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Case report

The role of ultrasound in the diagnosis of temporal arteritis

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ARTICLE INFO

Article history:
Received 5 August 2014
Accepted 10 March 2015
Available online 20 March 2015

Keywords:
Temporal arteritis
Giant cell arteritis
Ultrasound

ABSTRACT

Temporal arteritis (TA), also known as giant cell arteritis, is a chronic vasculitis of medium and large-sized blood vessels, in particular the main cervical branches of the aorta, with particular affinity to the temporal arteries and eye-supplying arteries. Temporal artery biopsy is still a gold standard for diagnosis, however in recent years colour duplex ultrasound examination has been proposed as a useful diagnostic screening tool in cases of TA suspicion. We report three cases of TA in which the ultrasonographical examination of the temporal arteries had a decisive role in the diagnosis.

1. Introduction

Temporal arteritis (TA), also known as giant cell arteritis, is a chronic vasculitis of medium and large-sized blood vessels, in particular the main cervical branches of the aorta, with particular affinity to the temporal arteries and eye-supplying arteries [1]. The involvement of the vertebral arteries, carotid arteries, the aorta itself and coronary arteries is less common [2].

The disease occurs in all cases in patients older than 50 years, the mean age at diagnosis being approximately 72 years [3].

Disease susceptibility has been associated with European descent, the highest incidence being found in Scandinavian countries and among Americans of Scandinavian descent [4,5].

The most feared complication of TA is visual loss, however in rare cases, stroke can occur (3–7% of cases) and is the leading cause of death in patients with TA [6,7]. Cerebral ischaemic events in patients with TA are mainly due to the involvement of the extradural vertebral and carotid arteries rather than to the vasculitic involvement of the intracranial vessels [6,8].

For the purpose of differentiating TA from other forms of vasculitis, the American College of Rheumatology formulated five classification criteria for TA: age ≥50 years at onset, localised headache of new onset, tenderness or decreased pulse of the temporal artery, erythrocyte sedimentation rate > 50 mm/h and biopsy revealing a necrotising arteritis. The presence of three of these five criteria is associated with 94% sensitivity and 91% specificity for the diagnosis of TA [9,10].

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http://dx.doi.org/10.1016/j.pjnns.2015.03.003

Temporal artery biopsy is still a gold standard for diagnosis, however in recent years colour duplex ultrasound examination has been proposed as a useful diagnostic screening tool in cases of TA suspicion [6,11,12].

We report three cases of TA in which the ultrasonographical examination of the temporal arteries had an important role in the diagnosis.

The examinations were performed by the same skilled investigator using a Siemens Acuson Antares ultrasound device with a VFX13-5 MHz linear transducer. The B-mode, Colour mode and duplex settings were adjusted for best assessment of the vessels. The temporal artery trunk, frontal and parietal branches were scanned in the axial and longitudinal sections. The latter section was used to detect stenoses. Stenosis was considered to be present if the flow velocity was two times higher comparing to the velocity measured in the area before stenosis.

2. Case reports

2.1. *Case 1*

An 80-year-old woman with negative medical history was admitted with a 1-month history of bitemporal headache, jaw claudication, diplopia and progressively decreasing visual acuity (left > right) that had appeared one week before. Clinical examination revealed necrotic cutaneous lesions in the temporal and frontal region of the head and induration on palpation of the temporal arteries. Neurological examination evidenced decreased visual acuity bilaterally (0.1 left eye, 0.6 right eye) and right-sided abducens nerve palsy. Laboratory analysis was relevant for increased erythrocyte sedimentation rate (ESR), 95/1 hour, thrombocytosis (507000 mm^{-3}), positive C reactive protein, elevated fibrinogen level (682 mg/dl) and mild mixed dyslipidaemia. Cerebral MRI revealed only mild cerebral atrophy, lacunary infarctions and leukoaraiosis. Duplex ultrasound examination of the cervical arteries revealed no pathological lesions. Duplex examination of the temporal arteries described the presence of the hypoechogenic halo (the halo thickness was between 0.6 and 0.9 mm) sign in both frontal and parietal branches of the temporal arteries (Fig. 1). Based on this workup the diagnosis of temporal arteritis was established without biopsy and high dose corticosteroid treatment was initiated (1 mg/kg/day prednison) followed by prednisone tapering. The outcome was favourable excepting the left-sided visual impairment.

2.2. *Case 2*

A 65-year-old woman with medical history relevant only for osteoporosis was admitted with a 2-month history of generalised headache, jaw claudication, pain and stiffness in the neck, shoulder and hip girdles and weight loss (3 kg in 1 month). Clinical examination revealed tender and thickened temporal arteries bilaterally, with right predominance, decreased pulse at this level and tenderness at the level of cervical spine and shoulder girdles. The neurological and ophthalmological examinations were normal. The laboratory workup was relevant for elevated ESR (76/1 hour) and mild normochromic anaemia. Duplex ultrasound examination of the temporal arteries revealed a hypoechoic halo sign in all branches of the temporal arteries (halo thickness between 0.3 and 1.1 m) (Fig. 2) and stenosis at the level of the left temporal artery trunk. Based on these findings, the diagnosis of temporal arteritis was established and treatment with corticosteroids was initiated (24 mg Medrol/day). The outcome was favourable, 2 weeks after the initiation of the corticotherapy the patient was asymptomatic.

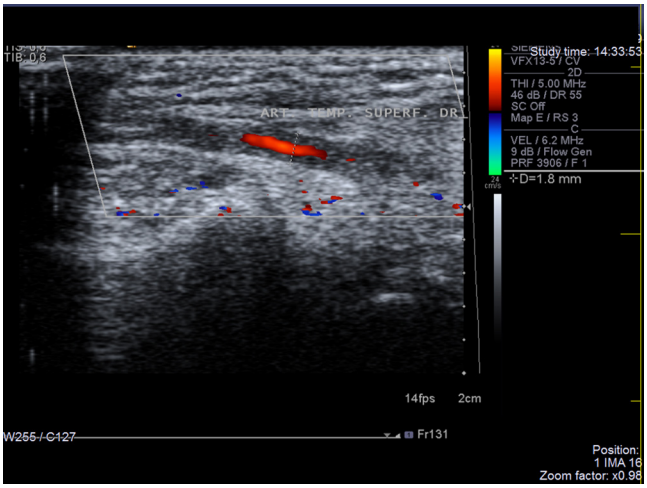

Fig. 1 – Case 1. Temporal arteritis in a 80-year-old woman. Colour duplex ultrasound examination of the temporal artery, longitudinal section, showing pronounced hypoechoic mural thickening ('halo sign').

2.3. *Case 3*

A 72-year-old hypertensive woman, without any other relevant findings in her medical history, was admitted for sudden onset vertigo, disequilibrium, nausea and vomiting, hiccups presented on awakening. Clinical examination was relevant for higher blood pressure values (156/100 mmHg). Neurological examination revealed a clinical picture suggestive of a right-sided Wallenberg syndrome. Cerebral MRI described a right-sided dorsal lateral medullary infarction. The laboratory workup was relevant only for thrombocytosis (667000 mm^{-3}); ESR, PCR and Fibrinogen were not performed. Cervical duplex ultrasound examination evidenced no stenotic lesions at the level of the carotid arteries and a significant hypoechoic mural thickening of the right vertebral artery in V1 and V2 level (the maximal thickness was 3.6 mm) (Fig. 3a). The first impression of the ultrasonographer was that this was a vertebral artery dissection, but the ultrasonographical examination of the temporal arteries revealed the same hypoechoic mural thickening (between 0.7 and 1.1 mm), that was suggestive for arteritis (Fig. 3b). Temporal artery biopsy revealed a granulomatous process with multinuclear giant cells and confirmed the diagnosis of temporal arteritis. The patient was treated with high dose corticosteroids (16 mg Dexamethasone/day, 10 days), followed by prednisone tapering, antiplatelets and antihypertensive medication with good outcome.

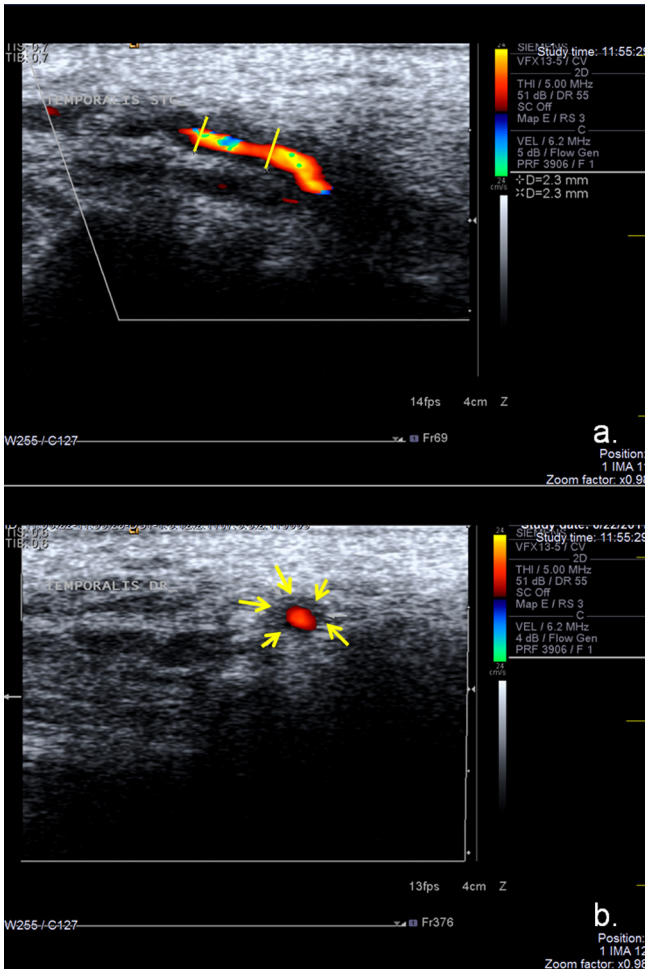

Fig. 2 – Case 2. Temporal arteritis in a 65-year-old woman. Colour duplex ultrasound examination of the temporal artery, longitudinal (a) and axial (b) sections, showing pronounced concentric hypoechoic mural thickening – 'halo sign' (lines and arrows).

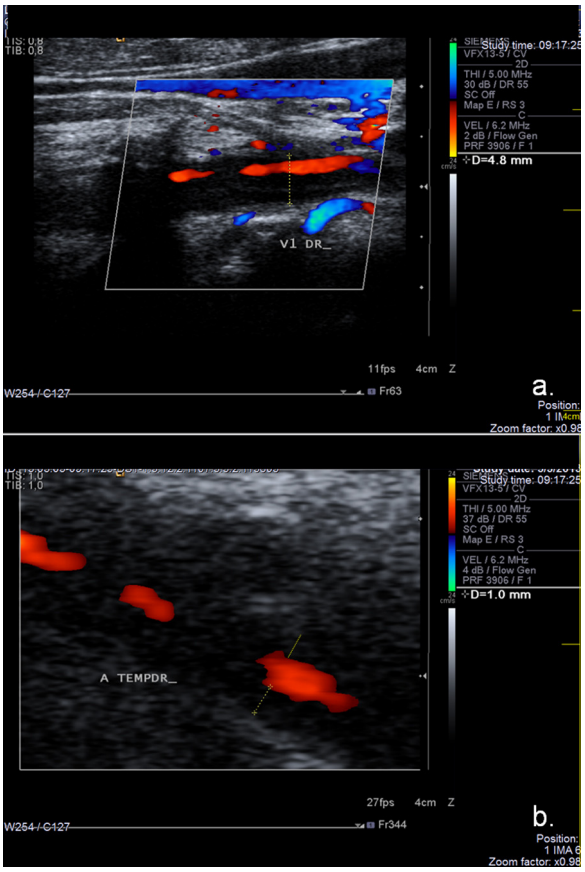

Fig. 3 – Case 3. Temporal arteritis in a 72-year-old woman. Colour duplex ultrasound examination of the right vertebral (a) and temporal (b) arteries (longitudinal sections) showing a pronounced eccentric hypoechoic mural thickening of the vertebral artery (a) and concentric hypoechoic mural thickening of the temporal artery (b).

3. Discussion

The usefulness of ultrasound examination for the diagnosis of TA is not yet well established. There are data in the literature debating its specificity and sensitivity compared with biopsy that is still the gold standard for diagnosis [9].

Schmidt et al. [13] first described the oedematous wall swelling of the temporal arteries, characterised echographically as a hypoechoic circumferential mural thickening localised around the lumen, with a diameter ranging from 0.3 to 0.5 mm. This finding was denominated as the 'halo sign' [13]. Two other parameters considered relevant for the diagnosis of TA were described: stenosis and occlusion. Stenosis, characterised by a narrowing of the lumen, was defined as a segmental increase in blood flow velocity two times greater than in the region before the stenosis. Acute occlusion is revealed by the absence of colour signal in a segment of temporal artery [13].

The same author [11] published a paper in 1997 about the usefulness of ultrasound in TA diagnosis in a cohort of 30 patients. Out of 30 patients with TA, 73% had a 'halo sign', 80% had stenosis or occlusion and 93% stenosis, occlusion or 'halo sign'. They concluded that in patients with a typical 'halo sign' it may be possible to make a diagnosis of TA and begin treatment without performing biopsy. Similar results were described on a larger cohort (101 patients) in a paper published in 2003 [14]. The sensitivity of ultrasound was highest when all three findings (halo, stenosis and occlusion) were present. The sensitivity of halo alone was lower, but the specificity was high [14].

In 2005, a meta-analysis including the most important studies published up to 2004 involving 2036 patients was published. The sensitivity and specificity of the three ultrasound criteria were calculated as follows: halo sign 69% and 82%, stenosis or occlusion 68% and 77%, halo, stenosis or occlusion 88% and 78% [9,15].

In a more recent meta-analysis including 17 studies containing 998 patients, Ball et al. found a 75% sensitivity and 83% specificity for the halo sign compared with biopsy [16].

Arida et al. [17] published a meta-analysis in the same year including eight studies with 575 patients, with similar results: unilateral halo sign achieved an overall sensitivity of 68% and specificity of 91%. Bilateral halo signs implied a 43% sensitivity and 100% specificity.

The typical halo sign can be observed also in other vessels affected by the vasculitic process such as vertebral, occipital or axillary arteries, however ultrasonographical examination is more difficult and positive findings are rare [6,18]. The vertebral artery halo sign, as in our third case, can be the first clue for early diagnosis of TA.

García-García J et al. [6] examined the ultrasonographical findings of vertebral arteries in patients with TA. They examined ultrasonographically 1237 acute stroke patients in the first 24 h from onset. They found concentric, homogenous, smooth hypoechoic mural thickening of the vertebral arteries (halo sign) in only five cases, all of which had vertebro-basilar territory infarction and, in all of the cases, temporal artery biopsy confirmed the diagnosis of TA. They concluded that the recognition of the halo sign in vertebral arteries may be of crucial interest in selected cases [6].

The most important differential diagnostic issue in cases of vertebral artery involvement is dissection, however in this latter situation, hypoechoic vessel wall changes are usually eccentric and crescent-shaped, often with spiralling course [19]. In our third case, the hypoechoic mural thickening was eccentric, resembling dissection, but the temporal artery findings and biopsy results were conclusive for the diagnosis.

For correct diagnosis, the examination technique and the experience of the sonographer is very important. There is a need for a high quality colour duplex ultrasound device, with standardised adjustments and a high frequency (>8 MHz) linear transducer [20]. According to Schmidt et al. it is recommended that a minimum of 50 normal temporal artery examinations are performed before starting the evaluation of patients with suspected TA [20].

False positive and negative halos may be seen in ultrasound examination. Ultrasound is not able to differentiate between TA and other vasculitis that can involve the temporal arteries [21].

Aschwanden M et al. published in 2002 a new ultrasound finding, called ''temporal artery compression sign'' that is largely operator-independent and elicits the contrasting echogenicity between the diseased artery wall and the surrounding tissue. They concluded that the halo sign and the compression sign were equal in their diagnostic performance [22].

In conclusion, we presented three classical cases of TA, with characteristic ultrasound findings, highlighting the role of this non-invasive examination in diagnosis. The third case demonstrated that the classical halo sign can be present at the level of the vertebral arteries and can be the key to correct diagnosis of temporal arteritis.

Conflict of interest

None declared.

Acknowledgement and Financial support

This publication was supported by the Internal Research Grant of the University of Medicine and Pharmacy Târgu Mureş, 26/11.12.2013.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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