A woman with progressive lethargy and sudden onset of shortness of breath

A 65-year-old Caucasian woman presented to the emergency department with rapidly worsening shortness of breath. On presentation she was tachycardic and tachypnoeic with reduced (85%) oxygen saturation. Cardiovascular examination revealed elevated jugular venous pressure with positive Kussmaul’s sign, pulsus paradoxus and muffled heart sounds. Her inflammatory markers were elevated; she had neutrophilia and deranged liver function tests. Imaging revealed cardiomegaly, a large fusiform thoracic aortic aneurysm, pericardial effusion and right ventricular free wall collapse during diastole (suggestive of tamponade). Urgent pericardiocentesis was performed with rapid symptomatic relief. She subsequently underwent aortic root and valve replacement surgery. Histology of the resected specimen showed inflammatory infiltrate with giant cell formation indicative of giant cell arteritis (GCA). This case highlights the need to consider GCA in the differential diagnosis of patients presenting with aortic aneurysm and pericardial effusion.

KEY WORDS: Giant cell arteritis, giant cell aortitis, aortic aneurysm, cardiac tamponade

Lesson

Case presentation

A 67-year-old Caucasian woman presented to the emergency department with a 3-day history of rapidly worsening dyspnoea on the background of a 3-month history of lethargy and exertional dyspnoea. Her medical history was significant for hypercholesterolaemia and diet-controlled type 2 diabetes mellitus. She was a current smoker. A myocardial perfusion scan performed 6 weeks before the current presentation had been unremarkable.

On initial assessment she was afebrile, with a pulse of 110 beats per min, respiratory rate 30 breaths per min and oxygen saturation 85% on room air. Cardiovascular examination revealed elevated JVP (jugular venous pressure) with positive Kussmaul’s sign (rise in JVP on inspiration). Her blood pressure was 100/70 mmHg on inspiration and 120/70 mmHg on expiration (pulsus paradoxus). Auscultation revealed muffled heart sounds. Respiratory examination was normal.

Investigations and treatment

A full blood count showed haemoglobin of 114 g/l, white cell count of 13.7 × 10⁹/l and normal platelet count. Differential white cell count revealed neutrophilia (neutrophil count 10.74 × 10⁹/l). Renal functions were normal but liver function tests were altered (normal bilirubin, elevated γ-glutamyl transpeptidase [171 U/l], alkaline phosphatase [269 U/l], alanine transaminase [109 U/l] and aspartate aminotransferase [84 U/l]). C-reactive protein was elevated at 140 mg/l. Autoimmune markers and syphilis screen were negative.

A chest radiograph (Fig 1) revealed cardiomegaly. Computed tomography displayed a large fusiform thoracic aortic aneurysm (Fig 2a) and evidence of pericardial effusion (Fig 2b). Echocardiography (Fig 3) confirmed pericardial effusion with right ventricular free wall collapse during diastole (suggestive of tamponade).

Pericardiocentesis yielded 770 ml haemorrhagic fluid and resulted in immediate symptomatic relief. The patient subsequently underwent aortic root and valve replacement surgery. Histology of the aortic tissue showed inflammatory infiltrate with giant cell formation suggestive of giant cell arteritis.

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giant cell formation (Fig 4). She was diagnosed as having giant cell arteritis (GCA) causing aortic aneurysm and cardiac tamponade.

Outcome and follow-up

The patient developed right middle cerebral artery infarct on postoperative day 7, manifested as left hemiparesis. She subsequently recovered well and was discharged home after intensive rehabilitation. Her inflammatory markers normalised and a PET (positron emission tomography) scan 12 months later did not show any features of arteritis.

Discussion

GCA is a systemic granulomatous vasculitic syndrome with an adverse impact on medium and large arteries and a predilection for older patients, particularly those aged >70 years. The male:female ratio is 1:3.8. The geographical spread of GCA favours European and American Caucasian individuals.

Aetiology of GCA remains unclear but the disease pathogenesis involves an interaction between genetic and environmental factors. The hallmark histopathological feature is transmural (intimal, medial and adventitial) inflammation, characterised by acellular islands of infarcted (devoid of smooth muscle cells) medial elastic tissue, typically cuffed by coalescing lymphocytes, giant cells and macrophages.

The main blood vessels affected are extracranial branches of the external carotid artery and the aorta and its major branches. Aortic involvement, being clinically silent, is usually undiagnosed and can manifest very late.

In the aorta, inflammation is more prominent in the adventitial and medial layers, causing minimal stenosis but increasing stiffness, weakness, dilatation and aneurysm formation. Thoracic aortic aneurysms are 17 times, and abdominal aortic aneurysms 2.5 times, more common in patients with GCA compared with the background population of the same age and sex. Being asymptomatic in the acute stages, the aneurysm is detected only by routine radiological tests or when an aortic dissection appears. Aortic dissection, complicating GCA, is rare but usually fatal. Patients with aortic involvement should be closely monitored and referred for surgical intervention if they develop significant aortic aneurysms.

Pericardial involvement in GCA appears to be immune mediated but could be complicated by concurrent infection. Most patients with pericardial complications are identified incidentally during other imaging studies, although some present with the classic signs and symptoms of pericarditis or a haemodynamically significant pericardial effusion. Rare complications include cardiac tamponade and constrictive pericarditis. These complications have been reported even when the patients have been on adequate immunosuppression.

Primary therapy of GCA is with high-dose corticosteroids (0.5–1 mg/kg per day) for a few weeks, depending on clinical response. Patients often need prolonged therapy with low-dose steroids.
GCA therefore needs to be considered in the differential diagnosis of patients presenting with aortic aneurysm and pericardial effusion.

**Conclusion**

Asymptomatic aortic involvement is common in GCA. Patients with GCA are at higher risk for catastrophic aortic complications. Aneurysms are relatively common but other complications such as cardiac tamponade are extremely rare. In patients presenting with aortic aneurysm or pericardial tamponade of unclear cause, a thorough histopathological examination of the resected specimen is essential to rule out GCA.

**References**


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