Multiple fusiform myxomatous cerebral aneurysms in a patient with Carney complex

Case report

KYOUNG SOO RYU, M.D., SUN-HO LEE, M.D., SEONG-Hyun PARK, M.D., JAECHAN PARK, M.D., SONG-KYOO HWANG, M.D., PH.D., AND IN-SUK HAMM, M.D., PH.D.

Department of Neurosurgery, School of Medicine, Kyungpook National University, Daegu, Republic of Korea

Carney complex is a rare autosomal-dominant familial tumor syndrome that involves the triad of myxoma, mucocutaneous pigmentation, and endocrine overactivity. To the best of the authors’ knowledge, there are no reports of multiple fusiform aneurysms coinciding with atrial myxoma.

The authors report the case of a 38-year-old woman with typical Carney complex who had multiple skin myxomas, endocrine abnormalities, and multiple brownish perioral lesions. Multiple fusiform aneurysms were also discovered after the recurrence of atrial myxoma. During a follow-up period of > 10 years, there have been no angiographic changes in the aneurysms and no progression of symptoms. (DOI: 10.3171/JNS/2008/109/8/0318)

KEY WORDS: • atrial myxoma • Carney complex • multiple aneurysms • pituitary adenoma

Case Report

History. At the age of 17 years, bilateral atrial myxomas were diagnosed in the patient when she was hospitalized for dyspnea after presenting with palpitation and symptoms of exertion. Echocardiography revealed well-pedunculated masses in both atria and left ventricle. This patient has been followed up for > 10 years. We also review the literature to provide additional clinical, radiological, and pathological information on this rare neurological disorder and to define the risk factors for the formation of cerebral aneurysms in patients with Carney complex.

Carney complex is a rare autosomal-dominant familial tumor syndrome involving the triad of myxoma, mucocutaneous pigmentation, and endocrine overactivity. There are several reports in the literature on multiple fusiform aneurysms occurring in patients with atrial myxomas, but few deal with the pathogenesis and clinical progression of the disease. To the best of our knowledge, there are no reports of Carney complex occurring concurrent with multiple fusiform aneurysms in atrial myxomas.

We report the case of a patient with Carney complex and multiple fusiform cerebral artery aneurysms that occurred after a successful resection of a recurrent myxoma in the left atrium and left ventricle. This patient has been followed up for > 10 years. We also review the literature to provide additional clinical, radiological, and pathological information on this rare neurological disorder and to define the risk factors for the formation of cerebral aneurysms in patients with Carney complex.

Examination, Operation, and Postoperative Course. When the patient was 27 years old she had a recurrence of the bilateral atrial myxomas, presenting with sudden-onset dizziness, headache, blurred vision, and tingling sensations in her tongue, arm, and the left side of her face. She was hospitalized, and echocardiography revealed 2 pedunculated masses in the left atrium and left ventricle, suggesting myxoma recurrence. Cerebral angiography revealed multiple fusiform aneurysms in the basilar artery, proximal PICA, left P2, and right P4 segments, temporal branch of the left MCA, and distal branches of the right MCA and ACA (Fig. 1A–C). The patient underwent surgery to excise the atrial masses; histological examination confirmed the masses to be myxomas. During the follow-up after surgery she only complained of intermittent headaches. At the age of 29 years she underwent MR angiography, which revealed the same fusiform aneurysmal mass lesion in the left P2 segment observed 2 years earlier; she had remained symptom free since the second operation.

At the age of 38 years, the patient was admitted for
evaluation of painful palpable masses in both breasts; the results of a biopsy confirmed that the masses were fibroadenomas. The physical examination revealed multiple brown perioral lesions ~1 mm in size, a subcutaneous mass on the right forearm, and a 5-mm extruding mass on the right nostril. A biopsy of the right nostril and right forearm masses confirmed them to be myxomas. Brain MR imaging revealed mass lesions in the pituitary gland. Serum analysis revealed an elevated insulin-like growth factor–I/prolactin level, but the serum cortisol, 24-hour urine 17-hydroxycorticosteroids, and growth hormone levels were within normal limits. An abdominal CT scan was unremarkable except for a small hepatic mass. She again underwent cerebral angiography, which revealed the same previously noted multiple fusiform aneurysms in the basilar artery, proximal PICA, left P₂ and right P₄ segments, temporal branch of the left MCA, and distal branches of the right MCA and ACA. There were no new lesions and no changes in the existing lesions (Fig. 1D–F).

Discussion

Carney complex is a triad of myxoma, mucocutaneous spotty pigmentation, and endocrine overactivity. It is a rare autosomal-dominant familial tumor syndrome that is more common in younger individuals (mean age 26 years) and females.⁶,¹⁸,²² As shown in our patient, cutaneous myxomas and spotty skin pigmentation are the most common clinical characteristics, occurring in 90% of patients with Carney complex.³,¹⁰ Atrial myxomas appear in 30–60% of patients with Carney complex. Although the initial symptoms include cardiac or systemic findings (fever, weight loss, malaise, and embolic strokes), fusiform, multiple cerebral aneurysms are sometimes observed before or after excising the mass in rare cases. Moreover, to the best of our knowledge, no case of fusiform aneurysm combined with Carney complex has been reported.

The pathogenesis of these aneurysms is also not well established. In the literature, the process of aneurysm formation has been grossly classified into 2 mechanisms: postembolic vascular damage and subsequent scarring resulting in a change in the flow dynamics, which promotes the formation of an aneurysm, and later active invasion of the vascular wall by a viable tumor embolus.⁷,³,⁹,¹² Sabolek et al.¹⁷ have reported an elevated interleukin-6 level at the time the aneurysm was detected. Although their use of interleukin-6 monitoring did not reach definite relevance, its use appears to be helpful in diagnosing and monitoring a myxomatous aneurysm.¹⁷ The angiographic appearance of myxomatous aneurysms are a typical finding, and characteristics include a filling defect, interruption of flow, and local arterial dilation. Morphologically, myxomatous aneurysms can appear as fusiform outpouchings or saccular dilations of various sizes⁵,¹³ and no preferential location. The lesions can appear as fusiform tubular dilations of the cerebral arteries within the sulci on T₁- and T₂-weighted MR images. Sometimes, enhancing tumor tissue is observed. Nucifora and Dillon¹³ recommended MR angiography of any tubular enhancing lesion to con-

**Fig. 1.** Angiograms obtained 11 years earlier (A–C) and at last follow-up (D–F). A: Left vertebral angiogram showing multiple fusiform dilations in the basilar artery, proximal PICA, and left P₂ and right P₄ segments. B: Left internal carotid artery angiogram revealing a fusiform aneurysm of the MCA temporal branch. C: Right internal carotid artery angiogram showing a fusiform aneurysm on the distal MCA and ACA. D–F: Follow-up cerebral angiograms obtained 11 years after the first detection. There was no difference in the diffuse aneurysm size and number.
firm the vascular nature and determine if there are any ischemic changes.

Because the natural course of these aneurysms is unclear due to their rarity, there are no established treatments for these aneurysms, particularly in cases combined with Carney complex. Roeltgen et al. have suggested that the clinical course of a myxomatous aneurysm can be divided into 3 phases: 1) resolution after treatment; 2) progressive enlargement and potential hemorrhage; and 3) spontaneous resolution or stabilization. Although there are reports of a successful aneurysmectomy—aneurysm wrapping and coil embolization for a moderate-sized or giant aneurysm—there are no feasible surgical interventions for multiple fusiform aneurysms in the distal branches of the major cerebral arteries, which are more common in cases of a myxomatous aneurysm. Under the hypothesis of dividing tumor cells being responsible for the formation of these aneurysms, chemotherapy has been performed alone or in conjunction with low-dose radiation therapy, but the results are not consistent.

Except for unproven treatment options, the natural course of these aneurysms probably occurs in 1 of 2 ways, although the courses are not well documented. Roeltgen et al. have described a progressively enlarging aneurysm during a follow-up examination, and Flemming et al. suggested that the predictor of an aneurysm rupture is an increase in diameter. However, there is no direct link between the size of the aneurysms and hemorrhage.

Furthermore, although the authors of a case series have shown that the immediate central nervous system manifestations are often severe and may be fatal, the manifestations also indicated a good prognosis with respect to the recurrence of neurological events. In contrast, spontaneous regression or stabilization has been demonstrated in patients in whom the primary atrial tumor was removed. Branch et al. have reported cases of an angiographically proven resolution of a fusiform cerebral aneurysm after removal of a cardiac tumor. However, recommendations for conservative treatment are troublesome due to the paucity of long-term angiographic follow-up results. Interestingly, in our patient the aneurysmes were uneventful for >10 years without any specific treatment, and there were no changes in the number or size noted on angiography. Therefore, this case shows that conservative treatment is a valid option for myxomatous aneurysms after the treatment of an atrial myxoma.

References


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Address correspondence to: Sun-Ho Lee, M.D., Department of Neurosurgery, School of Medicine, Kyungpook National University, 50 Samdul-2-ga, Jung-gu, Daegu, 700-721, Republic of Korea. email: sobotta@dreamwiz.com.