Moyamoya Disease Associated
With Pulmonary Sarcoidosis

—Case Report—

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Abstract

A 61-year-old female presented with a unique case of moyamoya disease associated with pulmonary sarcoidosis. She was admitted for sudden onset of left temporalgia with episode of numbness on face, tongue, and upper extremity on the right side. The next morning, she had symptoms of Gerstmann syndrome and her ability to speak was disturbed. Her medical history included radical resection of lung cancer on the right side. She had no symptoms of pulmonary sarcoidosis. Neuroimaging showed an infarction in the left occipital lobe. Angiography showed occlusions of the bilateral internal carotid arteries at the supraclinoid portions. Subsequently, a left superficial temporal artery-middle cerebral artery anastomosis with encephalo-myo-synangiosis was performed. Ninety-three days after admission, she suddenly developed dyspnea which resulted in death 3 hours later. Autopsy findings showed typical epithelioid granulomas of sarcoid type in the lymphnodes of the peribronchus, lung, and liver. Thrombotic emboli were found in the bilateral pulmonary arteries, and marked fibrous intimal thickening in the bilateral internal carotid arteries. Immunological reaction with inflammatory events may cause pathological changes in patients with moyamoya disease or sarcoidosis. The co-incidence in this case suggests that some common inflammatory events may be involved in the pathogenesis of these diseases.

Key words: moyamoya disease, immunological reaction, sarcoidosis

Introduction

Moyamoya disease may be associated with other disorders, such as intracranial aneurysm, arteriovenous malformation, disorder of the renal artery, phacomatosis, Down’s syndrome, brain tumors, etc.9) We describe a unique case of moyamoya disease associated with pulmonary sarcoidosis.

Case Report

A 61-year-old female was admitted to our hospital after suffering sudden onset of severe left temporalgia on July 28, 1995. Ten days before admission, she had had an episode of numbness in the face, tongue, and upper extremity on the right side. Her medical history included right lung cancer with radical operation at age 53 years and hypertension for 3 years. There was no family history of cerebrovascular or immunological disease.

Physical examination on admission showed hypertension (180/100 mmHg) with normal cardiac rhythm and no evidence of peripheral vascular disease or symptoms of pulmonary sarcoidosis. There was no cardiac murmur and no skin lesion. She was alert with normal speech but had slight impairment of memory for recent events. Examination of the visual field (Bjerrum screen) revealed complete right homonymous hemianopsia. Chest radiography, complete blood count, electrolytes, and liver and pulmonary function test results were within normal limits.

The next morning, the patient had symptoms of Gerstmann syndrome and her ability to speak was...
disturbed. Head computed tomography and magnetic resonance imaging showed an infarction in the left occipital lobe. Brain single photon emission computed tomography showed a severe (<70%) reduction in cerebral blood flow in the left hemisphere. Angiography disclosed occlusions of the bilateral internal carotid arteries around their terminal portions. Basal moyamoya vessels had developed, in which leptomeningeal anastomoses were observed. The left posterior cerebral artery was occluded at the interpeduncular segment. On October 5, 1995, a left superficial temporal artery-middle cerebral artery anastomosis with encephalo-myo-synangiosis was performed. No ischemic attacks occurred postoperatively and her symptoms gradually improved. On October 29, 1995, she suddenly developed dyspnea and bradycardia, and died 3 hours later. An autopsy was performed.

The general postmortem examination revealed mild atherosclerosis of the aorta. The heart, adrenal glands, and small systemic arteries were normal. The right lung was totally removed, but without recurrence in the residual left lung or lymphnodes across the bronchus. No metastatic or other neoplastic tumors were found. Thrombotic embolisms were found in the bilateral pulmonary arteries, which had caused acute pulmonary infarction. However, no intravenous thrombus was found elsewhere. The vertebral and basilar arteries had slight atherosclerosis. The internal carotid arteries appeared hypoplastic at the suprachinoid portion. Both anterior and middle cerebral arteries were hypoplastic at their proximal segments. The left occipital hemisphere showed infarction. There was no gross evidence of vascular malformations in the brain tissue.

Microscopic examination revealed moderate lymphocytic infiltration in the leptomeninges with occasional perivascular lymphocytes, and necrotizing changes in the left occipital lobe. The intrapetrosal segments of the internal carotid arteries exhibited marked concentric fibrous intimal thickening with reduplication-fragmentation of the elastic layers, especially the inner layer (Fig. 1). Similar lesions were found in the proximal segments of the anterior, middle, and posterior cerebral arteries and posterior communicating arteries, in which the intimal layer showed eccentric stenosis and elastosis. The left posterior cerebral artery showed eccentric stenosis. The vertebral and basilar arteries were normal, and no malignant cells were found in the residual left lung and lymphnodes. Typical epithelioid granulomas of sarcoid type, consisting of infiltration of giant cells, epithelioid cells, and lymphocytes without caseating necrosis, were found in the lymphnodes of the peribronchus, left lung, liver, and spleen (Fig. 2).

**Discussion**

Hereditary factors and/or acquired disorders, e.g. chronic arthritis due to immunological reaction, may be involved in the occurrence of moyamoya disease, but the etiology and pathogenesis are still unknown. Histological examination has showed the presence of macrophages and T cells in the super-
ficial layers of intimal thickening in the intracranial major arteries following inflammatory events. Disarrangement of endothelial cells and basement membrane layers was induced by injection of Propionibacterium acnes. P. acnes has been detected in the lymph nodes of patients with sarcoidosis or Kawasaki disease (mucocutaneous lymph node syndrome) and significantly elevated antibody titer against P. acnes in the serum of moyamoya disease patients.

Sarcoidosis is a chronic, multisystemic disorder characterized by non-caseating epithelioid granulomas consisting of giant cells, epithelioid cells, and lymphocytes. The etiology of the disease is still unknown, but immunological factors are involved in the pathogenesis. Microangiopathy in sarcoidosis depends on the immunological reaction to endothelial cells, and histological evidence shows that damage to capillaries of autoantibodies to endothelial cells occurs in the lesions of sarcoidosis. Production of various cytokines from T cells occurs in patients with sarcoidosis. Growth factors and/or cytokines such as basic fibroblast growth factor and interleukin-1 might cause endothelial cell damage or subendothelial cell proliferation, which could lead to moyamoya disease. Recently, the relationship between sarcoidosis and mycobacteria infection, especially mycobacterium tuberculosis or P. acnes, has been shown. Although the significance of such an association in a single case is unlikely, the coincidence in this case indicates that common inflammatory events might be involved in the pathogenesis of both moyamoya disease and sarcoidosis.

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