Moyamoya-Like Vascular Abnormality in Pulmonary Sarcoidosis

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Introduction
Sarcoidosis affects the central nervous system in about 5% of the patients usually in the form of cranial neuropathies, basal meningitis, intracranial masses, diabetes insipidus, encephalopathies and seizures [1]. Although cerebral vascular involvement is not uncommon, large cerebral artery involvement in sarcoidosis is extremely rare [2, 3]. We describe a patient with sarcoidosis who showed angiographic findings similar to moyamoya disease.

Case Report
A 45-year-old woman suddenly developed chest and back tightness associated with numbness on her right inguinal area (T10–12 dermatome), which spread gradually over the trunk. Brain MRI performed at a local clinic showed an old infarction in the right frontal area (fig. 1). She did not have a history of hypertension, diabetes mellitus, cardiac diseases, tuberculosis, strokes or seizures. Cigarette smoking, alcohol ingestion and any regular medications including birth control pills were denied. Family history of stroke was also denied. Physical and neurological examination revealed no abnormal signs except for bilaterally decreased superficial sensation in the distribution from the right 2nd thoracic and left 8th cervical to bilateral 2nd lumbar segments.

Laboratory results revealed decreased platelet count (125 x 10³/mm³) and an elevated serum angiotensin-converting enzyme level (67.8 U/l, normal range 8–52 U/l). Electrocardiogram was normal. Chest X-ray and CT scan showed bilateral hilar and mediastinal lymphadenopathy. Specimens obtained from transbronchial lung biopsy revealed noncaseating granulomas, which was consistent with sarcoidosis.

Transfemoral cerebral angiography showed bilateral occlusion of the internal carotid arteries (ICAs) at the terminal portion (fig. 2). There were no definite basal moyamoya vessels. Collateral circulation was provided by the verteobasilar system through posterior communicating arteries and by external carotid arteries through transdural anastomosis via meningeal arteries. Brain SPECT disclosed perfusion defect on the right frontal cortex and decreased perfusion reserve in the anterior cerebral artery (ACA) and middle cerebral artery (MCA) territory bilaterally. MRIs of the cervical and thoracic spine showed no significant abnormalities. CSF examination results were within normal limits. Needle EMG examination showed fibrillation potentials and positive sharp waves in 2–8 left thoracic and 3–7 right paraspinal muscles, indicating bilateral thoracic radiculopathy.

Fig. 1. Fluid-attenuated inversion recovery MRI shows old cortical infarcts in the right frontal area (arrows).
Case Report

Fig. 2. The frontal view of the right (a) and left (b) carotid angiogram demonstrates bilateral distal ICA occlusions at their terminal portion (arrows). There are no definite basal moyamoya vessels. c The lateral view shows that occlusion (black arrow) occurred just distal to the ophthalmic artery (white arrow). d The vertebral angiogram shows collateral circulation provided by the vertebrobasilar system.

Prednisolone 55 mg per day was started, and her sensory symptoms gradually improved within 2 weeks. The serum angiotensin-converting enzyme level became normalized (12.7 U/l) 3 months after the treatment and chest X-ray showed a decreased extent of bilateral hilar opacities. Steroid was tapered to 15 mg per day, and the patient remained uneventful in the 9 months of follow-up.

Discussion

Similar to a previous report [4], our patient presented with thoracic radiculopathy. In addition, MRI showed subclinical lesions which were presumed to be watershed infarcts between the right MCA and ACA territory. Cerebral angiography demonstrated occlusion of both terminal ICAs similar to moyamoya disease (fig. 2). It seems that the vascular occlusion was not related to the radiculopathy.

Despite the frequent finding of vasculitis and cerebral infarcts at autopsy, clinical ischemic events have been rarely reported in patients with sarcoidosis [2, 3, 5]. Characteristic postmortem findings of these patients were the presence of sarcoid granuloma in the leptomeninges and brain parenchyma, with the invasion of the arterial wall by epithelioid cell granulomas which disrupted the media and the internal elastica causing stenosis or occlusion. In many instances, small perforating and medium-sized arteries were primarily affected [1, 6], resulting in small, asymptomatic cerebral infarctions. Although large cerebral artery involvement was reported in one patient [6], angiogram findings were normal.

In patients with sarcoidosis, cerebral angiogram findings have been rarely positive even in those with symptomatic stroke. According to a previous study, an occlusion of the A1 segment of the ACA was considered to be caused by inflammation extending to the proximal ACA because the patient had a granulomatous mass adjacent to the distal ICA [2]. Another study reported segmental narrowing and dilatation of large cerebral arteries [7], but clinical correlation was not made. To our knowledge, there has been only one reported patient who had moyamoya-like vasculopathy associated with sarcoidosis [3]. Similar to our case, the patient had bilat-
er al ICA occlusion at the terminal portion. However, she had occipital infarction due to additional left posterior cerebral artery occlusion which resulted in episodes of numbness on the right side, right hemianopia and Gerstmann syndrome.

Considering the relatively young age and the lack of any vascular risk factors, atherosclerosis is an unlikely cause of ICA occlusion in our patient. There was no angiographic evidence of vasculitis, and evidence of meningitis was not present on CSF examination. Moreover, autopsy findings of the case mentioned above [3] showed marked fibrous intimal thickening in the bilateral ICAs but without active inflammation [3]. Therefore, moyamoya-like vascular abnormality associated with sarcoidosis may not be related to active inflammation. However, the possibility of a healed stage of long-standing vasculitis cannot be ruled out. Alternatively, considering that moyamoya-like vasculopathy has been shown to be related to various systemic diseases such as chronic meningitis, fibromuscular dysplasia, Down syndrome or von Recklinghausen’s disease, certain common genetic or inflammatory processes may be involved in the pathogenesis of both diseases. Unfortunately, the pathologic examination for the cerebral vessel or dura mater was not possible because our patient refused to undergo the invasive procedure. Finally, a simple coexistence of sarcoidosis and moyamoya disease was not completely ruled out in our patient. However, considering occasional reports of large cerebral vessel involvement in sarcoidosis, our case together with the previously reported one [3] suggests that sarcoidosis may be related to large vessel involvement in the pattern of moyamoya vasculopathy. More researches are required to elucidate the true relationship between the two rare conditions.

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