Takayasu's arteritis in an active man: burnt out or quiescent?

Introduction

Takayasu's arteritis is a chronic inflammatory disease of large and medium sized arteries, particularly affecting the aorta and its main branches. Histologically it is characterized by a panarteritis. The inflammation results in stenotic, occlusive or aneurysmal changes in affected arteries, giving rise to symptoms of ischaemia, infarction or secondary hypertension. Takayasu's arteritis is an unusual cause of ischaemic heart disease. Patients presenting early in life with ischaemic sounding chest pain, and/or symptoms of ischaemia elsewhere, should arouse suspicion of this diagnosis. As in this case, the clue is frequently an absence of or discrepancy in palpable limb or neck pulses.

Discussion

This article describes a very active young man with occlusive coronary and aortic disease, no recognized risk factors and no obvious inflammatory episode. The differential diagnosis could be premature accelerated atherosclerosis or 'burnt-out' Takayasu's arteritis.

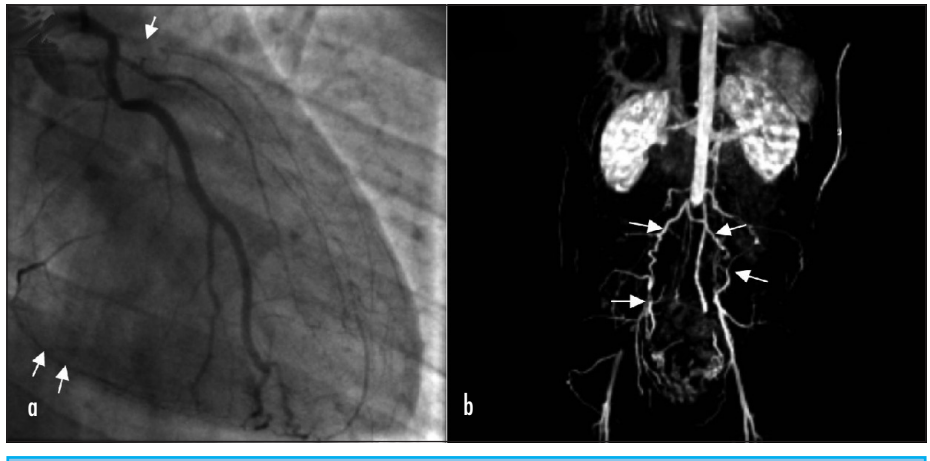
Takayasu's arteritis is a chronic inflammatory condition that affects the medium and large arteries with a predilection for the aorta and its major branches (Perera et al, 2013). Within the UK, the estimated mean prevalence is 4.7/million (Watts et al, 2009). Studies report a predominance of women (9 female:1 male) with onset of disease usually between the ages of 20 and 30 years (Brunner et al, 2010).

Dr Simon W Dubrey *is Consultant Cardiologist,* **Dr Richard Grocott-Mason** *is Consultant Cardiologist and* **Dr Sarah Ghonim** *is Cardiology Specialist Registrar in the Department of Cardiology, Hillingdon Hospital, Uxbridge, Middlesex UB8 3NN and* **Dr Tarun Mittal** *is Consultant Radiologist in the Department of Radiology, Royal Brompton and Harefield Hospitals, Harefield Hospital, Harefield, Middlesex*

Correspondence to: Dr SW Dubrey (simon.dubrey@thh.nhs.uk)

Although no specific aetiology has been identified, there is an abundance of evidence to support an autoimmune process. Inflammatory markers (C-reactive protein and erythrocyte sedimentation rate) are of some use but there are no auto-antibodies or specific biomarkers that identify active Takayasu's arteritis. Angina is present in 6–16% of cases (Lupi-Herrera et al, 1977). Imaging modalities capable of detecting pre-stenotic disease include computed tomographic angiography, high-resolution ultrasound and magnetic resonance angiography, with specific focus on changes in thickness of the aortic wall (Steeds and Mohiaddin, 2006). 18-fluorodeoxyglucose positron emission tomography has proved useful in diagnosing and monitoring disease activity in Takayasu's arteritis (Andrews et al, 2004).

Figure 1. a. Coronary angiography demonstrating obstruction of the left anterior descending (single arrow) and right coronary artery, evidenced by extensive cross filling (two arrows). b. Magnetic resonance angiography showing total occlusion of the distal aorta with extensive collaterals (arrows) to both lower limbs.



Case Report

A 22-year-old man presented with atypical chest pain. He was fully active. On examination, all pulses were absent in the lower limbs. Inflammatory markers fluctuated with his serum C-reactive protein level ranging from <1 mg/litre to as high as 80 mg/litre. Troponin-I was elevated at 7 ug/litre (normal range <0.04). The serum glucose, glucose tolerance test, calcium and lipid profile were normal. An electrocardiogram showed sinus rhythm with inferior lead saddle-shaped ST segments, suggestive of myopericarditis. There was no family history of premature atherosclerotic disease.

Echocardiography revealed impaired left ventricular function (estimated 40–50%) with regional wall motion abnormalities. Cardiac magnetic resonance imaging with delayed gadolinium imaging demonstrated subendocardial enhancement, indicative of infarction.

Computed tomography of the coronary arteries revealed extensive atheroma in the proximal left anterior descending and right coronary artery. Invasive angiography confirmed proximal occlusion of both these vessels (*Figure 1a*). Magnetic resonance angiography demonstrated occlusion of the distal aorta and both common iliacs with extensive collateralisation (*Figure 1b*). 18-fluorodeoxyglucose positron emission tomography showed no evidence of active vasculitis.

Coronary artery bypass grafting, using both internal mammary arteries, was successfully performed. Histology showed no evidence of active vasculitis. The patient is now taking aspirin, simvastatin and ramipril, is fully active and has not required immunosuppressive therapy.

The mainstay of treatment of Takayasu's arteritis is immunosuppression. This is initially instituted with corticosteroids, which alone will induce remission in about 40–60% of patients. Additional agents may include methotrexate, azathioprine cyclophosphamide, mycophenolate mofetil, anti-tumour necrosis factor therapies (infliximab or etanercept) and the anti-interleukin-6 receptor monoclonal antibody tocilizumab, to control disease activity or facilitate reduction in corticosteroid dose. Short stenotic vascular lesions may be treated by percutaneous transluminal angioplasty. Surgical options include coronary artery bypass surgery and endarterectomy. Restenosis is a major complication when considering surgical intervention, with control of disease activity a priority before and following revascularization (Perera et al, 2013).

Sometimes described as 'burnt-out' it is important to realize that Takayasu's arteritis may be quiescent and can re-activate (Mañá et al, 2003). Continued clinical surveillance is advised. BJHM

LEARNING POINTS

- Takayasu's arteritis should be considered in young patients presenting with ischaemic symptomatology and/or absent pulses.
- A diagnosis is reached using clinical criteria, advanced imaging modalities and available histology.
- The disease may rarely remit spontaneously and present in a quiescent phase, but importantly cannot be relied on to remain 'dormant'.

Andrews J, Al-Nahhas A, Pennell DJ et al (2004) Non-invasive imaging in the diagnosis and management of Takayasu's Arteritis. *Ann Rheum Dis* **63**: 995–1000 (doi: 10.1136/ard.2003.015701)

Brunner J, Feldman BM, Tyrell PN et al (2010) Takayasu arteritis in children and adolescence. *Rheumatology* **49**: 1806–14 (doi: 10.1093/rheumatology/keq167)

Lupi-Herrera E, Sanchez-Torres G, Marcushamer J et al (1977) Takayasu's arteritis: clinical study of 107 cases. *Am Heart J* **39**: 744–50 (doi: 10.1016/S0002-8703(77)80178-6)

Mañá J, Montero A, Vidal M, Marcoval J, Pujol R (2003) Recurrent sarcoidosis: a study of 17 patients with 24 episodes of recurrence. *Sarcoidosis Vasc Diffuse Lung Dis* **20**(3): 212–21

Perera AH, Mason JC, Wolfe JH (2013) Takayasu arteritis: criteria for surgical intervention should not be ignored. *Int J Vasc Med* 618910 (doi: 10.1155/2013/618910)

Steeds RP, Mohiaddin RH (2006) Takayasu arteritis: role of cardiovascular magnetic resonance. *Int J Cardiol* **109**: 1–6 (doi: 10.1016/j.ijcard.2004.09.023)

Watts R, Al-Taiar A, Mooney J, Scott D, MacGregor A (2009) The epidemiology of Takayasu arteritis in the UK. *Rheumatology* **48**(8): 1008–11 (doi: 10.1093/rheumatology/kep153)

Varicella pneumonia

A 21-year-old male smoker presented with a 10-day history of headache, myalgia, arthralgia, malaise, sore throat and loss of appetite after exposure to a relative with chickenpox.

Varicella pneumonia is estimated to occur in one in 400 cases of chickenpox infection and is increasing in incidence in the UK (Mohsen and McKendrick, 2003).

Varicella pneumonia presents 1–6 days after onset of the rash and is associated with tachypnoea, cough, dyspnoea, fever and occasionally pleuritic chest pain and haemoptysis. Chest symptoms may start before the skin rash appears. Physical findings are often minimal and chest radiographs (*Figures 1* and *2*) typically reveal nodular or interstitial pneumonitis (Mohsen et al, 2001).

Smokers, the immunocompromised and patients who have chronic lung disease are at increased risk of developing pneumonia (Popara et al, 2002).

Varicella pneumonia can progress rapidly to fulminant respiratory failure despite maximum conventional support. Treatment with aciclovir has been successful. Varicella vaccine is effective in preventing or modifying the severity of varicella infection if used <5 days after exposure (Hall et al, 2000). BJHM

Dr Tamer Shalaby *is ST5 in Acute Medicine in the Department of Acute Medicine and* **Dr Ravish Mankragod** *is Consultant in Respiratory Medicine, Maidstone Hospital, Maidstone, Kent*

Correspondence to: Dr T Shalaby, Gastroenterology Department, Royal Surrey County Hospital, Guildford, Surrey GU2 7XX (t.shalaby@nhs.net)

Figure 1. Chest X-ray showing small bilateral multiple nodules.

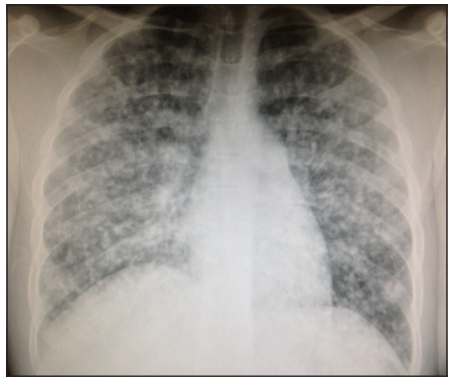
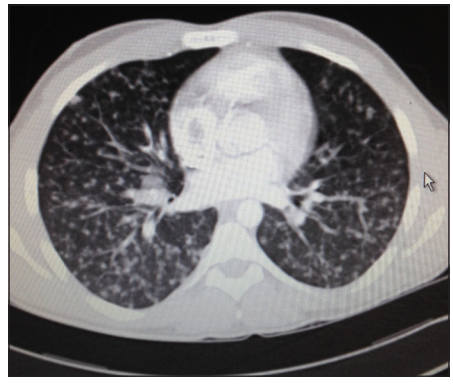


Figure 2. Computed tomography of the chest showing small bilateral multiple nodules.



Hall S, Galil K, Watson B, Seward J (2000) The use of school-based vaccination clinics to control varicella outbreaks in two schools. *Pediatrics* **105**: 111

Mohsen AH, Peck RJ, Mason Z, Mattock L, McKendrick MW (2001) Lung function tests and risk factors for pneumonia in adults with chickenpox. *Thorax* **56**: 796–9

Mohsen AH, McKendrick M (2003) Varicella pneumonia in adults. *Eur Respir J* **21**(5): 886–91

Popara M, Pendle S, Sacks L, Smego RA, Mer M (2002) Varicella pneumonia in patients with HIV/AIDS. *Int J Infect Dis* **6**: 6–8