Left main stem disease in a patient with Takayasu's arteritis

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Takayasu's arteritis is a chronic vasculitis of unknown aetiology involving the aorta and its main branches, the pulmonary and coronary tree. Women are affected more often than men (80 to 90% of the cases) with an age onset between 10 and 40 years. This case report demonstrates the limitations of exercise testing and stress echocardiography in diagnosing the extent of coronary artery disease in patients with inflammatory disease in the left main stem coronary artery. (*Neth Heart J* 2007;15:260-2.)

Keywords: Left main stem disease, Takayasu's arteritis, coronary artery bypass surgery

A 19-year-old female presented to the emergency room with a two-month history of exercise-induced chest pain, which was getting worse in intensity and frequency. Regarding her past medical history, she had documented Takayasu's arteritis, hypertension, nephrosclerosis of the left kidney and a right renal artery stenosis. Her physical examination was unremarkable. Laboratory tests, chest X-ray and the electrocardiogram were normal. Before her visit to the emergency room, a treadmill exercise test and a dobutamine stress echocardiogram had been performed in the outpatient clinic because of chest pain. Both investigations showed no evidence of myocardial ischaemia. The echocardiogram revealed a mild aortic regurgitation.

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Because she was known to have Takayasu's arteritis and her symptoms were typical for angina pectoris, coronary artery disease was suspected and coronary angiography was performed, despite normal stress echocardiography and treadmill exercise test. The angiogram revealed a pinpoint lesion of the left main stem, unresponsive to intracoronary nitroglycerin. The left anterior descending, circumflex and the right coronary arteries showed no abnormalities (figures 1A and B).

Subsequently the patient underwent coronary artery bypass surgery using venous conduit materials.

Discussion

Takayasu's arteritis is a chronic vasculitis of unknown aetiology, mainly affecting the aorta and its primary branches, the pulmonary and coronary tree. Takayasu's arteritis is also known by other terms such as pulseless disease, aortic arch syndrome, occlusive thromboangiopathy, Martorel syndrome and non-specific arteritis. Takayasu's arteritis is one of two types of large-vessel vasculitis, the other one being giant cell arteritis. It should be distinguished from medium- and small-vessel vasculitic syndromes such as polyarteritis nodosa, Kawasaki disease, Churg-Strauss arteritis, Wegener's granulomatosis and Henoch-Schönlein purpura. Takayasu's arteritis was first described in 1908 by Dr Mikito Takayasu in a Japanese patient with arteriovenous anastomosis around the papillae of the ocular fundi.¹ It is a systemic chronic inflammatory and obliterative vasculitic disease that usually involves the large and medium-sized arteries with preferential involvement of the aorta and its main branches. The pulmonary arteries are affected in 50% of patients. In a small number of patients the coronary tree is involved.² In 10 to 20% of cases aortic regurgitation may be present due to aortic root dilatation. Our patient had a mild aortic regurgitation, without aortic root dilatation. Renal artery stenosis and hypertension may occur in 30% of cases.³ This was also the case in our patient. The age of onset is between 10 and 40 years and the disease mainly affects females: 80 to 90% of the cases.⁴ The estimated worldwide incidence is 2.6 cases per million per year. Although it has a world-

A. A 90 to 99% pinpoint lesion of the left main stem (LMCA=left main coronary artery). Intracoronary nitroglycerin was used to exclude coronary vasospasm. The remaining coronary arteries were normal. **B.** Right coronary artery (RCA) was normal.

wide distribution the greatest prevalence is in Asia with 150 new cases per year in Japan.⁵ In Europe and the United States the incidence is one to three new cases per million per year.^{6,7} The pathogenesis of Takayasu's arteritis remains largely unclear. Genetic aetiology, autoimmune mechanisms, active inflammation, and antiendothelial antibodies may all play a role in this disease. Aneurysmal dilatation of the affected vessel is caused by destruction of the elastic lamina and the muscular media.⁴ Intimal proliferation and active inflammation can lead to the development of stenotic arterial lesions. The clinical symptoms can vary considerably. Systemic symptoms such as fatigue, low-grade fever and weight loss can be the first manifestation of Takayasu's arteritis. Other symptoms include vascular symptoms (subclavian steal syndrome, intermittent claudication, renal failure due to renal artery stenosis, diminished or absent peripheral pulsations), neurological symptoms, arthralgias and/or skin lesions. According to the American College of Rheumatology a diagnosis of Takayasu's arteritis can be established when at least three of the six criteria in table 1 are met.⁸

Treatment of Takayasu's arteritis consists of glucocorticoids with successful suppression of the systemic

symptoms, and slowing or even arrest of the progression of the vasculitis. At an advanced stage of the disease, when irreversible arterial stenosis has occurred, angioplasty or bypass surgery may be necessary. Overall total cardiac involvement (myocardial ischaemia or even myocardial infarction, coronary ectasia, valve pathology) in patients with Takayasu's disease is around 40%. As said before, coronary artery involvement is rare, about 5 to 15%.² When the coronaries do become affected it is usually the proximal part of the coronary artery that becomes stenotic due to direct extension of inflammation of the aortic wall. In the study by Endo et al.⁹ more than 85% of the patients with coronary involvement had ostial lesions. Coronary stenotic lesions in Takayasu's arteritis can be treated with percutaneous coronary intervention, but the restenosis rate is over 20 to 30%, especially when bare metal stents are used.^{11,12} So far there is little experience with drug-eluting stents. Coronary bypass surgery after pretreatment with glucocorticoids to suppress active inflammation has a good prognosis with a survival rate of 87% at five years and 81% at ten years.⁹ Because of persistence of arterial inflammation in up to 40% of cases, even after glucocorticoid treatment, with subsequent endangerment of insertion sites, venous rather than arterial conduit is used in coronary bypass surgery.

Table 1. The American College of Rheumatology 1990 criteria for the classification of Takayasu's arteritis.

Patient <40 years of age at disease onset
 Claudication of extremities
 Decreased brachial pulse
 Blood pressure difference between right and left arms >10 mmHg
 Bruit over subclavian artery, aorta or both
 Arteriographic abnormal appearance

At least three criteria must be present for the diagnosis of Takayasu's arteritis.

In our patient we decided to perform coronary artery bypass surgery rather than percutaneous coronary intervention.

In conclusion, although the incidence of coronary artery involvement in Takayasu's disease is low, in young, particularly female patients presenting with angina pectoris and especially when peripheral pulsations are diminished or absent, the diagnosis of Takayasu's arteritis with coronary artery involvement should be considered. ■

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