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CASE REPORT

Atypical presentation of Takayasu arteritis with chest pain

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Abstract

Takayasu arteritis (TAK) is a progressive granulomatous necrotizing vasculitis that affects large vessels. Angina, acute myocardial infarction or stroke as primary manifestations of TAK are rarely reported in the medical literature. A 48-year-old man with the metabolic syndrome complained especially of exertional chest pain (angina-like) for the past 6 months. Apart from asymmetric pulse in the upper limbs, the physical examination was normal. Inflammation tests were positive, and ultrasound of the carotid arteries found a significant (3 mm) uniform thickening of the intima of carotid arteries, without atheroma plaques. Heart ultrasound and conventional chest radiography were normal. Angio-computed tomography of total aorta revealed a typical TAK aspect with circumferential parietal thickening of the ascending aorta, aortic arch and proximal descending thoracic aorta, corresponding to TAK type IIB. Treatment included intravenous methylprednisolone, followed by oral methylprednisolone and subcutaneous methotrexate, with positive outcome. The particularity of the case is the atypical presentation of TAK with angina-like chest pain which was initially labelled as cardiac ischemic disease in a patient with the metabolic syndrome. Specifically, chronic chest pain in TAK can be caused by aortic involvement (aortitis), coronary artery involvement (stenosis), pulmonary artery involvement (pulmonary hypertension) or aortic aneurysm and dissection (which cause sudden, severe chest pain). The case also underlines the importance of multidisciplinary collaboration in vasculitis and the rising role of vascular ultrasound.

Keywords: Takayasu arteritis, vasculitis, chest pain.

Introduction

Takayasu arteritis (TAK) is a progressive granulomatous necrotizing vasculitis that affects large vessels and leads to various manifestations depending on the type of vessel involved. TAK is diagnosed by clinical history of claudication, absent pulse, discordant blood pressure, aortic bruit and typical angiographic findings. Angina, acute myocardial infarction or stroke as primary manifestations of TAK are rarely reported in the medical literature. Coronary involvement occurs in 10-15% of patients, stenosis of the right coronary being the most common lesion. Isolated involvement of only the coronary arteries is extremely rare [1, 2].

Case presentation

A 48-year-old man, who was a former high-performance athlete, was transferred to the rheumatology clinic after being investigated and treated in the cardiology and vascular departments for previous 8 years. He complained of asthenia, fatigue, weight loss (10 kg in one year), which worsened in the prior 6 months, but his main complaint was exertional chest pain (angina-like) with no irradiation. Patient's comorbidities were related to the metabolic syndrome, namely obesity, hyperuricemia, hypercholesterolemia, arterial hypertension and chronic ischemic heart disease.

His medical history began in May 2016 with unstable angina symptoms. After coronary angiography was performed, he required interventional therapy with angioplasty and placement of 4 stents on the right and anterior interventricular artery. In December 2016, as the symptoms reappeared, the restenosis of the same arteries was discovered and the patient was treated with myocardial revascularization by triple bypass. Between 2017 and 2024 the patient was periodically evaluated by the cardiology department. Angina symptoms improved, but in the last year he periodically accused fatigue and dilatation of the jugulars. Doppler ultrasound of the cervical arteries performed in the cardiology department noted a "uniform thickening of the carotid wall", which subsequently lead to the patient being referred to the rheumatology department.

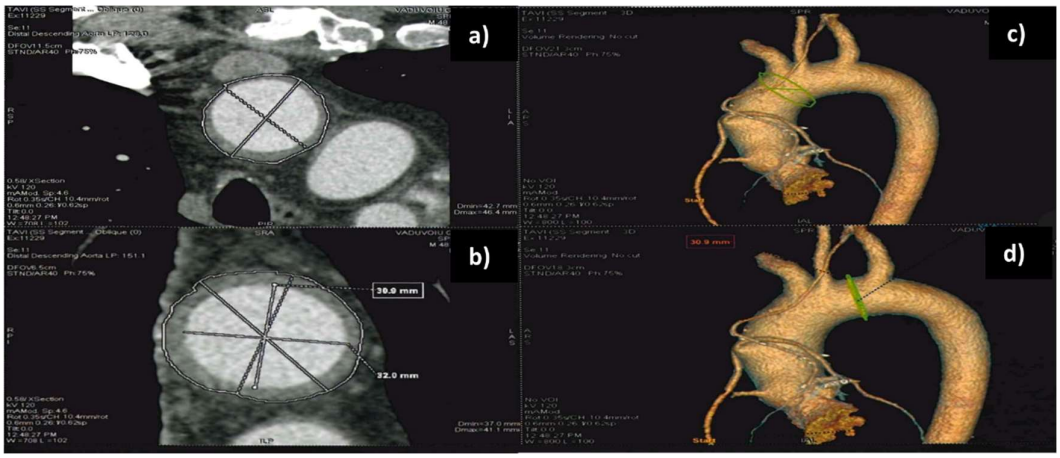

Figure 1. *Aortic angio-CT. Aorta, ascending portion (a) and transverse segment (b) with 3D reconstruction (c, d).*

His first rheumatology consult revealed a good general condition, no fever, no skin manifestations and no oedema. Clinical examination of the osteo-articular system was quasi-normal. Systolic blood pressure was 140 mmHg in the left arm and 130 mmHg in right arm and he had asymmetric pulse in the upper limbs, with no heart/arterial murmurs. The rest of the physical examination was normal.

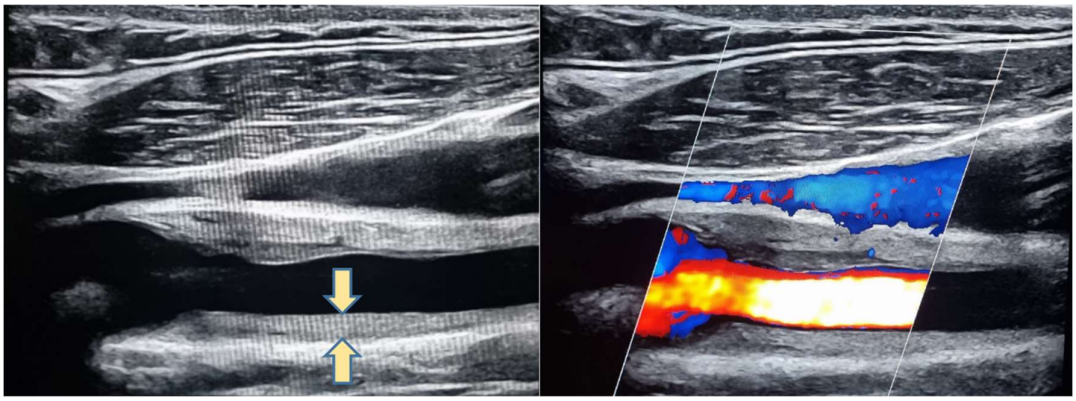

Figure 2. *Ultrasound of the right carotid artery. Wall thickening illustrated by arrows.*

Laboratory workup was performed: complete blood count was normal and there were no positive antibodies, including antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA). Inflammation tests were positive, with a C-reactive protein of 31.4 mg/L (normal < 5 mg/L) and ESR of 34 mm/h (normal < 20 mm/h). Imaging findings were vital for the diagnosis. Ultrasound of the carotid, axillar and temporal arteries found a significant (3 mm) uniform thickening of the intima of carotid arteries, without atheroma plaques. The echo-cardiography was normal. Conventional chest radiography was also normal. Angio-computed tomography of total aorta was performed, revealing a typical TAK aspect with circumferential parietal thickening of the ascending aorta, aortic arch and proximal descending thoracic aorta, corresponding to TAK type IIB [3]. Therefore, the positive diagnosis was of TAK type IIB with coronary involvement.

Treatment included a 3-day pulse-therapy (500 mg per day) of intravenous methylprednisolone, continued with oral methylprednisolone 64 mg/day (0,7 mg/kg/day) with gradual decrease until 16 mg/day after 3 months. A concomitant sparring agent was initiated according to the latest EULAR guidelines in large vessels vasculitis management [4], namely methotrexate with initial doses of 15 mg/week and then 20 mg/week, that was well tolerated by the patient. Substantial improvement was observed within the first week of treatment. The patient also received cardiological medication with aspirin 75 mg/day, statin and antihypertensive treatment. The EULAR recommendations (2018) indicate that antiplatelet or anticoagulant therapy should not be routinely prescribed for treating patients with TAK unless indicated for other reasons (the patient had multiple revascularisation interventions).

Discussions

TAK is a rare large vessel vasculitis, with reported worldwide incidence rates of only 1–2 per million with a predominance for female patients and Asian countries. It is considered a disease of the young, but it has also been described in adults and the elderly [1, 5]. Clinically TAK is characterized by varied

ischaemic symptoms, the most common being dizziness, neurological disturbances, syncope, absent pulses, differences in systolic blood pressure between arms [6, 7]. Chest pain (stable or unstable angina), myocardial infarction and even sudden death are cited in case reports as uncommon manifestations of TAK, especially at the beginning of the disease. In a patient with other cardiovascular comorbidities, it is hard to identify if the coronary lesions are fully caused by the pathogenesis of TAK or by atherosclerosis. Cardiovascular complications resulting from accelerated atherosclerosis and vascular calcification are a significant cause of morbidity and mortality in patients with large vessel vasculitis (LVV) [8, 9]. In patients with LVV, coronary artery pathology may develop not only due to inflammation, but also as a result of accelerated atherosclerosis and endothelial dysfunction [10].

Specifically, chronic chest pain in TAK can be caused by aortic involvement (aortitis), coronary artery involvement (stenosis), pulmonary artery involvement (pulmonary hypertension) or aortic aneurysm and dissection (which cause sudden, severe chest pain). Coronary artery lesions, due to the nonspecific clinical manifestations can lead to a delayed diagnosis, and the difficulty of treatment. The literature is scarce in cases with TAK presenting predominantly with chronic chest pain [11-14].

In the latest recommendations for the use of imaging in LVV, published in 2023, compared to the 2018 version, ultrasound (US) is now recommended as first-line imaging test of patients with suspected giant cell arteritis (GCA) [10]. In this case, US was used to discriminate between these two entities (GCA and TAK), since it is easily accessible, non-irradiating and non-invasive and can be used in monitoring the patient. In TAK, magnetic resonance imaging (MRI) is the preferred imaging modality, with PET-CT, CT or ultrasound mentioned as alternatives. Imagining modalities are not routinely recommended for follow-up, but ultrasound, PET-CT or MRI may be used for assessing vessel abnormalities in patients with suspected relapse [10] or persistent symptoms.

Treatment was also improved over time in LVV. In last EULAR guide for the treatment and management of TAK, second line medication in patients with TAK resistant to conventional therapy includes tumour necrosis factor (TNF) inhibitors and interleukin (IL)-6 inhibitors (e.g. tocilizumab – TCZ [15]) with a good prognosis in TAK. TCZ treatment led to an improvement in imaging findings of TAK patients with coronary artery involvement and can be used as a perioperative treatment strategy in revascularization [16]. There are also more biologic agents that are being studied, but with no recommendation to date, such as JAK-inhibitors and rituximab, with good results in some case reports [17, 18].

Conclusions

The main particularity of the case is the atypical presentation of TAK with angina-like chest pain which was initially labelled as cardiac ischemic disease in a patient with the metabolic syndrome. The case underlines the importance of multidisciplinary collaboration in LVV and the rising role of vascular ultrasound.

Conflicts of Interest: The authors declare no conflicts of interest.

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