

Amaurosis and Pulselessness in a Young White Woman: A Case of Takayasu Disease

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Takayasu disease, first described by a Japanese ophthalmologist in 1908, is a chronic inflammatory arteriopathy characteristically diagnosed in young Oriental women.¹ It might be more common among the white population than previously appreciated, as its incidence is estimated to be 2.6 in 1,000,000 each year. The median delay from the first symptom to the time of diagnosis in the United States is 18 months.² The underappreciation of this disease might contribute to this delay in diagnosis. We describe a young white woman whose ocular symptoms led to the diagnosis of Takayasu disease.

Case Report

A 33-year-old North American white woman complained to her ophthalmologist of recurrent episodes of painless loss of vision in the right eye. Each episode lasted approximately 1 minute, and she had experienced two to three episodes each month for the preceding 6 months. Her symptoms were associated with episodes of migraine-like headaches that relieved spontaneously after 20 minutes. She admitted to smoking two packs of cigarettes a day but denied using cocaine. Her medical history was unremarkable except for occasional migraines and an episode of bronchitis. She was not taking any medications. She had no family history of premature atherosclerosis or lipid disorders.

Findings on an ophthalmic examination were within normal limits, including vision, pupils, ocular motility, and confrontational visual fields. She had normal vessels, macula, and peripheral retina in both eyes on a dilated fundus examination. The optic discs appeared pink and healthy.

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Systolic bruits were heard bilaterally on carotid auscultation. Her radial pulses were markedly diminished and asymmetric. Her blood pressure could not be detected in the upper extremities but was measured to be 140/108 mmHg in the lower extremities. Cardiac, musculoskeletal, and neurologic findings were normal. Tests for lupus erythematosus, coagulopathy, syphilis, tuberculosis, and anticardiolipin antibodies had negative results.

Echocardiography showed no source of emboli. Arteriography showed a high-grade irregular narrowing of the innominate and left subclavian arteries. There was reduced flow in the right common carotid and subclavian arteries. A biopsy specimen from the innominate artery showed lymphocyte, plasma cell, and fibroblast infiltration consistent with Takayasu disease. Langhans giant cells, which occasionally accompany the inflammatory cells in Takayasu disease, were not visualized.³ Surgical revascularization was performed by aortic innominate bypass using a soft woven Dacron graft. The episodes of amaurosis ceased following the procedure, and the patient has had no visual symptoms since then. Three years after this procedure she did, however, develop lesions in other midsize arteries, including the left subclavian and coronaries, which required multiple bypass procedures.

Discussion

Not all family physicians will see a case of Takayasu disease, but they will encounter symptoms and physical findings that require considering it as part of the differential diagnosis. The vigilant physician who makes the diagnosis will earn the patient's gratitude, for Takayasu arteritis is a devastating disease with a 10-year mortality of 15 to 20 percent. Prompt diagnosis and treatment can be lifesaving. Vascular surgery or systemic steroids have been the traditional treatment choices.

Before considering Takayasu disease as the diagnosis, other infectious and inflammatory causes

of arteriopathy, such as syphilis, tuberculosis, and lupus erythematosus, must be ruled out. Coagulopathies, antiphospholipid antibody syndrome, and possible arterial embolic disease should also be considered. Our patient tested negative for all these disease processes.

Amaurosis, which was her original symptom, is a reported clinical feature in 13 percent of the cases. Other, more common clinical features are carotid bruits (72 percent), myalgia or arthralgia (56 percent), arm claudication (47 percent), fever (44 percent), postural dizziness (41 percent), and headache (28 percent). Arterial bruits are uncommon in young patients, and their occurrence in association with these symptoms should alert clinicians to the possibility of Takayasu arteritis. When a patient has these symptoms, the physician should be sure to auscultate the carotid arteries and palpate the radial pulses. Because of compromised peripheral pulses, hypertension can go unrecognized until late in the course of the disease. Hence, some of these patients might have hypertensive end organ damage, such as retinopathy, without any antecedent history of hypertension. Young women are most affected by migraines, which can sometimes be associated with visual symptoms. Our patient's amaurotic episodes were associated with migraine-like headaches. These patients deserve to be examined

carefully before ascribing the visual symptoms to migraine.

Findings from angiography help to confirm Takayasu disease. The combination of the arteriographic pattern and location of the lesions is usually diagnostic.² Characteristic patterns include irregular vessel walls, stenosis, poststenotic dilatation, aneurysm formation, occlusion, and evidence of increased collateral circulation.⁴ Histopathologic evidence of inflamed vessels add confirmatory data.⁴ We need to have a heightened awareness of Takayasu disease, an uncommon entity but one that can mimic other common and less threatening diseases.

References

1. Subramanyan R, Joy J, Balakrishnan KG. Natural history of aortoarteritis (Takayasu's disease). *Circulation* 1989;80:429-37.
2. Hall S, Barr W, Lie JT, Stanson AW, Kazmier FJ, Hunder GG. Takayasu arteritis. A study of 32 North American patients. *Medicine* 1985;64:89-99.
3. Titus J, Kim H. Blood vessels and lymphatics. In: John M. Kissane, editor. *Anderson's pathology*. 9th ed. St. Louis: Mosby, 1990:774.
4. Fauci SA. The vasculitis syndromes. In: Isselbacher KJ, Braunwald E, Wilson JD, Martin JB, Fauci AS, Kasper DL, editors. *Harrison's principles of internal medicine*. 13th edition. New York: McGraw-Hill, 1994:1677.