CASE REPORT

Giant cell arteritis presenting with arm claudication

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Abstract

Case report: a 72-year-old woman presented after a series of falls with a history of malaise, lethargy, cold hands and arm weakness for several months. She had undergone extensive investigation for what remained an unexplained anaemia during this period. Examination revealed cold hands with absent radial pulses. Axillary Doppler studies confirmed grossly diminished systolic pressures. The erythrocyte sedimentation rate was markedly elevated. An arch aortogram showed oblitative changes selectively affecting the subclavian and axillary arteries. In spite of clinically normal temporal arteries, and the absence of headache, temporal artery biopsy revealed giant cell arteritis.

Discussion: aortic arch syndrome is an important but under-appreciated complication of giant cell arteritis and may be the presenting feature. In this case, it resulted in falls consequent upon arm claudication when using a walking frame.

Keywords: arm claudication, giant cell arteritis

Introduction

Although most clinicians are familiar with the focal features associated with obliterative endarteritis of the cranial branch arteries that occur in giant cell arteritis (GCA), it is less well appreciated that this disorder also prominently affects the aorta and its proximal branches in a centripetal fashion.

A limited view of this disease as one presenting with symptoms of cranial artery involvement results in missed and delayed diagnoses as typical signs and symptoms are often absent when the patient first seeks medical attention. A spectrum of aortic involvement occurs, ranging from aortitis (which is asymptomatic or associated with systemic rather than local symptoms) to an aortic arch syndrome or aneurysm formation [1–9]. We report a case of GCA presenting as arm claudication.

Case report

A 72-year-old woman with a history of several months of malaise, lethargy, cold hands and arm weakness was admitted after several falls. She had previously had outpatient investigation of microcytic anaemia and raised erythrocyte sedimentation rate which remained unexplained: haematinies, myeloma screen, auto-antibodies, upper and lower gastrointestinal tract endoscopy and marrow biopsy were all normal or negative.

On examination, she had cold hands with impalpable radial and brachial pulses and was unable to abduct either arm beyond 90°. Other pulses were unremarkable. Systolic pressure at the ankle was 130 mmHg, but axillary artery Doppler studies revealed systolic pressures of only 30 mmHg on the left and 60 mmHg on the right.

Haemoglobin was normal after a transfusion 6 weeks previously, before which it had been 8.6 g/dl (12–15) with a mean corpuscular volume of 72 fl (80–100) Platelet count was 51x5103/l (150–400), erythrocyte sedimentation rate was 80 mm/h and C-reactive protein 107 mg/l (<7). Other results were normal, apart from a slightly elevated serum alkaline phosphatase and reduced serum albumin.

Chest radiograph and computed tomography were normal, with no evidence of cervical ribs or chronic aortic dissection. Aortography showed severe bilateral disease of the subclavian and axillary arteries, with smooth, diffusely narrowed segments suggesting vasculitis rather than atheroma (Figure 1). In spite of the absence of scalp tenderness or headache at any stage and clinically normal temporal arteries, temporal artery biopsy revealed GCA (Figure 2) and she was given prednisolone.
initial presentation with falls was explained on the basis of arm claudication when using a walking frame.

**Discussion**

GCA is readily recognized when cardinal features such as headache, jaw claudication, visual loss or polymyalgia rheumatica occur. However, it is under-recognized that up to 40% of patients present in a non-classical or occult manner with prominent systemic features or local symptoms resulting from large artery involvement [1, 2]. In one series, 16% of patients presented with pyrexia (GCA is now the single commonest cause of pyrexia of unknown origin, accounting for 15% of cases in the over-50s [3]) and a further 4% with unexplained anaemia [1]. Other important systemic symptoms which may dominate the clinical picture include anorexia and weight loss, depression and failure to thrive [1, 2].

Multiple investigations and diagnostic delays are the rule in these patients, as the correct diagnosis may only be considered when local symptoms resulting from cranial branch artery involvement supervene. Yet these patients are as steroid-responsive as those with classical presentations [1, 2].

GCA affects the aorta and its proximal branches as well as the cranial branch arteries supplying extracranial structures. This results in clinical evidence of an aortic arch syndrome in 10–15% of patients, ranging from an arm blood pressure differential to manifest ischaemia [4, 5]. For this reason, the condition is inadequately described by the terms temporal arteritis or cranial arteritis, which refer to the common anatomical domains of symptom expression and do not suggest the systemic nature of the vasculitis.

The subclavian and axillary arteries are the proximal aortic branches most commonly affected [6, 10] and 1–2% of patients with GCA may present predominantly with arm claudication [1, 4]. This shows the importance of not automatically attributing all occlusive arterial disease to atherosclerosis in older patients [10]. Although local signs and symptoms of temporal artery involvement may be absent, temporal artery biopsy is positive in around 80% of these patients [6]. Response to steroids is generally good and surgical intervention is rarely necessary [4–7].

Dissecting and non-dissecting aneurysms of the thoracic aorta are also important complications of GCA, occurring 17 times more frequently than in age-matched controls [8]. They almost always affect the ascending aorta and are usually late sequelae, with ongoing aortitis demonstrable in two-thirds of cases. Late degeneration presumably occurs on a background of previous inflammatory damage in the rest [9]. In one study of 205 patients with GCA, 8% of patients died of aortic rupture over a mean follow-up period of 7 years, although overall mortality did not differ significantly from that of the general population [11]. Although there
is no consensus on screening patients with GCA for this complication, surgically-fit patients should perhaps be followed up annually with a physical examination and chest X-ray [8].

Takayasu’s arteritis shares many clinical and histopathological findings with GCA. However, they are separate entities which can usually be differentiated on the basis of clinical, aortographic and sociodemographic factors. Aortic involvement tends to be more severe in Takayasu’s, with narrowing of the aortic lumen as well as the aortic branch arteries. Aneurysm formation is rare. The cranial branch arteries are rarely involved, so that symptoms such as headache and visual loss are unusual and temporal artery biopsies are usually normal. Affected patients are rarely over 40. By contrast, GCA hardly ever occurs below the age of 50 [7, 12, 13].

The incidence of aortic involvement in GCA is unknown, as patients with early inflammation do not manifest focal signs or symptoms until obliterative disease occurs and therefore do not undergo aortography [8, 9]. However, several strands of evidence suggest that subclinical aortic disease may be frequent. First, subclavian and axillary bruits are unusual in controls but occur in one-third of affected patients [5]. They may appear during the active phase of disease and disappear with treatment. Secondly, post mortem studies indicate that most patients dying during active disease have thoracic aortic involvement, although these patients represent a selected series who might be expected to have more severe and widespread disease [14]. Thirdly, a study evaluating the utility of positron emission tomography in 25 patients with GCA or polymyalgia rheumatica found increased uptake in the thoracic arteries in 56% compared with 2% of controls [15]. Patients presenting with an aortic arch syndrome may therefore represent one end of a disease spectrum [3]. Others may have non-obstructive aortitis, which may account for the important group of patients with GCA who present with predominantly systemic rather than focal features [9].

A broader view of GCA as a disease affecting the aorta and its proximal branches (rather than only its branch arteries) and an appreciation that atypical presentations are in fact common will result in fewer missed and delayed diagnoses.

Key points

- Up to 40% of patients with giant cell arteritis do not present with typical symptoms.
- 10–15% of patients with giant cell arteritis have clinical evidence of aortic arch syndrome.
- Many other patients probably have a non-occlusive aortitis, which may be associated with systemic symptoms.
- It is important to consider this diagnosis in patients with arm ischaemia.

**Acknowledgement**

We are grateful to Barry Morgan for providing a photograph of the temporal artery biopsy specimen.

**References**


Received 21 September 2000; accepted in revised form 2 October 2000