Colour duplex ultrasonography in the diagnosis of Takayasu's arteritis: a case report

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Introduction

Takayasu's arteritis (TA) is an inflammatory arteritis involving the aorta and its major branches. Clinical manifestations depend on the site of the vascular lesions. A major complication of the disease is brain ischaemia, which may be related to artery embolism, local thrombosis or, more frequently, haemodynamic deficit. The diagnosis of TA is based on the criteria of the American College of Rheumatology, which take into account clinical and angiographic findings. We highlight the importance of ultrasonography in the clinical diagnosis and management of TA. We describe the case of a 19-year-old woman in which ultrasound was useful in the diagnosis of the disease (in its early phase) and in its management.

Case Report

A 19-year-old woman was admitted to hospital because of fatigue, weight loss and piastrinosis. Her medical history was unremarkable. Physical examination showed a great difference in systolic blood pressure between the two arms together with decreased pulse in the left one. Laboratory tests showed systemic inflammation (ESR 100 mm/1st h and a CRP level of 50 mg/l, nv < 5). Echo-colour Doppler ultrasonography of the upper limb, extracranial and intracranial arteries showed a homogeneous, midechoic, circumferential wall thickening of the common and internal carotid arteries on both sides and of the vertebral, axillary and humeral arteries on the left side (Fig. 1). Positron emission tomography (PET) demonstrated inflamed areas on the subclavian artery. During the follow up, by monitoring the wall thickness we were able to assess the response to therapy.

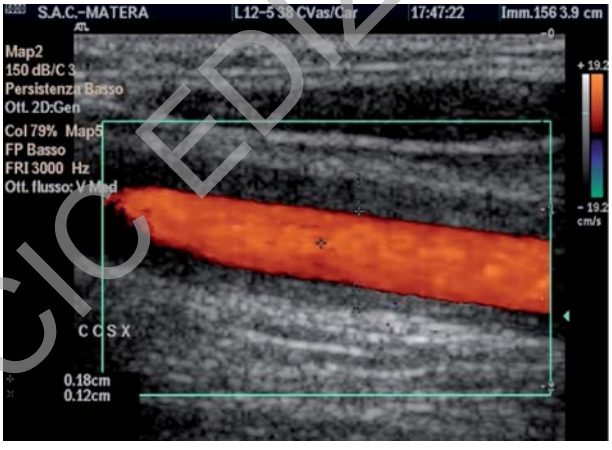

Fig. 1 - Colour duplex ultrasound showing the "macaroni sign" (homogeneous, midechoic and circumferential wall thickening) at the left common carotid artery.

Discussion

TA is an inflammatory disease of the wall of the aorta and its main branches. Vessel wall inflammation leads to stenosis and occlusion or aneurysm, or both, of the involved artery. The diagnosis is based on the criteria of the American College of Rheumatology (1). Three of the following six criteria must be present:

1) Onset at age < 40 years
2) Claudication of an extremity
3) Decreased brachial artery pulse
4) Greater than 10 mmHg difference in systolic blood pressure between the right and left arms
5) A bruit over the subclavian arteries or the aorta
6) Arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities.

Several studies have highlighted the diagnostic potentialities of duplex ultrasonography in TA (2). Nevertheless, so far there is no general agreement on the advantages of this method compared with other diagnostic tools, such as angiography or PET.

Duplex ultrasonography has certain advantages over angiography and PET in the detection of TA. Indeed a homogeneous, midechoic, circumferential wall thickening of the arteries on ultrasound examination is typical of TA and has been called the "macaroni sign". This finding is brighter than the typical "dark halo" sign of giant cell arteritis. Nowadays, improved ultrasound technology makes it possible to visualise discrete wall vessel alterations as well as to distinguish inflamed from atherosclerotic wall lesions. In TA, angiography can show important luminal stenoses in many vessels including small arteries. However, this technique cannot visualise the vessel wall and can miss mild wall lesions without stenosis. PET shows inflamed arterial areas but cannot provide information on vessels with a diameter <4mm and details of the vessel wall.

Moreover, colour duplex ultrasonography can easily detect the activation of the compensatory flow of the extracranial and intracranial vessels in cases of haemodynamic stenosis or occlusion of any cerebral vessel. This is very important because the most important cause of brain ischaemia in TA is a haemodynamic deficit (3).

Although the intracranial arteries are infrequently involved compared with the extracranial internal/common carotid artery, there are studies that highlight intracranial vessel involvement in TA (4). We therefore think that transcranial colour Doppler should be mandatory not only for the study of compensatory flow but also to detect intracranial stenosis in TA.

Moreover, ultrasonography is suitable for long-term repeated follow up of TA because is a non-invasive, efficient, low-cost method that can describe the temporal profile of vascular lesions. By monitoring the homogeneous, midechoic, circumferential wall thickening of the arteries we were able to assess more efficiently the response to therapy.

In conclusion, we regard ultrasound as an adequate method for the diagnosis of Takayasu's arteritis in young patients with chronic inflammation of unknown origin.

References

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