

## Clinical Notes

### Subarachnoid hemorrhage revealing Takayasu's disease

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Takayasu arteritis (TA) is a chronic inflammatory disease of large arteries, usually affecting the aorta and its large branches and the pulmonary arteries. It is a vasculitis and can manifest systemically, involving any or all the major organ systems.<sup>1</sup> Fibrosis and thickening of the arterial wall often occur at later stages, resulting in a cerebrovascular accident. A cerebrovascular accident may be an important and predictive complication for the prognosis in TA disease.<sup>2</sup> Subarachnoid hemorrhage (SHA) has not been reported frequently compared with ischemic stroke in these patients. Non-aneurysmal SHA revealing Takayasu's disease is uncommon.<sup>3</sup> A case of TA revealed by SHA is reported.

A 25-year-old Moroccan woman was admitted to the Neurosurgery Department of Mohamed V Military Hospital, Rabat, Morocco, with a 5-day brutal headache and trouble in consciousness with nuchal stiffness. She had been on medication for hypertension since her pregnancy with eclampsia one year ago. On admission she had a Glasgow Coma scale of 12, with no deficit but asymmetrical blood pressure (175/120 mm Hg on the right arm and 150/85 mm Hg on the left arm). No pulse on right radial artery and feeble pulse on left radial artery. No carotid bruit was noted. Cardiac examination was normal. Somatic examination showed no abnormalities. Cranial CT scan demonstrated hypodense lesion of the left putamen and capsule consistent with infarction with frontal ventricle horn retraction with suspected SHA. The SHA was confirmed by lumbar puncture. Aortography and cerebral angiography showed proximal stenosis of the right subclavian artery (**Figure 1**). No intracranial artery aneurysm was identified. The extracranial carotid arteries and thoracic aorta were normal. These findings were compatible with Takayasu's disease. An immunologic work-up gave the following results: lab peripheral white blood cell count was 14,000/mm (normal 9500-10,000) and erythrocyte sedimentation rate (ESR) was elevated at 60 mm in the first hour. C-Reactive protein was 12.2 mg/l, (normal < 4 mg/l). Lab study complement 3 (C3) dosage was normal: 1.39 g/l and complement C4 was normal. Rheumatoid factor, antinuclear antibodies, anticardiolipin antibodies, and antineutrophil cytoplasmic antibodies were negative. The venereal disease research test, and treponema pallidum hemagglutination assay was negative. Intra-dermo reaction to 10% tuberculin was 12 mm at 72 hours. Exhaustive research of *Mycobacterium tuberculosis* in gastric aspiration, spit, and urine were

unable to confirm any tuberculosis localization. Protein electrophoresis showed a hypoalbuminemia at 33.8 g/l (43-51). Alpha 1, alpha 2 are elevated at 4.1 g/l and 10.1 g/l (normal reference 1-2 and 5-8). Gamma globulin was elevated: 16.5 g/l (6-11). Other laboratory data did not reveal any relevant abnormalities. The ECG and echocardiography were normal. The patient was treated by corticotherapy. Follow up was unremarkable. She was oriented to the internal medicine department for complement investigation and follow-up.

Takayasu arteritis is a chronic vasculitis of the aorta and its primary branches. Cases have been reported worldwide, but TA is most common in Southeast Asia and the Indian subcontinent. Takayasu arteritis is primarily a disease of young women. Men rarely are affected. For the classification of TA, the American College of Rheumatology requires 3 of the following 6 criteria to be met: age at disease onset  $\leq 40$  years, claudication of extremities, decreased brachial artery pressure, blood pressure difference  $>10$  mm Hg, bruit over subclavian arteries or aorta, and abnormal arteriogram.<sup>1</sup> These criteria are not met in early disease. Usually, there is a 'prestenotic stage' ('prepulseless stage') with the symptoms of fatigue, malaise, arthralgia, myalgia, weight loss, low-grade fever, and an elevated ESR. This phase can persist for months or even years before characteristic stenoses occur (pulseless stage). As no bruits or other signs of arterial obliteration are found in the prestenotic stage, it is very difficult to establish the diagnosis of early Takayasu's disease. Takayasu 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 occurs



**Figure 1** - Aortogram showing stenosis of proximal right subclavian artery.

in later stages, resulting in a cerebrovascular accident. Clinical manifestations vary depending on the sites and severity of the occlusive vascular lesions. The etiology of TA is unknown. The underlying pathologic process is inflammatory, with several etiologic factors having been proposed, including spirochetes, *Mycobacterium tuberculosis*, streptococcal organisms, and circulating antibodies due to an autoimmune process. In a case report, the role of *Mycobacterium tuberculosis* and its 65-kDa heat shock protein has been implicated in the etiology. Patients with TA were found to have higher immunoglobulin (Ig) G, IgM, and IgA titers against the *Mycobacterium tuberculosis* extract than patients without the condition. Our patient had a positive intradermoreaction with 10% tuberculin without any tuberculosis. Neurological dysfunction due to cerebrovascular accident is an important component of TA and may be a presenting manifestation, but more often it occurs later in the disease course. However, cerebrovascular disease has not been reported to be a frequent complication, despite multiple occlusion of major cervical arteries recurrence of stroke is uncommon, probably due to the gradual development of abundant collaterals.<sup>2</sup> Thyrocervical trunk, anterior spinal arteries, several unnamed vessels, or the circle of Willis may play an important role. Minor ischemic stroke is more frequently observed in the early stages of TA. Although intracranial bleeding or aneurysmal rupture may be the cause of mortality in TA, hemorrhagic stroke including SAH has not been reported frequently compared with ischemic stroke. Most cases of SAH in TA have been reported to be related to intracranial aneurysm rupture. Based on the literature review, the total number of SAH due to a proven intracranial aneurysm rupture was 25 aneurysms in 16 patients.<sup>4</sup> Four out of 16 patients with SAH due to ruptured intracranial aneurysm had occlusive cerebrovascular disease. But whether the aneurysm and TA occur independently, the co-existence of TA and aneurysm can be fortuitous. However, some others authors pointed out the role of

altered hemodynamic forces on circle of Willis. The role of abnormal intracranial hemodynamic forces plays a role in the development of the aneurysm. Immune destruction of lamina of elastic lamina in TA may be a possible mechanism. Immune mechanisms play an important role in the development of disease. However, no evidence of arteritis in intracranial vessels or any pathological change in the intracranial arteries adjacent to the aneurysm was reported in TA.<sup>2-4</sup>

The SAH revealing TA disease is very rare. The TA should be suspected in young women with neurological dysfunction with abnormal findings on cardiovascular examination and elevation of ESR. A SAH is not necessarily associated with aneurysm bleeding, but it must be eliminated by intracranial angiography. In this case, no intracranial aneurysm was found in exhaustive and 4 pedicles study of circle of Willis.

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