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CASE REPORT

Takayasu arteritis revisited☆

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KEYWORDS
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Abstract Takayasu arteritis is a large vessel vasculitis with various clinical presentations depending on the territories affected. We report the case of a 47-year-old woman who was diagnosed with Takayasu arteritis following rapid progression of coronary disease. The condition evolved rapidly under corticosteroid therapy, with formation of new arterial stenoses within the carotid and splanchnic circulations. Disease remission was achieved with cyclophosphamide pulses and percutaneous angioplasty of the affected vessels was performed.

PALAVRAS-CHAVE
Arterite de Takayasu;
Vasculite de grandes vasos;
Vasculite;
Ciclofosfamida;
Doença coronária

Arterite de Takayasu: a propósito de um caso clínico

Resumo A arterite de Takayasu é uma vasculite de grandes vasos que cursa com apresentações clínicas diversas consoante os territórios afetados. Apresenta-se o caso clínico de uma mulher de 47 anos a quem foi feito o diagnóstico de arterite de Takayasu na sequência de doença coronária rapidamente progressiva. O quadro evoluiu rapidamente, sob corticoterapia, com formação de estenoses a nível da circulação carotídea e esplâncnica, tendo-se conseguido estabilização da doença com pulsos de ciclofosfamida e angioplastia percutânea das lesões.

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Introduction

Takayasu arteritis is a rare vasculitis that mainly affects females before the age of 50.[1] Clinical presentation varies according to the territories affected; the left subclavian artery is most often involved, followed by the aorta and the carotid, renal and vertebral arteries.[2] Coronary artery involvement is well documented, but revascularization of coronary lesions is a challenge if the underlying disease is not controlled. Since there are no markers specific to the disease, in most cases diagnosis is based on identification of vascular lesions in imaging studies of patients with suggestive symptoms. Age of onset under 40, limb claudication, differences in pulses and systolic blood pressure between the arms, murmur over the subclavian arteries and stenosis of the great vessels on angiography are the diagnostic criteria used to distinguish Takayasu arteritis from the other vasculitides.[1]

Case report

We report the case of a 47-year-old woman, white, with a history of hypertension and chronic iron deficiency anemia but no other known medical history. Three weeks before admission she began to suffer chest pain radiating to the jaw, relieved by rest. Following assessment at another institution she was prescribed aspirin, transdermal nitroglycerin and nebivolol and was referred for urgent cardiological consultation. Worsening symptoms in the previous 48 hours prompted her to go to the emergency department of our hospital, where on admission she was eupneic and hypertensive, with pain triggered by minimal exertion and a II/VI apical systolic murmur; there were no other alterations on physical examination. Laboratory tests showed normocytic normochromic anemia with hemoglobin 10.4 g/dl and elevated troponin (rising from 0.21 to 0.32 ng/dl). Serial ECGs revealed dynamic repolarization changes without ST-T segment elevation, while echocardiography showed mild concentric ventricular hypertrophy but no wall motion abnormalities, and ejection fraction was normal.

She was provisionally diagnosed with non-ST-elevation acute coronary syndrome and underwent urgent coronary angiography, which revealed stenosis in the ostium of the left main but no other lesions. During double bypass surgery (left internal mammary-anterior descending and saphenous vein-obtuse marginal), findings suggestive of aortitis were observed, and so a biopsy was performed, and anatomopathological study showed a nonspecific lymphoplasmacytic inflammatory infiltrate. Syphilis serology was negative. Postoperative recovery was uneventful and no further investigation was carried out.

Around four months later, there was recurrence of exertional angina. At this time a difference in blood pressure between the arms was noted (20 mmHg less in the left arm), with a very weak pulse on the left and evident skin pallor immediately after exertion or when raising the left arm. Laboratory testing showed slight thrombocytosis and a rise in erythrocyte sedimentation rate to 100 mm/h. Echocardiography revealed new-onset hypokinesia of the left ventricular inferior wall, and coronary angiography was repeated, which showed preocclusive lesions of the right coronary artery and ostial saphenous graft. She underwent percutaneous coronary intervention with placement of two drug-eluting stents; during the procedure occlusion of the left subclavian artery was observed with retrograde filling from the ipsilateral vertebral artery (Figure 1), which raised the suspicion of Takayasu arteritis. Cranial computed tomography (CT) angiography confirmed proximal occlusion of the left subclavian artery upstream of the origin of the left vertebral artery (Figure 2), an atheromatous plaque in the distal segment of the right common carotid causing preocclusive stenosis of the external carotid, and tortuosity and kinking of the cervical course of the internal right carotid artery. Carotid Doppler ultrasound showed coiling of the right internal carotid and 70% stenosis, preocclusive stenosis of the

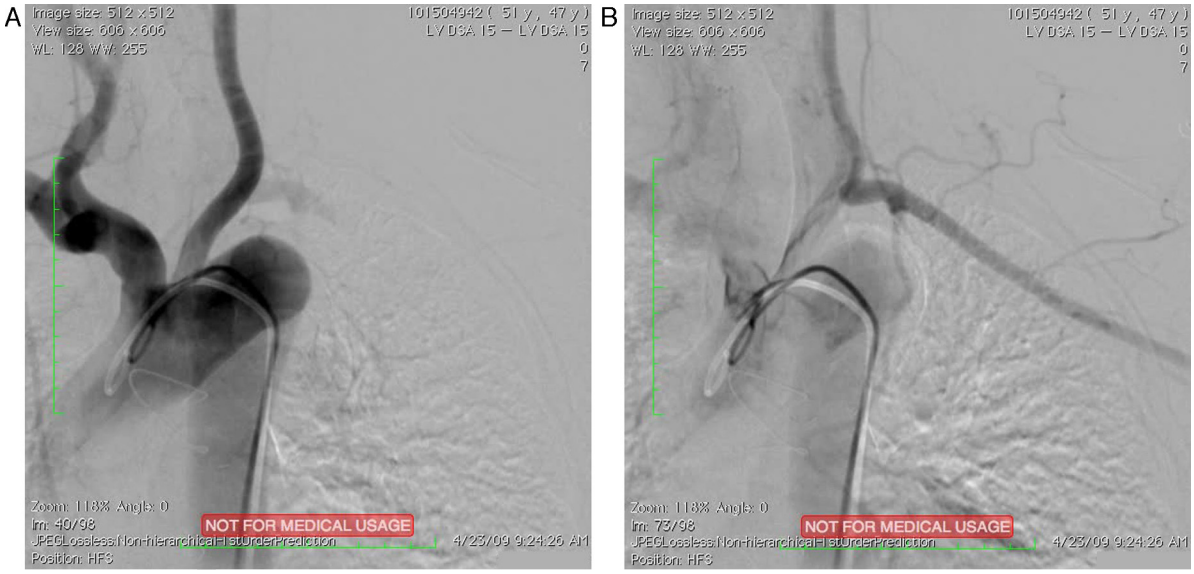

Figure 1 Aortography showing stenosis of the left subclavian artery (A) and retrograde filling by the ipsilateral vertebral artery (B).