Stroke as a Manifestation of Takayasu’s Arteritis Likely due to Distal Carotid Stump Embolism

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Abstract

The clinical symptoms of Takayasu’s arteritis (TA), which mainly affects the aorta and major aortic branches, vary widely depending on the site and degree of arterial lesions. We present herein the case of a young man whose initial symptom was pulmonary artery occlusion and who manifested TA 6 years later as cerebral embolism. Angiography confirmed bilateral common carotid artery (CCA) occlusion and a well-developed collateral circulation. The stump of the occluded CCA has both proximal and distal ends. The possibility of emboli from the occluded CCA (distal stump) seems to be the most probable explanation, as turbulent flow was detected at distal stump on color Doppler sonography. The carotid stump can be a potential source of emboli in TA as well as in atherosclerosis.

Key words: Takayasu’s arteritis, carotid stamp, ischemic stroke, pulmonary artery occlusion

(DOI: 10.2169/internalmedicine.49.3033)

Introduction

Takayasu’s arteritis (TA) is a chronic inflammatory arteritis that affects the aorta and main aortic branches, resulting in stenosis and obstruction secondary to thrombus formation or dilation due to aneurysmal formations of involved arteries (1, 2). Stroke in TA patients causes serious neurological deficits that negatively affect the prognosis (3). Nevertheless, literature regarding stroke due to TA is sparse and consists only of case reports and case series (4-7).

In the absence of specific laboratory tests for TA, the diagnosis depends predominantly on clinical findings, which should be subsequently confirmed by angiography. Initial isolated involvement of the pulmonary artery (PA) in TA is very rare and difficult to diagnose (8, 9). In the present study, we report the case of a young man whose initial symptom was PA occlusion and who manifested TA 6 years later as cerebral embolism. We consider that the source of the emboli was probably a left occluded CCA (suspected distal stump). The carotid stump is a potential source of emboli that results in transient ischemic attack or ischemic stroke (10, 11). To our knowledge, this is the first report of carotid stump embolism with TA.

Case Report

A 33-year-old man was admitted to our hospital with sudden onset of weakness on the right side. His medical history revealed a diagnosis of right PA occlusion based on progressive exertional dyspnea six years previously. Physical examination revealed a medium-sized man with clubbed fingers and no apparent distress. Pulse asymmetry between the right and left arms was absent and heart rate was regular and 80 beats/min. His blood pressure was 121/79 mmHg in the right arm, 125/80 in the left arm. Bruit was subtly audible over the course of the right subclavian artery. Arterial blood gases on room air revealed PaO₂ 76 mmHg and PaCO₂ 42 mmHg. Neurological examination demonstrated global aphasia and right-side hemiparesis with normal ocular fundi. Magnetic resonance (MR) imaging revealed infarction in the territory of the left middle cerebral artery (MCA) (Figs. 1A, 1B). Occlusion in the horizontal segment of the left MCA and an indistinct bilateral internal carotid artery (ICA) were revealed by MR angiography (MRA) (Fig. 1C). He was treated with intravenous edaravone and osmotic...
therapy. Routine laboratory tests, including fasting blood glucose and a lipid panel, were normal; the erythrocyte sedimentation rate (ESR) and C-reactive protein were 6 mm/h and <0.5 mg/dL, respectively. Antinuclear antibody, rheumatoid factor, anti-double-strand DNA antibody, anticardiolipin antibody, lupus anticoagulant, C-ANCA and P-ANCA, as well as protein C and S activities were all within normal limits. A conventional serological determination revealed A2, A24, B35 and B52 HLA loci. Chest X-ray revealed unremarkable findings (Fig. 2A). Electrocardiography showed normal sinus rhythm (Fig. 2B) and atrial fibrillation did not occur at any time during the clinical course including Holter monitoring. Transthoracic echocardiography for the detection of embolic source showed mild aortic regurgitation (AR) (Fig. 2C), normal left ventricular systolic function, no evidence of ventricular hypertrophy or pulmonary hypertension, and normal relaxation. Sonographic findings showed homogeneous, midechoic and circumferential wall thickening of the right ICA. Color Doppler findings showed occlusion of both the common carotid arteries (CCAs), filling of the both ICAs by reversal flow in the each external carotid artery (ECA) and turbulence at the ‘distal stump’ of the occluded CCA (Fig. 2D). Craniocephalic CT angiography on post-admission day 3 demonstrated occlusion of the bilateral CCAs ‘proximal stump’, but the bilateral ICAs were perfused with an extensive collateral circulation (Fig. 3A). Thoracoabdominal CT angiography on post-admission day 8 demonstrated right PA occlusion (Fig. 3B) and an intact renal artery. We diagnosed TA type I: P(+), which involves branches from the aortic arch and pulmonary artery (12). The absence of left MCA occlusion on repeated MRA on post-admission day 10 (Fig. 1D) suggested reperfusion after the emboli. Clinical symptoms were not improved by reperfusion. Angiography on post-admission day 20 confirmed bilateral CCA occlusion and bilateral collateralization from the vertebral artery (VA) via the posterior communicating artery (Pcom) to the MCA (Fig. 4A), via the occipital artery (OA) and retrogradely via the ECA to the ICA, and via the deep and ascending cervical arteries to the OA and ascending pharyngeal artery (APA) (Figs. 4B, 4C). In addition, the right inferior thyroid artery flowed mainly to the contralateral ICA via the retrogradely superior thyroid artery (Fig. 4D). Left MCA was not occluded (Fig. 4E). Contrast saline transcranial Doppler (TCD) examination on post-admission day 25 detected no right-to-left shunt, including patent foramen ovale and pulmonary arteriovenous fistula. The complex collateral and hemodynamic changes were produced by bilateral CCAs involvement in the TA. The patient was prescribed with clopidogrel.

**Discussion**

Takayasu’s arteritis is idiopathic disease that mainly affects the aorta and major aortic branches. Clinical symptoms of TA vary widely, depending on the site and degree of arterial lesions. Although subclinical PA involvement in TA is
Figure 2. A. Chest X-ray revealed unremarkable findings. B. Electrocardiography showed normal sinus rhythm. C. Transthoracic echocardiography showed mild aortic regurgitation. D. Sonographic and color Doppler findings showed the right ICA with homogeneous, midechoic and circumferential wall thickening, filling by reversal flow in the ipsilateral ECA and turbulence at the ‘distal stump’ of the occluded CCA (circle).

Figure 3. A. Craniocervical CT angiography on post-admission day 3 shows bilateral CCA occlusion ‘proximal stump’ (circles in the square inset), but bilateral ICA are perfused due to extensive collateral circulation. B. Thoracoabdominal CT angiography on post-admission day 8 shows right PA occlusion (arrow). CCA: common carotid artery, ICA: internal carotid artery, PA: pulmonary artery.

frequent, pulmonary symptoms caused by PA lesions are rare and typically occur as late manifestations (8, 13). The present patient demonstrated unusual symptoms associated with PA occlusion as the initial sign of TA. Most often, pulmonary involvement of TA has no or poor clinical impact, as it did in our patient (14). Some TA patients with initial severe PA involvement mainly show respiratory disorder, including chest pain, pleural effusion and hemoptysis (8, 9, 13). These symptoms are often difficult to distinguish from those of primary lung disease and have been initially diagnosed as primary pulmonary hypertension, chronic pulmonary embolism, tuberculosis (8, 13) and congenital PA occlusion, as in the present patient. The interval between the onset of symptoms and the diagnosis of TA in these other patients ranged from 1 to 16 years. However, the presence of PA lesions supports a diagnosis of TA when arteritis and atherosclerosis are difficult to differentiate (13).

Although 10 to 20% of patients with TA have ischemic stroke or transient ischemic attacks (15), reports of stroke as the first clinical manifestation before diagnosing TA and starting therapy are rare (4). Collateral blood flow is relatively well-developed in TA (16) which was evident in the present patient. Possible causes of ischemic stroke in TA include embolism from stenotic or occlusive lesions in the aortic arch and its main branches or cardiac diseases such as AR (1, 17). In addition, a recent case report has described TA with intracranial arteritis (6). The exact etiology of ischemic stroke in our patient is unknown. After excluding a cardiac source and embologenic changes in the other ves-
sels, we consider that the source of the emboli was probably a left carotid stump of the occluded CCA. The stump has both proximal and distal ends. The proximal stump is a demonstrable patent remnant of the occluded CCA, the distal stump is above the occluded CCA and continues to the patent ICA. Turbulent flow in both stumps can cause platelet-fibrin aggregation and theoretically lead to embolization (10). Carotid stump syndrome is defined as the persistence of cerebral or retinal ischemic symptoms after occlusion, usually of an ipsilateral ICA (11); however, our patient had an occluded ipsilateral CCA (18, 19). Stenotic or occlusive lesions in TA develop and may resemble arteriosclerosis except for a distribution (20). The possibility of emboli from the distal stump of the occluded CCA seems to be the most probable explanation, as turbulence at distal stump was detected by color Doppler echo. It was estimated that the emboli passed from the distal stump to patent ICA, and occluded MCA. Isolated instances of emboli arising from the distal end of a thrombus in an occluded carotid artery have been reported (21, 22). The other acceptable possibility can be the embolization from proximal stump of the occluded CCA through VA-Pcom-MCA or VA-OA-ECA-ICA-MCA, though the thrombus in the present case appeared too large to have traveled via collateral circulation. The present case suggests there can be potential stroke risk in TA even after CCA has become occluded.

In patients with TA, the proximal portion of a vessel is affected, with continuation of the pathological process into the more distal portion. During the acute phase, inflammatory infiltrates usually consist of lymphocyte, plasma cells, and giant cells and involve all layers of arterial walls. Thrombi are frequently observed distal to the site of inflammatory changes. At the chronic phase, fibrous tissue replaces the damaged intima, media, and adventitia. The previous study using TCD ultrasonography demonstrated that microembolic signals were detected not only in the active phase of TA but also in the chronic phase (23). In the present report, the patient developed cerebral infarction in spite of a normal ESR, suggesting that an active inflammatory process in TA could continue during the chronic phase and cause cerebral infarction by embolic materials.

We present herein the case of a young man whose initial symptom was PA occlusion and who manifested TA 6 years later as carotid stump embolism. The carotid stump can be a potential source of emboli in TA as well as in atherosclerosis. The frequency of this clinical form could be underestimated, given the difficulties of diagnosis and features that are similar to those of pulmonary diseases. As glucocorticoids are a potentially effective treatment, a diagnosis of TA should also be considered when another pulmonary disease is suspected, even when direct evidence of systemic artery involvement is absent.

Disclosure: The authors report no conflicts of interest

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