

Takayasu's Arteritis

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ABSTRACT Takayasu's arteritis is a chronic vasculitis of unknown etiology that typically occurs in women. It affects the aorta and its main branches, leading to either arterial stenoses or aneurysms. Here we report a case of a 42-year-old woman presenting with loss of peripheral vision in her left eye. On ultrasound imaging, she had distinctive findings of Takayasu's arteritis with marked circumferential thickening of the intimal-medial layer in the left common carotid artery and characteristic sparing of the carotid bulb and internal carotid artery. The diagnosis was further supported by computed tomography findings of aneurysms in the subclavian artery and aorta. Awareness of these ultrasound findings may be of particular importance to many echocardiographers who are extending their practice into vascular imaging.

Case Report

A 42-year-old woman, a former tobacco smoker with no history of hypertension, diabetes mellitus, or hyperlipidemia, was referred for carotid duplex ultrasonography after the loss of peripheral vision in the left eye.

B-mode images showed marked circumferential increase in intimal-medial thickness (up to 2.0 mm) in the left common carotid artery (CCA) and proximal left subclavian artery (SCA). Intimal-medial thickness was normal in the right CCA (Figure 1 and Video 1). No structural abnormalities were seen in the visualized portions of remaining neck arteries on either side. Doppler ultrasonography demonstrated no hemodynamically significant stenosis in any of the neck arteries on either side, including the left CCA or SCA.

The finding of localized homogeneous circumferential arterial thickening of the proximal CCA or subclavian arteries with sparing of the carotid bulb and internal carotid artery in a young female was consistent with Takayasu's arteritis.

After this presumptive diagnosis, noncontrast computed tomography (CT) imaging of the chest and abdomen was performed. Focal aneurysmal dilation of the left subclavian artery was seen, measuring approximately 1.9 cm in diameter. Also aneurysmal was the proximal left subclavian artery, measuring 1.6 cm in diameter.

A contrast-enhanced CT angiogram of the abdominal aorta and pelvic arteries showed two distinct aneurysms involving the distal thoracic and proximal abdominal aorta (Figure 2). The first aneurysm, located at the distal thoracic aorta, had a maximal diameter of 4 cm and a length of 7.5 cm. The second aneurysm, located at the level of the renal arteries, had a maximal diameter of 4.7 cm and a length of 5.8 cm. The aortic segment between the two aneurysms was normal. The remaining distal 9.4 cm of the aorta and the iliac arteries were also normal.

Discussion

Takayasu arteritis (TA), first described in 1908¹ and first named in 1942,² is a chronic and rare inflammatory vasculitis affecting large vessels with unknown etiology.^{3,4} Although TA is rare, its incidence is distributed globally,⁵ with the greatest prevalence seen in Japan, South East Asia, India, and Mexico.⁶ The incidence in Europe and the United States ranges from 0.12 to 0.26 per million per year.^{7,8} In Japan, TA is considered an intractable disease and a database is

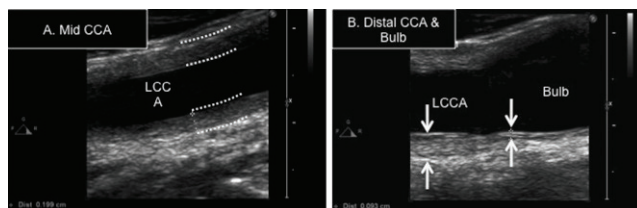


Figure 1

B-mode carotid ultrasonography of left carotid artery. (A) Marked circumferential intimal-medial thickening in the mid portion of the left common carotid artery. (B) Marked-intimal medial thickening in the distal left common carotid artery that abruptly tapers to normal thickness in the left carotid bulb.

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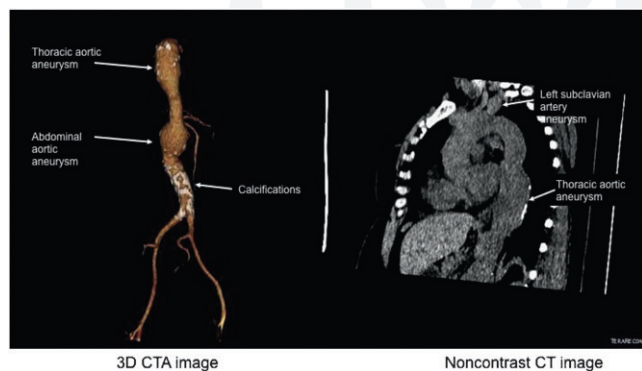


Figure 2

Three-dimensional computed tomography angiography (left) and noncontrast two-dimensional computed tomography (right) showing aortic and left subclavian artery aneurysms.

maintained by the government, with approximately 5,000 patients registered as of 1996.⁹ Disease presentation commonly occurs in the second or third decade of life, and diagnosis is often delayed. From a study of 107 TA patients, 80% of participants were between 11 and 30 years of age and 77% of those were between 10 and 20 years when disease symptoms presented.¹⁰ In 78% of the patients, diagnosis took from 2 to 11 years.¹⁰ The disease was once believed to be limited to women, but has since been shown to present in both sexes worldwide, with various manifestations in different populations.³

The aorta and its principal arch branches, such as the CCA, SCA, and coronary arteries, commonly are affected, and, less commonly, the pulmonary arteries. TA may lead to either arterial stenosis or aneurysm formation.

Arterial stenosis is caused by inflammation of vessel walls and thrombus formation, which may lead to significant flow obstruction. These characteristics gave rise to the term “pulseless disease.”¹⁰ Monocytes and T cells enter the arterial wall through the vasa vasorum and cause inflammation; thus, TA may be defined as a vasa vasoritis.¹¹ This leads to destruction of the elastic fibers of smooth muscle cells of the media and is associated with atrophy, disappearance, and replacement with fibrosis. Calcification in those areas and thickening of the intima are also commonly seen.

Rapid development of arteritis can also lead to a dilated artery and formation of an aneurysm and aortic valve regurgitation.^{4,12} A two-stage process has been described in which a pre-pulseless phase, characterized by a lack of nonspecific inflammation, is followed

by a chronic phase with vascular insufficiency.³ Our patient had an intima-media thickening of the left CCA, a characteristic feature of TA. This atypical ultrasonographic finding was previously referred to as the “macaroni sign.”¹³

TA also leads to arterial media destruction, which combined with an inadequate fibrotic response results in dilation and aneurysm formation.¹⁴ A focal aneurysmal dilation of the left subclavian artery was identified along with two aortic aneurysms: one at the distal thoracic aorta and another near the renal arteries. Calcification, commonly found in TA, was identified in the abdominal aorta of our patient.

In summary, our case illustrates the typical ultrasonographic findings in the CCA. Awareness of such a finding may be of particular importance to many echocardiographers who are extending their practice into vascular imaging. In addition to clinical findings, diagnosis of TA may be supported by characteristic CT findings as illustrated by our case.

References

1. Takayasu M. A case with peculiar changes in retinal central vessels. *Acta Soc Ophthalm Jpn* 1908;12:554–555.
2. Numano F. The story of Takayasu arteritis. *Rheumatology (Oxford)* 2002;41:103–106.
3. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: A review. *J Clin Pathol* 2002;55:481–486.
4. Brady J, Esrig BC, Hamirani K, Baisre A, Saric M. Severe chronic aortic insufficiency requiring valve replacement: an infrequent complication of Takayasu’s disease. *Echocardiography* 2006;23:495–498.
5. Liang P, Hoffman GS. Advances in the medical and surgical treatment of Takayasu arteritis. *Curr Opin Rheumatol* 2005;17:16–24.
6. Moriwaki R, Noda M, Yajima M, Sharma BK, Numano F. Clinical manifestations of Takayasu arteritis in India and Japan—New classification of angiographic findings. *Angiology* 1997;48:369–379.
7. Vanoli M, Bacchiani G, Origg L, Scorza R. Takayasu’s arteritis: A changing disease. *J Nephrol* 2001;14:497–505.
8. Hall S, Barr W, Lie JT, Stanson AW, Kazmier FJ, Hunder GG. Takayasu arteritis. A study of 32 North American patients. *Medicine (Baltimore)* 1985;64:89–99.
9. Toshihiko N. Current status of large and small vessel vasculitis in Japan. *Int J Cardiol* 1996;54(Suppl):S91–S98.
10. Lupi-Herrera E, Sanchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE. Takayasu’s arteritis. Clinical study of 107 cases. *Am Heart J* 1977;93:94–103.
11. Kobayashi Y, Numano F. 3. Takayasu arteritis. *Intern Med* 2002;41:44–46.
12. Nasu T. Pathology of pulseless disease. A systematic study and critical review of twenty-one autopsy cases reported in Japan. *Angiology* 1963;14:225–242.
13. Maeda H, Handa N, Matsumoto M, et al. Carotid lesions detected by B-mode ultrasonography in Takayasu’s arteritis: “Macaroni sign” as an indicator of the disease. *Ultrasound Med Biol* 1991;17:695–701.
14. Andrews J, Mason JC. Takayasu’s arteritis—Recent advances in imaging offer promise. *Rheumatology (Oxford)* 2007;46:6–15.