and topical eye drops. Forty-eight days after the aneurysmal rupture, he was discharged without further neurological deficit.
Case 2
A 34-year-old woman was transferred to our hospital for management of known subarachnoid hemorrhage. She had a past history of Behçet’s disease since she was 32 years old and had had oral ulcers, genital ulcers, and folliculitis-like skin lesions, which had been well-managed with medications (prednisolone, Colchicine). Five days before the transfer, she visited the emergency room of nearby hospital presenting sudden headache, nausea and vomiting. Her brain CT scan showed a small amount of subarachnoid hemorrhage, and by the cerebral angiography, a $4.5 \times 3.7$ mm-sized, superolaterally projecting aneurysm in the bifurcation area of the right middle cerebral artery was diagnosed. At the time of visit to our hospital, her clinical status was fit to Hunt-Hess grade II and the effacement of right sylvian fissure was shown on the follow-up brain CT scan, which was suitable for Fisher grade II (Fig. 2). She underwent the operation, and the aneurysm was successfully clipped. We were worry about the delay of wound healing or secondary infection/inflammation due to Behçet’s disease, but there was no problem.

Twenty-one days after the aneurysmal rupture, she was discharged without neurological deficit.

Discussion
Behçet’s disease is characterized by specific clinical manifestations including recurrent aphthous stomatitis, genital ulcers, anterior uveitis, and, vasculitis. There are many vascular lesions and aneurysms in the major systemic arteries associated with Behçet’s disease. But cerebral aneurysms are rare. Al-Dalaan, et al and Benamour, et al found only one cerebral aneurysm in 119 cases and 316 cases with Behçet’s disease, respectively, whereas they did not routinely perform cerebral angiographic studies.\(^1\)\(^2\)

In vasculo-Behçet’s disease, the histological findings of aneurysms include ruptured internal and external elastic laminae with thickening of the tunica media, and vasculitis of the vasa vasorum with perivascular lymphocytic infiltration.\(^3\) But Katoh, et al and Tsuji, et al pointed out that in cerebral aneurysms with Behçet’s disease, histological examination in two patients showed no feature of vasculitis.\(^3\) So it is difficult to exclude the possibility of coincidental association in most of the cases. However, as Nakasu, et al proclaimed, the high frequency of multiple aneurysms in Behçet’s disease may support the likelihood that Behçet’s disease has some role in the formation of cerebral aneurysms.\(^6\) Furthermore, Nakasu, et al reported that cerebral
aneurysm associated with Behçet’s disease disappeared after steroid therapy. And it is reported that recurrence of the cerebral aneurysms in the Behçet’s disease is frequent.

Considering all these facts and the severity of aneurysmal rupture, intracranial aneurysms in Behçet’s disease deserve to be concerned particularly and the efforts to reveal the pathogenesis of aneurysm formation in Behçet’s disease and the role of steroid for prevention of aneurysm formation should be made.

Conclusion

We treated two patients with ruptured aneurysm associated with Behçet’s disease. One was treated with endovascular intra-aneurysmal coil embolization, and the other was treated with direct clipping of aneurysmal neck. Both patients showed good outcome, but long-term follow-up and observation for formation of de novo aneurysm should be necessary.

REFERENCES