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## **Takayasu's Arteritis Complicated With Subarachnoid Hemorrhage and Hematomyelia**

—Case Report—

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### **Abstract**

**A 44-year-old woman presented with severe headache, drowsy mentality, and right hemiparesis. Brain computed tomography and magnetic resonance angiography revealed non-aneurysmal subarachnoid hemorrhage (SAH). Thoraco-abdominal and pelvic computed tomography angiography showed multiple steno-occlusive lesions involving the aorta and its large branches suggesting Takayasu's arteritis. Spine magnetic resonance imaging was taken because of prominent right hand muscle atrophy on the 14th hospital day, which showed subacute stage of hematomyelia in the cervical cord and conus medullaris. Aneurysmal or non-aneurysmal SAH is rare in patients with Takayasu's arteritis but SAH with coincidental hematomyelia is even more unusual. This case emphasizes the rarity of the coincidental spinal hematomyelia and its importance in the differential diagnosis.**

Key words: Takayasu's arteritis, subarachnoid hemorrhage, aneurysm, hematomyelia, diagnosis

### **Introduction**

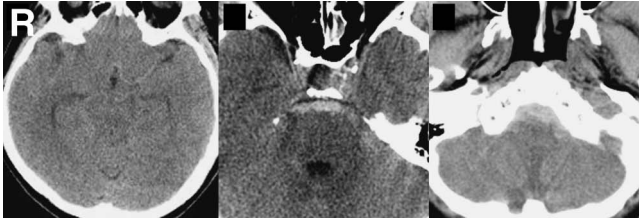
Takayasu's arteritis is an inflammatory vasculitis of unknown etiology that involves the aorta and its major branches, the pulmonary arteries and coronary arteries. Vascular inflammation may cause arterial stenosis, occlusion, thrombosis, dilatation, or aneurysms.<sup>2,12)</sup> Takayasu's arteritis affects young people, generally Asian women of reproductive age. Clinical manifestations vary depending

on the site of the blood vessels affected, organs involved, and collateral circulation. Cerebrovascular accident may be an important and predictive complication for the prognosis in such patients.<sup>2,4)</sup> We describe a woman who presented with non-aneurysmal subarachnoid hemorrhage (SAH) with coincidental hematomyelia and occlusion of the carotid or vertebral artery associated with Takayasu's arteritis.

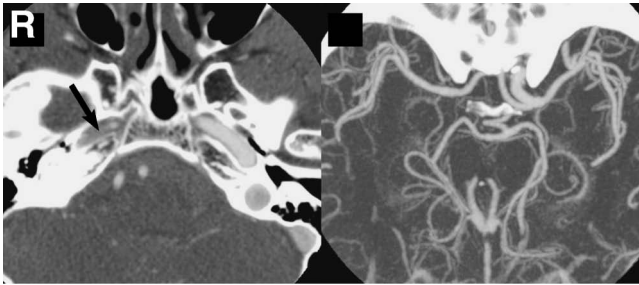
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**Fig. 1** Axial computed tomography scans showing subarachnoid hemorrhage in the prepontine and perimedullary cisterns.

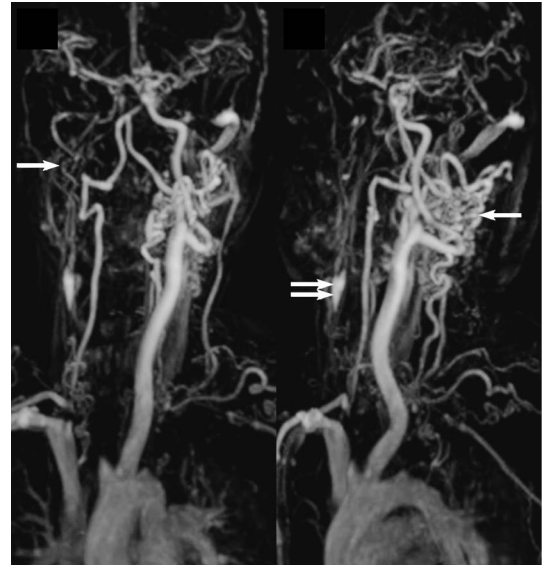


**Fig. 2** Computed tomography angiograms showing decreased blood flow in the right petrous portion of the right internal carotid artery (arrow). No intracranial aneurysm is noted.

### Case Report

A 44-year-old woman presented with sudden onset of severe headache and drowsy mentality while she was playing cards. She had suffered a mitral valve prolapse and had been taking antihypertensive medications over the last 5 years. On admission, she was drowsy with grade 2 right hemiparesis. Pain, temperature, and touch sensations were decreased on the right side of her body. Left foot and leg were deformed, and muscles were atrophic due to an accidental injury in her early life. Brain computed tomography (CT) demonstrated a small amount of SAH in the prepontine and basal cisterns (Fig. 1). Brain CT and magnetic resonance (MR) angiography showed significant stenosis of the basilar artery and bulbous dilatation of the left internal carotid artery, but no saccular aneurysmal lesion (Figs. 2 and 3). Our impression was basilar artery dissection. Lipid profile, coagulation study, hepatitis markers, cardiac markers, and electrocardiography were all within normal limits. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were initially normal but later constantly elevated: ESR (> 30 mm/hr) and CRP (> 1 mg/dl), and mild leukocytosis (10,000–15,000/ $\mu$ l) was noted.

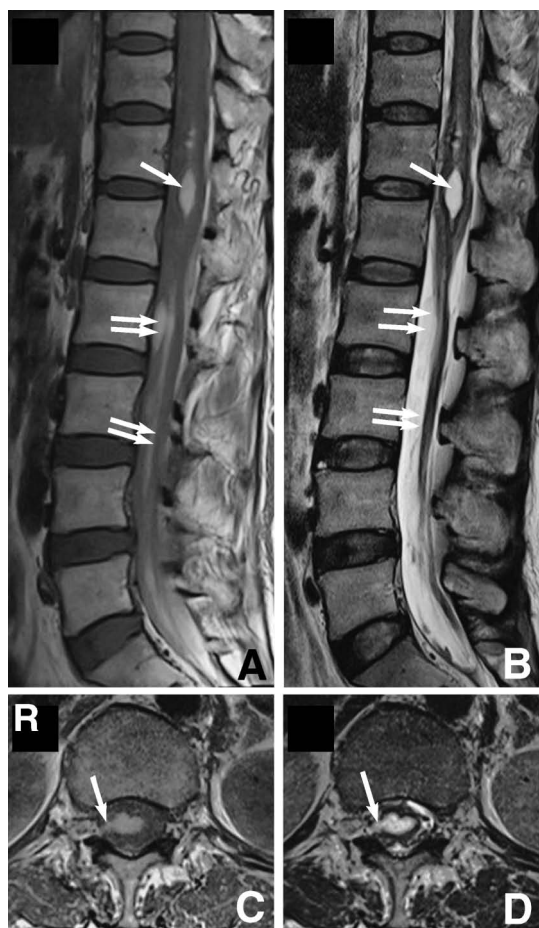
On the 2nd hospital day, she began to complain of abdominal and flank pain. Thoraco-abdominal and pelvic CT angiography showed segmental luminal narrowing of the distal thoracic aorta, marked stenosis of the celiac axis, and near total occlusion of the right brachiocephalic and left subclavian arteries with distal filling by collateral supply. The distal thoracic aorta showed focal stenosis



**Fig. 3** Brain and neck magnetic resonance angiograms showing the occluded right brachiocephalic and left subclavian arteries with multiple collateral vessels (arrows), and partial filling of the right distal common carotid artery (double arrow).



**Fig. 4** Sagittal T<sub>1</sub>-weighted (A) and T<sub>2</sub>-weighted (B) magnetic resonance (MR) images showing subacute stage hematomyelia in the cervical spinal cord (arrows). Axial T<sub>2</sub>-weighted (C) and gradient-recalled echo T<sub>2</sub>-weighted (D) MR images depicting hematomyelia in the right anterior and posterior gray column (arrows).



**Fig. 5** Sagittal T<sub>1</sub>-weighted (A) and T<sub>2</sub>-weighted (B) magnetic resonance (MR) images showing subacute stage hematomyelia in the conus medullaris (arrows) and intradural-extramedullary space (double arrows). Axial T<sub>1</sub>-weighted (C) and T<sub>2</sub>-weighted (D) MR images at the conus medullaris level depicting rupture of hematomyelia into the subarachnoid space (arrows).

with prominent collaterals such as bronchial or intercostal arteries. Bilateral pleural effusion and dependent atelectasis were also noted. Her carotid and femoral pulsations were very weakly palpated bilaterally. Her brachial blood pressure was 114/66 mmHg on the right and 121/69 mmHg on the left. Her pedal blood pressure was 80/57 mmHg on the right and 95/68 mmHg on the left.

Under the impression of Takayasu's arteritis and SAH, she underwent percutaneous stent insertion into the distal thoracic aorta. However, stent insertion into the right brachiocephalic artery failed because of marked tortuosity of the artery. Steroid therapy was started and the motor power of her right arm was improved to grade 4, but sensations and right leg weakness were unchanged. On the 14th hospital day, her prominent right hand intrinsic muscle atrophy led us to perform spinal MR imaging, which showed subacute stage of hematomyelia in the cervical cord (C2–C7) and conus medullaris (T12–L1) (Figs. 4 and 5). The hematoma in the conus medullaris had ruptured out into the spinal subarachnoid space. Her neurological

status was stable until discharge on the 66th hospital day.

## Discussion

Patients with Takayasu's arteritis usually have constitutional symptoms such as fever, general malaise, and weight loss before the development of various ischemic symptoms. ESR and CRP are usually elevated in the acute phase but remain normal in one third of patients.<sup>9)</sup> As the disease progresses, the subclavian, innominate, carotid, vertebral, coronary, renal, and other arteries are progressively occluded. As a result, ischemic vascular symptoms are most common depending on the affected arteries. Acute or chronic abdominal pain may be the presenting symptom due to involvement of the mesenteric artery.<sup>8,13)</sup> The most common symptoms are claudication of the legs, and weakness and coldness of the arms. If the renal arteries are involved, hypertension may develop, and if the carotid and vertebral arteries are involved, neurological signs and symptoms may develop.

Most neurological symptoms are due to occlusion of the cervical carotid and/or vertebral arteries, but are rarely reported,<sup>9)</sup> possibly due to the abundant cerebral arterial collateral vessels. Takayasu's arteritis rarely presents with aneurysmal or non-aneurysmal SAH.<sup>2,5)</sup> The relationship between cerebral aneurysm formation and Takayasu's arteritis has not yet been clearly defined. Hemodynamic stress on the cerebral arteries, immune destruction and necrosis of the elastic lamina, and hypertension may be causes of aneurysm formation in Takayasu's arteritis.<sup>5,13)</sup> However, aneurysmal SAH may be an incidental event unrelated with Takayasu's arteritis because the incidence of cerebral aneurysm in Takayasu's arteritis is not different from the natural incidence and microscopic studies of the aneurysms have shown no vascular inflammation.<sup>6)</sup>

If pathological processes of the intracranial arterial wall are involved in aneurysm formation in Takayasu's arteritis, dissecting aneurysm may also be formed and result in SAH. Our patient had SAH mainly in the prepontine cistern and her basilar artery was very stenotic compared with the dilated vertebral arteries. Some patients with Takayasu's arteritis present with aneurysmal or non-aneurysmal SAH, so our impression on admission was dissecting basilar artery or non-aneurysmal SAH. However, the spinal MR imaging, taken 2 weeks later, showed hematomyelia in the cervical cord and conus medullaris. The immediate right hemiparesis and subsequent right hand muscle atrophy were thought to be due to cervical hematomyelia. Spontaneous onset of hematomyelia is usually due to vascular malformation in the spinal cord, intramedullary cavernoma, intrasyringal hemorrhage, hematologic disorders, or anticoagulant therapy.<sup>1,9)</sup> However, none of these were present in our patient. SAH with coincidental hematomyelia is extremely unusual, even compared to the rarity of aneurysmal or non-aneurysmal SAH in patients with Takayasu's arteritis.

Our patient immediately developed dense right hemiparesis despite the small amount of SAH, so the cause of hemiparesis was thought to be cervical hematomyelia. Since the hematomyelia of the conus medullaris ruptured

into the spinal subarachnoid space without visible cerebral saccular aneurysm, the cranial SAH might be due to basilar artery dissection or ruptured hematoma from the conus medullaris into the spinal subarachnoid space. The subarachnoid blood was mainly located in the preponine cistern, supporting the possibility of basilar artery dissection. Whether the bleeding in the cervical cord and conus medullaris occurred simultaneously with intracranial SAH is still unknown due to the lack of initial spinal CT or MR imaging.

Based on our speculation of hematomyelia and Takayasu's arteritis, we suggest several plausible causes of hematomyelia as follows: hemorrhagic infarction due to embolism; hemorrhagic conversion secondary to spinal cord infarction; and systemic vasculitis affecting small vessels such as the radiculomedullary arteries. From the neurosurgical point of view, Takayasu's arteritis is associated with cerebral and spinal cord infarctions. In a previous longitudinal study of 16 patients with Takayasu's arteritis, two patients experienced cerebral infarction but none had spinal infarction.<sup>8)</sup> Another series included one patient (5%) who suffered paraplegia due to spinal infarction.<sup>7)</sup>

However, whether the hematomyelia resulted from systemic pathologic processes in the arterial walls associated with Takayasu's arteritis remains unclear. Systemic vasculitis can present as a tumor-like lesion although not usual. Granulomatous vasculitis of tumor-like lesion associated with spinal cord compression has been reported in patients with Takayasu's arteritis, sarcoidosis, giant cell arteritis, and Wegener's granulomatosis.<sup>3,10,11)</sup> Takayasu's arteritis is now known to affect small vessels as well, although radiculomedullary vessel involvement has not been reported.<sup>12)</sup> Therefore, we suggest that the multi-centric hematomyelia of our patient probably developed from inflammation associated with Takayasu's arteritis.

Aneurysmal or non-aneurysmal SAH occurs rarely in the patients with Takayasu's arteritis, but this case emphasizes the rarity of the coincidental spinal hematomyelia and its importance in the differential diagnosis.

### Declaration of Interest

None of the authors have any financial interest in the subject under discussion in this paper.

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