Unusual association of diseases/symptoms

Large-vessel GCA or a late presentation of Takayasu's arteritis?

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Summary

Giant cell arteritis (GCA) is a vasculitis that involves medium- and large-sized vessels. Typically presenting with localised headache, temporal artery tenderness, jaw claudication and ophthalmological complications, it can also affect large vessels, leading to limb claudication. We describe a patient presenting with acute coronary syndrome but who was also noted to have absent upper limb pulses, leading to a speculative diagnosis of GCA. According to the American College of Rheumatology, the patient met the criteria for both giant cell arteritis and Takayasu's arteritis. The gold standard investigation for diagnosing GCA is the temporal artery biopsy. Although contemplated, it was argued by the reviewing rheumatologist that this would not alter her management. She was thus commenced on prednisolone 60 mg daily, which led within 2 days to a rapid fall in erythrocyte sedimentation rate to 66 mm/h, and by day 11 this had dropped further to 4 mm/h.

BACKGROUND

This case is very important as it highlights the limitations of the current diagnostic modalities used in investigating vasculitides. It also demonstrates that there is a significant overlap between Takayasu's arteritis (TA) and giant cell arteritis (GCA), and clinically it is not always possible to differentiate the two conditions and thus the American College of Rheumatology (ACR) criteria used in diagnosing these conditions should be revised.

CASE PRESENTATION

A 75-year-old woman presented with ischaemic sounding chest pain, troponin elevation and ECG changes consistent with non-ST elevation myocardial infarction. In addition, she had a chronic anaemia of unknown aetiology. She complained of a long-standing history of 'cold fingers and toes' and occipital headache. On examination, she had impalpable brachial and radial pulses bilaterally and blood pressure readings were un-recordable in the upper limbs.

INVESTIGATIONS

Investigations revealed an elevated erythrocyte sedimentation rate (88 mm/h) and eosinophilia. On the basis of her presentation, a large-vessel vasculitis was suspected and a CT angiogram was performed that revealed absent flow in the left subclavian artery and minimal flow in the right subclavian artery with collateral formation (figure 1) confirming the diagnosis of vasculitis.

DIFFERENTIAL DIAGNOSIS

- ▶ Large-vessel GCA
- ▶ Takayasu's arteritis
- ▶ Atherosclerosis

OUTCOME AND FOLLOW-UP

This patient has been started with prednisolone and is under review by rheumatologists. Threemonths after diagnosis and

commencing treatment, her blood pressure recordings have become recordable. However, her upper limb pulses are still absent. She continues to be chronically anaemic.

DISCUSSION

The ACR criteria for the diagnosis of GCA are presented in table 1. The patient under discussion met three of the five ACR criteria for establishing this diagnosis. She also met the criteria for diagnosing TA, although TA is most prevalent in patients under the age of 40. Furthermore, recent studies have also suggested that TA and GCA may exist on a spectrum within the same disease due to strong similarities in the distribution of arterial disease in both conditions.¹

Nonetheless, the presentation is also consistent with a diagnosis of large-vessel GCA.² This is a variant of classic GCA and patients must meet the criteria for diagnosing GCA and, in addition, present with symptoms such as intermittent claudication of the arms, impalpable pulses and an inability to measure blood pressure. Typically 5% of GCA patients present this way.³

Temporal artery biopsy (TAB) has several limitations in the assessment of patients with suspected large-vessel vasculitis, most notably false negatives due to skip lesions and a possibility for auriculotemporal nerve damage. Furthermore, TAB cannot distinguish between GCA and TA as both have similar histological features. In recent years, other non-invasive techniques have emerged that are good alternatives to TAB. These include colour Doppler ultrasound of the temporal arteries and positron emission tomography (PET) studies.

Ultrasound of the temporal artery reveals a characteristic halo in patients with GCA. The halo sign represents temporal artery oedema and, when present bilaterally, has a specificity of 100% for diagnosing GCA, although the sensitivity is much lower (41%).

The halo sign has been shown to disappear in patients following successful treatment with steroids,⁴ suggesting

Figure 1 CT angiogram showing bilateral subclavian stenoses.

that this non-invasive technique can be used repeatedly to monitor the response to therapy. It can also be used to guide the region of arterial segment to biopsy and thus reduce the rate of false negatives. Ultrasound is underutilised in diagnosing GCA, owing to its limitations of user dependency and difficulty in interpreting the halo sign under inexperienced hands.

Arterial ultrasound can also reveal distinct findings in TA patients. Ultrasound has been shown to delineate characteristic findings of long segments with homogeneous circumferential arterial wall thickening, described as the macaroni sign.⁵

PET studies are an alternative non-invasive technique to angiography and provide information regarding the vascular wall.

Table 1 American College of Rheumatology criteria for giant cell arteritis (GCA) and Takayasu's arteritis (TA)

GCA	TA
Age >50	Age <40
New onset localised headache	Claudication of the extremities
Temporal artery tenderness/decreased temporal artery pulse	Decreased brachial pulse
Erythrocyte sedimentation rate >50 mm/h	Bruits, for example subclavian bruit
Abnormal artery biopsy specimen characterised by mononuclear infiltration or granulomatous inflammation	Inability to measure blood pressure/ blood pressure difference in the arm
>3/5 required for diagnosis	>3/5 required for diagnosis

This technique has also been shown to assist in the assessment of vasculitis involving intrathoracic vessels, which cannot be readily visualised by Doppler ultrasound. The sensitivity and specificity of PET to detect large-vessel vasculitis are approximately 85% and 95%, respectively.⁶

Learning points

- ▶ Differentiating the different vasculitides is difficult due to overlapping symptoms.
- ▶ The American College of Rheumatology needs to be revised as patients can meet both giant cell arteritis and Takayasu's arteritis criteria.
- ▶ Alternative investigations into the invasive temporal artery biopsy need to be performed in order to help confirm diagnoses.

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Competing interests None.

Patient consent Obtained.

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