CASE REPORT

Thrombosed aneurysm as the initial manifestation of Takayasu arteritis

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Accepted 25 May 2014

SUMMARY

Takayasu arteritis (TA) is a large-vessel vasculitis characterised by stenosis, dilation and/or aneurysm formation. We present a case of a 43-year-old man with an initial manifestation of an acute thrombosed aneurysm. He was found to have a thrombosed right common femoral artery aneurysm on Doppler ultrasound. Physical examinations revealed a substantial difference in blood pressure level between bilateral upper extremities, and absent pulses at right upper and lower extremities. The diagnosis was confirmed by angiography, which revealed 100% occlusion of the right common femoral artery. Biopsy of the thrombosed aneurysm was consistent with a diffuse lymphocytic subendothelial infiltrate. The patient was treated with high-dose corticosteroids and had a good response to treatment, as demonstrated by a decrease in sedimentation rate, and recovery of the right side pulses. This case helps to create awareness among physicians that TA may present with an acute occlusion of an aneurysm without the typical warning symptoms of TA.

pressure (BP) between bilateral upper extremities (right arm BP, 111/78 mm Hg and left arm BP, 142/95 mm Hg) and no palpable pulses in the right brachial, radial, femoral, popliteal, dorsalis pedis or posterior tibialis arteries compared with +2 pulses in his left upper and lower extremities. The patient's initial laboratory workup was remarkable for a persistent increase in white cell count ($>16\,000$ cells/mm³), microcytic anaemia and polyclonal gammopathy. The erythrocyte sedimentation rate was 72 mm/h. Screening tests for tuberculosis and syphilis were negative. Laboratory workup for connective tissue diseases, including: antinuclear antibody, rheumatoid factor, proteinase-3 antibodies and myeloperoxidase antibodies, was negative. He had a positive partial thromboplastin time-lupus anticoagulant screen and a hexagonal phase consistent with lupus anticoagulant. Anticardiolipin antibodies, β -2 glycoprotein I antibodies, prothrombin 20 210 mutation and factor V Leiden mutation were all negative. The patient had normal protein S and C activity levels.

BACKGROUND

Takayasu arteritis (TA) is a large-vessel vasculitis characterised by stenosis, dilation and/or aneurysm formation.¹ The initial symptoms are usually fever, weight loss, night sweats and general malaise, and disease onset generally occurs before the age of 40, with a marked female predominance.² Although aneurysms may occur as part of the disease process, they tend to occur later in the course of the disease.³ We present the case of a 43-year-old man diagnosed with TA who initially presented with a thrombosed common femoral artery aneurysm.

CASE PRESENTATION

A 43-year-old, previously healthy Hispanic man, presented with a sudden-onset right hip pain that irradiated to his right leg. Symptoms gradually worsened over 2 days, culminating with a cold and numb right lower extremity from the knee level. The patient had a Doppler ultrasound, which found a thrombosis of an aneurysm of the right common femoral artery. The patient reported no previous episodes of fever, general malaise, arm claudication, headaches, visual loss, abdominal pain, renal disease, chest pain, shortness of breath or other symptoms of a typical case of TA.

Physical examination in the emergency department revealed severe pain in the right inguinal area, without a pulsatile mass or discolouration of the skin of the right leg. Measurement of his vital signs revealed a substantial difference in blood

INVESTIGATIONS

Transthoracic echocardiography revealed a normal ejection fraction, no pericardial effusion or valvulopathy. Moreover, renal ultrasound findings were consistent with left renal atrophy and decreased flow to the left renal artery (60%). Our patient underwent angiography, which revealed 100% occlusion of the right common femoral and superficial femoral arteries, as well as dilation of the left common femoral and superficial femoral arteries (figure 1). He had a graft placed in the occluded arteries and achieved successful reperfusion. Tissue obtained from the affected segments of the common and superficial femoral arteries had a diffuse lymphocytic subendothelial infiltrate by H&E stain (figure 2). An additional angiography identified mild dilation at the abdominal aorta (figure 3), coronary artery aneurysm and occlusion of left renal artery.

TREATMENT

The patient received 80 mg of prednisone daily for 1 week then decreased to 60 mg; it was continued on re-evaluation at the rheumatology clinic to continue taper down slowly.

OUTCOME AND FOLLOW-UP

Once treatment began his sedimentation rate decreased to 24 mm/h, and he recovered his right side pulses. The patient remained stable without

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To cite: Ojeda J, Rodríguez Y, Ríos G. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-203523

Figure 1 Right lower extremity angiography showing complete occlusion of the right common femoral artery.

systemic symptoms and had an unremarkable physical examination at his follow-up visit 3 weeks after discharge.

DISCUSSION

Our patient had acute onset of right hip pain as the presenting symptom of TA. TA typically affects young adults and mostly females; only 20% of the cases present after age 40.²⁻⁴ We report the case of a 43-year-old man who presented at the time of disease onset with an initial manifestation of an acute thrombosed aneurysm. The most common initial presentation of TA is an insidious onset of fever, arthralgia, night sweats, weight loss and general malaise.² In a study of 60 patients, the most common presenting vascular symptoms were claudication, absent pulses, carotid bruits, hypertension, carotidynia, light-headedness and asymmetric BP readings in the upper

Figure 2 Medium-power, H&E stain of the right common femoral artery surgical specimen revealed subendothelial inflammation with diffuse lymphocyte infiltration.

Figure 3 Abdominal aorta angiography presenting with mild dilation.

extremities.^{4, 5} Although our patient had objective evidence of asymmetric BP and pulses, he was asymptomatic prior to his initial presentation of right hip pain.

The main feature of TA is vascular involvement.⁴ In a study of patients with TA, 68% had stenotic lesions, which were 3.6 times more common than aneurysms, and 98% had long aortic stenotic lesions.⁴ Aneurysm tends to be diagnosed later in the disease course. A study conducted in Mexico reported 15% of patients with TA died during the 19-year follow-up, with one death caused by a subclavian aneurysm rupture.⁶ In another study, two patients with TA died during a 13-year follow-up, of which one death was caused by a ruptured aortic aneurysm.⁷ Although there are reported cases of acute rupture of an aneurysm in patients with TA, most of these were subclavian, carotid, thoracic or abdominal aortic aneurysms.^{6, 8-11} Interestingly, our patient had aneurysms of the coronary arteries and the bilateral femoral artery, the latter of which is an uncommon manifestation in TA.^{3, 12}

Our case, as well as other previously published cases, helps to create awareness among physicians that TA may present with acute occlusion of an aneurysm without the typical warning symptoms of TA. Increased physician awareness of atypical presenting symptoms combined with the use of non-invasive imaging modalities may help improve diagnosis of TA in atypical cases, such as our case.

Learning points

- ▶ Takayasu arteritis may present without constitutional symptoms such as fever, weight loss, fatigue and general malaise.
- ▶ Takayasu arteritis should be considered in the differential diagnosis of a patient presenting with occlusion of the right common femoral artery.
- ▶ Thrombosed aneurysm may occur as the initial manifestation in the course of disease.

Acknowledgements All the authors would like to thank Dr Carmen Gurrea Rosas for her collaboration with the biopsy report.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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