

Takayasu's Arteritis Presented with Subarachnoid Hemorrhage : Report of Two Cases

Takayasu's arteritis is a chronic inflammatory disease that produces a narrowing of the aorta and its major branches. Fibrosis and thickening of the arterial wall often occur in later stages, resulting in a cerebrovascular accident. The authors report two young women patients who presented with subarachnoid hemorrhage (SAH) and occlusive cerebrovascular disease associated with Takayasu's arteritis. Both patients had sudden headache and hemiparesis. Physical examination showed weak radial pulse, carotid bruit, and asymmetrical blood pressure. Erythrocyte sedimentation rate (ESR) was elevated in both patients. SAH was confirmed by brain computerized tomography (CT) or lumbar puncture. Occlusive cerebrovascular disease was diagnosed by brain magnetic resonance imaging (MRI), brain magnetic resonance angiography (MRA), and cerebral angiography. The findings of aortography and cerebral angiography were compatible with Takayasu's arteritis, but intracranial aneurysm was not found in either patient.

Key Words : *Takayasu's Arteritis; Subarachnoid Hemorrhage; Blood Sedimentation; Aortography, Diagnosis*

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INTRODUCTION

Takayasu's arteritis is an inflammatory disease of unknown etiology that involves the aorta and its major branches. Takayasu's arteritis affects young people, generally Asian women. Clinical manifestations vary depending on the site and severity of occlusive vascular lesion (2, 14, 15, 19). Cerebrovascular accident may be an important and predictive complication for the prognosis in such patients (4).

In this report, we describe two women who presented with subarachnoid hemorrhage (SAH) of unknown origin and occlusion of the carotid or the vertebral artery associated with Takayasu's arteritis.

CASES REPORT

Case 1

A 44-yr-old woman presented with drowsy mentality after a sudden bursting headache and vomiting on the day before admission. She had been on medication for hypertension and cerebral infarction since her left hemiparesis had developed 5 yr before. Left side weakness was gradually improved.

On admission, she had asymmetrical blood pressure (130/100 mmHg on the right arm and 100/80 mmHg on the right leg), both carotid bruits, and weak pulsations of both

radial arteries. She showed multiple cranial nerves (III, IV, VI, IX, X) palsy and left hemiparesis (arm/leg: Grade II/Grade III). Peripheral white blood cell count was 20,500/ μ L and ESR was 39 mm/hr. Echocardiogram showed moderate to severe concentric left ventricular hypertrophy. SAH and massive intraventricular hemorrhage were found on brain CT (Fig. 1A, B). Aortography showed proximal occlusion of both subclavian arteries (Fig. 1D). Both vertebral arteries were also occluded at their proximal levels (Fig. 1D). Multiple narrowings of the abdominal aorta and proximal occlusion of the right renal artery were shown (Fig. 1E). Bilateral carotid angiographies showed proximal narrowings of external carotid arteries and posterior circulation supplied from collateral through the circle of Willis. Upper portion of the basilar artery was visualized on the right internal carotid angiography (Fig. 1F) and the second segment of the vertebral artery was visualized on the left external carotid angiography via through the posterior cervical branches. There was no evidence of intracranial aneurysm. Brain MRA obtained three day following the admission showed no evidence of aneurysm or vascular malformation, but T2-weighted image revealed high signal intensity in the pons, which was slightly increased in size compared with the previous MRI that was obtained 5 yr before (Fig. 1C).

Steroid therapy was initiated on her admission. She fully recovered consciousness and her left side weakness was improved (Grade IV/Grade IV). But multiple cranial nerve (IV, IX, X) palsy remained.

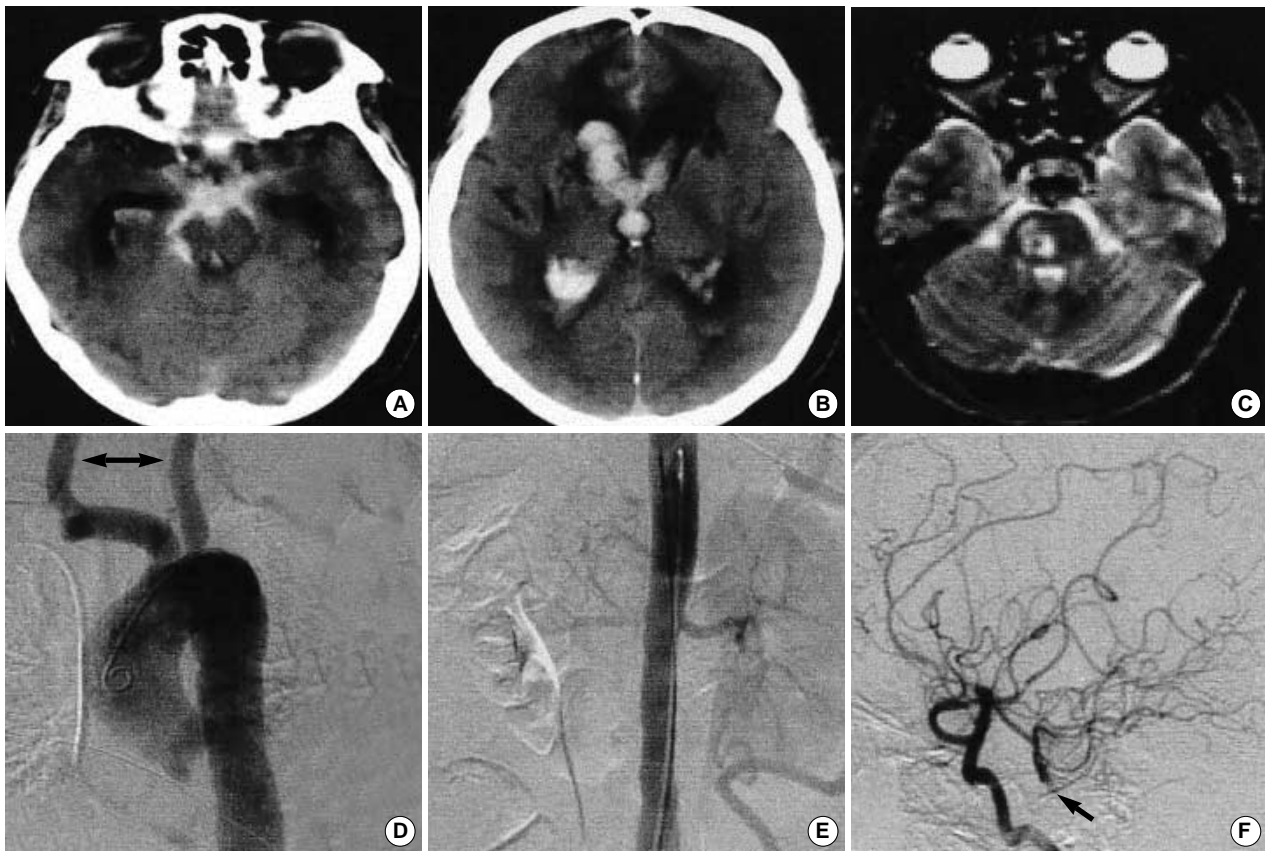


Fig. 1. (A) and (B): Brain axial CT revealing SAH in basal cistern and intraventricular hemorrhage in the lateral, the third and the fourth ventricle. (C) Axial T2 weighted image of brain MRI obtained 5 yr ago is showing high signal in the pons. (D) and (E) Aortography: Both subcalvian arteries and both vertebral arteries are not visualized but both carotid arteries (arrow) are well visualized. Multiple focal narrowing of abdominal aorta and proximal occlusion of the right renal artery are shown. (F) Right internal carotid angiography (lateral projection): Upper portion of the basilar artery (arrow) is visualized but intracranial aneurysm is not shown.

Case 2

A 31-yr-old woman experienced sudden headache and left hemiparesis 7 days before admission, which had been improving without any medical treatment. She experienced again sudden headache with nausea and vomiting on the day before admission. She was transferred to the emergency room of because of her headache and remained mild motor weakness.

On admission, her blood pressure was 87/46 mmHg on the left upper arm and 120/60 mmHg on the right upper arm. Physical examination revealed carotid bruit and carotid tenderness on the left side and weak pulsation of left radial artery. Neck stiffness was equivocal. Left hemiparesis was Grade IV⁰. Deep tendon reflex was hyperactive on the left side. Brain computerized tomography (CT) showed no evidence of SAH, but lumbar puncture revealed xanthochromic cerebrospinal fluid. Peripheral white blood cell count was 10,900/ μ L and erythrocyte sedimentation rate (ESR) was 38 mm/hr. Echocardiogram showed no definitive abnormality. Aortography and cerebral angiography showed diffuse narrowing of the left axillary artery and brachial artery with abun-

dant regional collateral arteries on the left shoulder area. Stenosis and dilation of the left subclavian artery were identified distal to the origin of the left vertebral artery (Fig. 2F). Diffuse narrowing of the left internal and external carotid arteries (Fig. 2A) and proximal occlusion of the right common carotid artery were shown. Right cerebral hemisphere was supplied with collaterals through the left internal carotid artery and vertebrobasilar system (Fig. 2B, C). No intracranial aneurysm was found on cerebral angiography (Fig. 2B-E). Brain MRA and MRI did not show the right internal carotid artery and the right middle cerebral artery (Fig. 2G) and the evidence of hemorrhagic transformation of cerebral infarction.

Steroid therapy was initiated on her admission. But she refused further treatment and discharged on the 12th day after admission.

DISCUSSION

Takayasu's arteritis (pulseless disease) is a chronic inflammatory disease that involves large trunks and branches of the

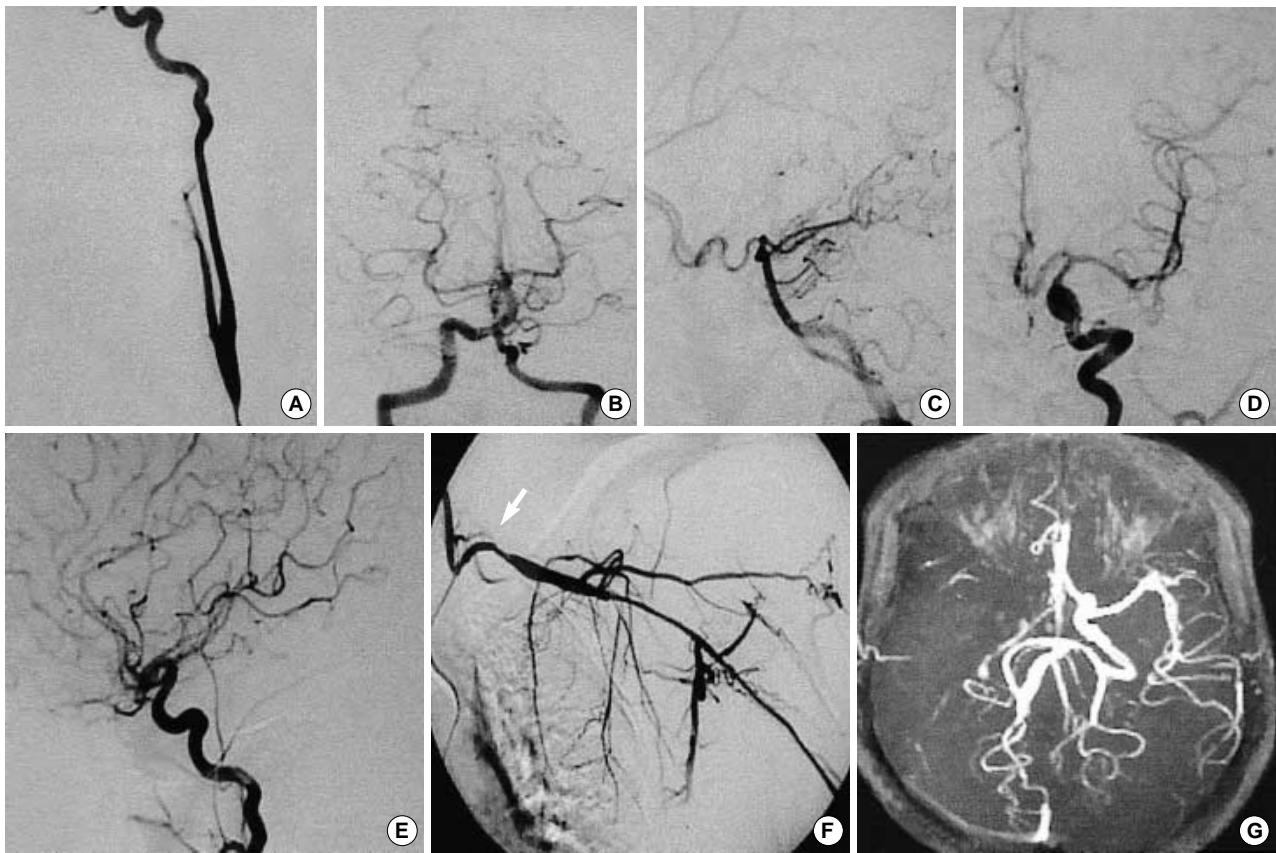


Fig. 2. (A) Left common carotid angiography: Diffuse narrowing of the left internal and external carotid arteries. (B, C, D) and (E) Vertebral and left internal carotid angiography, Towne and lateral projection: No intracranial aneurysm is shown. (F) Left subclavian angiography: Stenosis (arrow) and dilatation of the left subclavian artery distal to the origin site of the left vertebral artery. (G) Brain MRA: Right carotid artery and right middle cerebral artery are not visualized. Also no intracranial aneurysm is shown.

aortic arch and abdominal aorta, leading to a severe narrowing or occlusion of large arteries and ischemic symptoms. Clinical manifestations vary depending on the sites and severity of the occlusive vascular lesions (2, 14, 15, 19). Neurologic dysfunction due to cerebrovascular accident is an important component of Takayasu's arteritis and may be a presenting manifestation but more often it occurs later in the disease course (1, 13, 19). However, cerebrovascular disease has not been reported to be a frequent complication despite multiple occlusion of major cervical arteries and recurrence of stroke is uncommon, probably due to the gradual development of abundant collaterals (15). Thyrocervical trunk, anterior spinal arteries, several unnamed vessels, or the circle of Willis may play an important role (14, 15). Minor ischemic stroke is more frequently observed in the early stage of Takayasu's arteritis (within one to two years after development of arteritis), which may be related to insufficient collaterals (15). Also cerebral hemodynamics may be chronically insufficient in Takayasu's arteritis (15, 19) and a small reduction in blood pressure may develop cerebral ischemia even in the chronic stage of the disease (21).

Although intracranial bleeding or aneurysmal rupture may

be the cause of mortality in Takayasu's arteritis (9, 17), hemorrhagic stroke including SAH has not been reported frequently compared with ischemic stroke. Most cases of SAH in Takayasu's arteritis have been reported to be related with intracranial aneurysmal rupture. Based on a literature review, the total number of SAH due to a proven intracranial aneurysmal rupture was 25 aneurysms in 16 patients (3, 5, 7-12, 16, 20, 22). Four out of sixteen patients with SAH due to ruptured intracranial aneurysm had occlusive cerebrovascular disease (3, 5, 9, 12). But whether the aneurysms and Takayasu's arteritis occur independently or whether the aneurysm is related with Takayasu's arteritis is unknown. Shigemori et al. pointed out the importance of altered hemodynamic forces on the circle of Willis produced by agenesis of the internal carotid artery as one of the causative effects of the formation of aneurysms (18). Masuzawa et al. reported two cases of intracerebral aneurysms occurring with Takayasu's arteritis. They concluded that abnormal intracranial hemodynamic forces play a role in the development of the aneurysm (9). Immune destruction of elastic lamina in Takayasu's arteritis may be a possible mechanism. Necrosis of the vessel wall with rupture and hemorrhage may also occur (6). Sano and

Saito found no aneurysmal formation or subarachnoid hemorrhage in either clinical or radiological examination in up to 77 cases of Takayasu's arteritis. Therefore they concluded that the immune mechanism plays an important role in the development of the disease (15). However, no evidence of arteritis in intracranial vessels nor any pathological change in the intracranial arteries adjacent to the aneurysm has been reported in Takayasu's arteritis (9).

In our cases, SAH was confirmed by lumbar puncture or brain CT but no intracranial aneurysm was found by cerebral angiography. MRA showed neither intracranial aneurysm nor vascular malformation.

Takayasu's arteritis should be suspected in young women presenting with neurologic dysfunction with abnormal finding of cardiovascular examination and elevation of ESR. The aortography and cerebral angiography should be performed for a definitive diagnosis. Based on our cases, SAH associated Takayasu's arteritis was not necessarily related with intracranial aneurysm.

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