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Case Report

Temporal superficial arteritis as differential diagnosis in patients with atherosclerotic changes due to advanced chronic renal disease: case report and review of the literature☆

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ABSTRACT

Giant cell arteritis is an autoimmune disease that affects medium and large caliber vessels, creating deposits of inflammatory clusters on the arterial wall. It is the most common form of large vessel vasculitis, but given the variability of biopsy efficiency and of other diagnostic strategies employed, the diagnosis of this disease is challenging. We report the case of a 69-year-old female patient who presented with neurological deficit and increased bilateral sensation in the temporal region associated with excruciating headache. Workup revealed calcification of the superficial temporal, vertebral and ophthalmic arteries, as well as suggestive findings on Doppler ultrasound such as the halo sign, pointing to superficial temporal arteritis though not excluding the possibility of those calcifications being consistent with atherosclerosis in a patient with advanced chronic renal disease, which has been reported as giving rise to false-positive results. Knowledge of the main differences between the 2 diagnoses is important, given the wide range of diagnostic imaging possibilities which can avoid the need for biopsy.

Introduction

Giant cell arteritis (GCA) is an inflammatory vasculitis affecting medium and large vessels and it is the most frequent form of vasculitis that occurs in the elderly. It is characterized for being systemic and of granulomatous type and is more frequent among women, with a median age of onset of 75 years [1]. It must be suspected in the presence of clinical signs such as headache, amaurosis fugax, transient ischemic attacks or

Abbreviations: GCA, GIANT CELL ARTERITIS.

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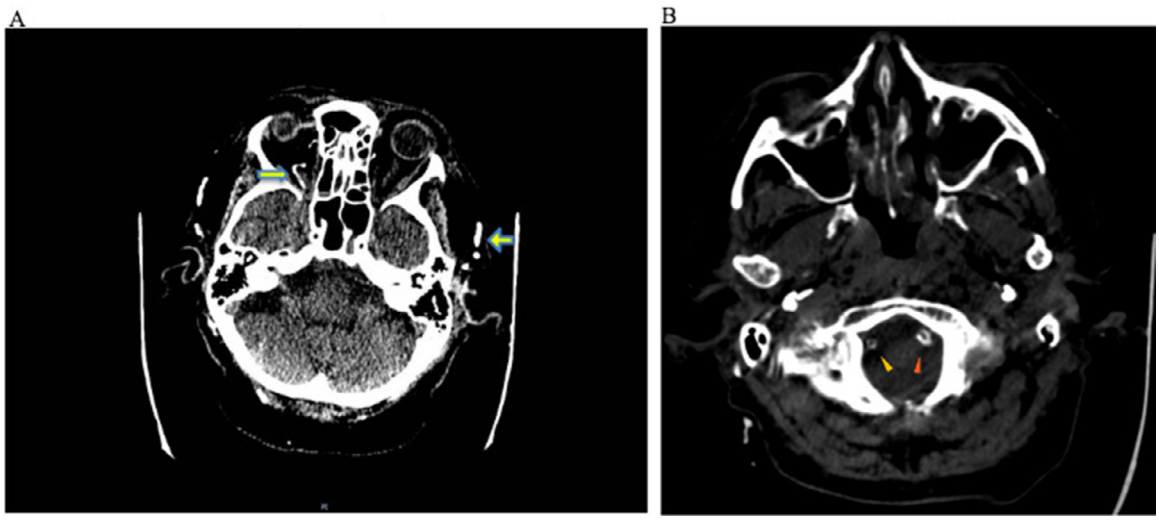

Fig. 1 – Plain cranial CT scan. (A) Calcification of temporal arteries and of the right ophthalmic artery. (B) Bilateral calcification of the vertebral arteries.

stroke, the latter occurring as early symptoms of the disease, 73% at the time of diagnosis, and 27% within the first 2 weeks [2]. It is associated with radiological signs such as temporal artery (TA) calcifications, the halo sign on Doppler ultrasound and the absence of arterial compressibility, the most frequent being the latter 2 [3]. We report the case of patient who presented with neurological deficit and was found to have bilateral calcifications of the superficial temporal artery, the ophthalmic artery and the vertebral arteries, as well as the halo sign on superficial temporal artery Doppler.

Case description

A 69-year-old female patient attended a tertiary level hospital due to surgical site infection one month after supracondylar amputation of the right femur and left transtibial amputation secondary to bilateral peripheral arterial occlusive disease. The patient also presented on admission, signs of neurological focalization in the form of dysarthria, facial palsy and left hemiparesis, predominantly of the left upper limb and new onset biparietal throbbing headache of medium intensity. Her medical history included chronic renal disease needing renal dialysis every other day, diabetes mellitus type II, arterial hypertension, heart failure with preserved left ventricular ejection fraction, acute myocardial infarction without coronary disease, and cor pulmonale. Plain computed tomography (CT) imaging of the brain showed an ischemic cerebrovascular event in the right posterior cerebral artery territory in the right occipital region, NIHSS score of 3, with no indication for thrombolysis. Dual antiplatelet therapy was initiated before referring the patient to our institution for comprehensive management.

During the review by systems on admission the patient reported dyspnea with mild exertion, recovery of her strength in the left side of the body. She did not report visual acuity abnormalities, mandibular claudication or constitutional symptoms. On initial physical examination, vital signs were normal, with mildly diminished temporal pulses and increased bilateral tenderness on palpation over the temporal region, symmetrical chest with evidence of systolic murmur, no stridor on lung auscultation, and normal abdominal examination; left lower limb with transtibial amputation stump and right lower limb with supracondylar amputation stump, both with single suture margin approximation, in the process of healing. On neurological examination, the patient was alert, focused, oriented in 3 spheres, with mild dysarthria, could repeat and understand, and had preserved remote and episodic memory. Cranial nerves with normal reactive isocoric pupil, preserved ocular movements, facial symmetry and preserved sensation. Strength 5/5 in right upper limb and 2/5 in left upper limb. Osteotendinous reflexes ++/++ bilateral flexion strength, no cerebellar signs, normal gait and no dissymmetry or meningeal signs.

On admission to our institution, acute phase reactants were high, with elevated erythrocyte sedimentation rate (ESR) (65 mm/h); a follow-up plain cranial CT scan showed periventricular hypodensity possibly due to small vessel hypoxic/ischemic attack, an 8 mm hypodensity of the cortical-subcortical junction in the frontal region in the middle cerebral artery territory, and poorly defined acute phase hypodensity of ischemic type in the right occipital region, in the territory of the right posterior cerebral artery. Other relevant findings included the presence of calcifications of the vertebral arteries, of critical grade in the left vertebral artery, as well as multiple bilateral calcifications of the superficial temporal artery, involvement and calcification of the ophthalmic arteries, suggesting temporal arteritis as the primary cause, versus atherosclerotic changes secondary to chronic renal disease.

On follow-up, the patient refused to undergo any invasive intervention, precluding the performance of the superficial temporal artery biopsy. However, a Doppler ultrasound of the superficial temporal artery was performed and showed

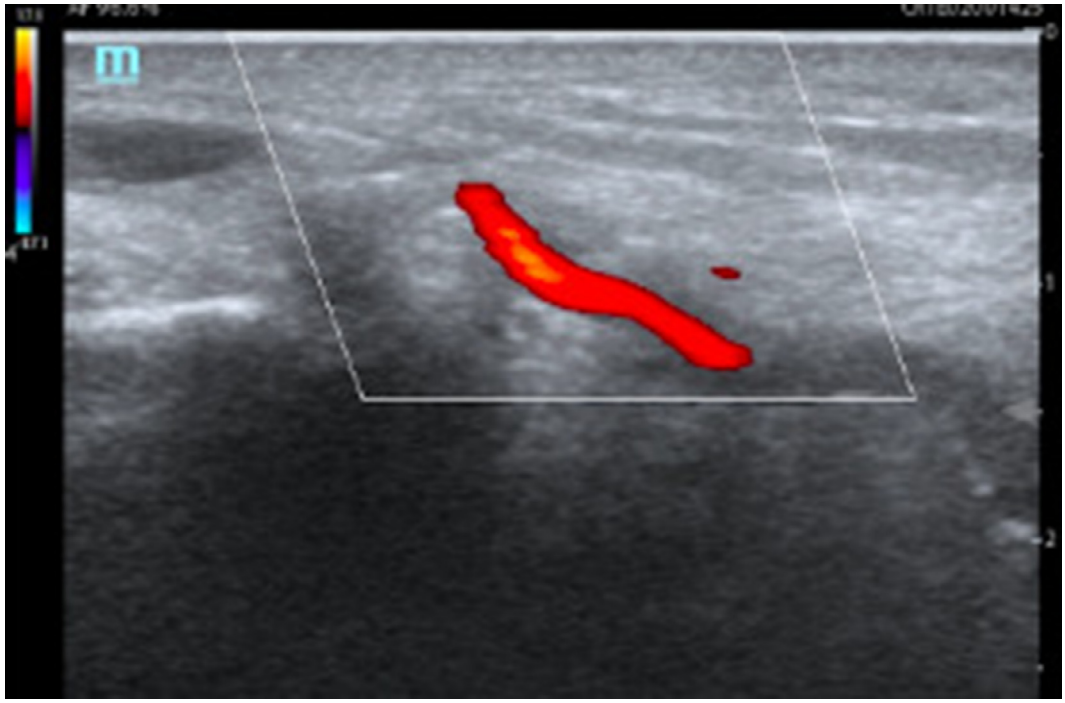

Fig. 2 – Doppler ultrasound in where it seems a temporal artery wall thickened.

characteristic findings such as thickening of the external carotid artery wall and increased resistance in the temporal arteries.

Discussion

Temporal arteritis, also known as giant cell arteritis, is a form of vasculitis affecting medium and large caliber blood vessels. It can be intra or extracranial, depending on the branches involved, but has greater predilection for the temporal and vertebral arteries, hence the associated clinical manifestations [5]. It occurs more frequently in white women (2:1 F:M ratio) and in individuals over 50 years of age. It is the most common systemic vasculitis of the granulomatous type with chronic presentation; the annual incidence varies between 1.6 and 32.8 cases × 100,00 inhabitants in Europe, USA, Israel, and Oceania; epidemiological data for Colombia are nonavailable [4]. The triggering agent for this autoimmune disease occurring in patients with predisposing factors and HLA class II polymorphisms is unknown [8]. Histologically, there is vascular infiltration of CD4+ T cells and macrophages that fuse to form "giant cells" that are deposited on the vessel wall, leading to arterial intimal hyperplasia and thickening, reduced blood flow due to stenosis, and arterial occlusion, and giving rise to the typical symptomatology of this disease [2,4].

The main clinical manifestations have been divided into 2 presentation patterns, depending on the affected vessel. The most frequent with severe manifestations and more prevalent in the elderly population is the cranial presentation characterized by headache, predominantly temporal in 70%-80%, with scalp hypersensitivity and mandibular claudication; it can even trigger visual loss secondary to ischemic optic neuritis. On the other hand, the extracranial phenotype, also called giant cell arteritis, affects younger patients and involves the aorta and its proximal branches, the main associated symptoms being fever, upper limb claudication, aortitis, and constitutional symptoms such as weight loss, asthenia and myalgia [4,6].

Table 1 – Clinicopathological criteria from the American College of Rheumatology: 3 out of 5 criteria are necessary to suspect medium vessel vasculitis [4]

1. Age of onset of the disease of 50 years or more
2. New onset headache or location type
3. Tenderness on palpation of the temporal artery or reduced pulse not related to cervical arterial atherosclerosis
4. Erythrocyte sedimentation rate 50 mm/H or greater
5. Positive temporal artery biopsy

In 1990, the American College of Rheumatology established clinical criteria to help with the classification and suspicion of this condition. These criteria are still valid and consist of 4 of the cranial symptoms and one temporal artery biopsy and the disease is suspected if 3 of these are present (Table 1) [5].

The test of choice for the definitive diagnosis of this condition is still a subject of debate. Superficial temporal artery biopsy is found throughout the literature as the gold standard for confirming the diagnosis. However, its sensitivity varies between 39% and 91% given that involvement is segmental and prior glucocorticoid treatment can result in a high number of false negative results [7]. As part of the search for

Table 2 – Radiological characteristics of temporal superficial arteritis on Doppler ultrasound, the pathognomonic finding is the halo sign [7]

Signs	Characteristics
Halo	Eccentric hypoechoic circumferential ring around the vessel (Fig. 2) [7]. Thickness greater than 1.5 mm, highly predictive of GCA [5]. It is the most specific sign.
Vascular occlusion	Secondary to vessel wall edema. Aliasing is seen due to focal velocity increase with low-resistance beyond the stenosis [3].
Stenosis	Peak systolic velocity (PSV) at the site of the TA stenosis is 2 or more times greater than PSV distal or proximal to the stenosis [3].
Arterial compression	Persistence of the halo with compression [3] (Fig. 3).

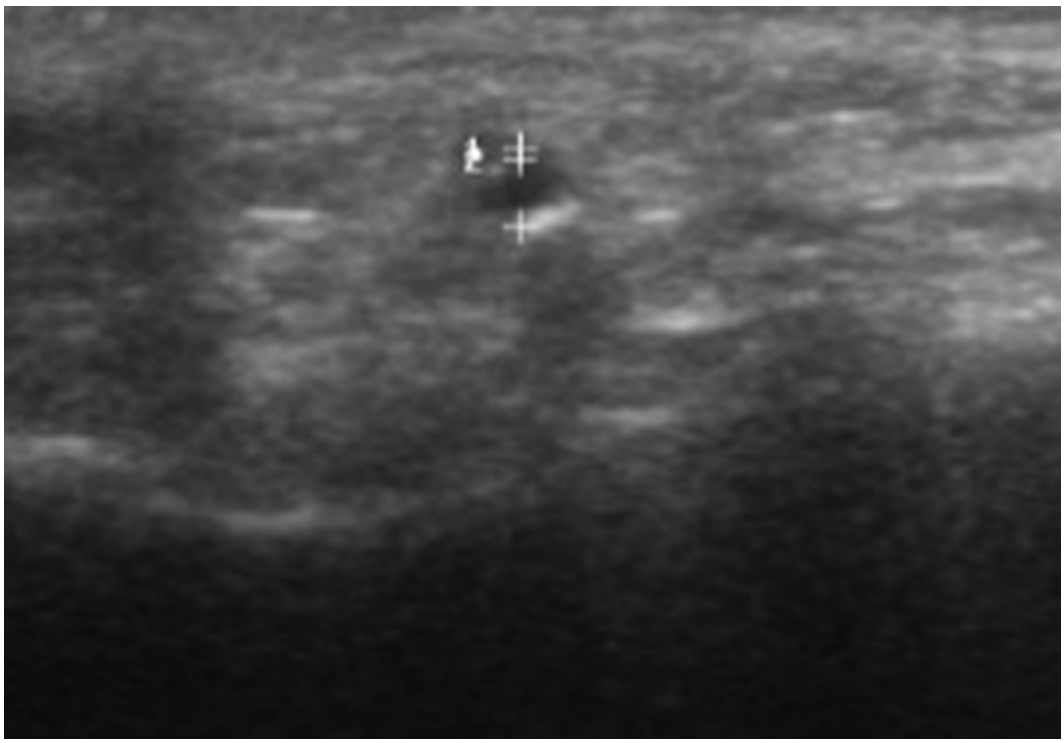

Fig. 3 – Persistent TA hypoechoic halo despite compression made by echographic transducer.

new, less invasive diagnostic options with high sensitivity, the European League Against Rheumatism, in its latest release, recommends Doppler ultrasound as the first imaging study of choice when temporal arteritis is suspected, avoiding the need for biopsy [5].

Doppler has shown to be a very useful diagnostic test because of accessibility, noninvasiveness and low cost, although it is operator-dependent. The ultrasound assessment consists of examining the temporal artery and its branches, the external carotid artery and the axillary artery mainly, but it can also include the occipital artery or the facial arteries [8]. The most common signs are the presence of a hypoechoic halo, noncompressible vessel, stenosis and calcification (Table 2). CT and magnetic resonance imaging also help as part of the diagnostic workup, showing calcifications of the temporal, ophthalmic and vertebral arteries (Fig. 1), taking into consideration the main differential diagnoses, namely, atheromatosis and changes due to chronic renal disease.

In 2014, Diamantopoulos et al., in a prospective, noncontrolled cohort study, were able to determine that signs like the temporal artery halo had a sensitivity of 96% and a specificity of 90% for diagnosis; the halo in the temporal or the axillary artery, a sensitivity of 98% and a specificity of 91%, and the halo in the temporal + axillary + common carotid arteries had a sensitivity of 100%. On the other hand, Aschwanden et al., in another noncontrolled prospective cohort study in 2015, found that the noncompressibility sign had a sensitivity of 79% and a specificity of 100%. This shows that the use of imaging-based diagnosis is supported in the literature, although the interpretation and any potential limitations of Doppler ultrasound depend on the technique and the expertise of the radiologist in identifying the findings [4].

Moreover, there is the question of whether imaging findings may be consistent with atherosclerotic changes in a patient with end-stage chronic renal disease, given that calcification of medium caliber vessels has also been described, creating false-positive results. In a retrospective study, Anwar et al. found that 9.9% of a population of 453 patients with end-stage chronic renal disease showed TA calcification, with calcification of the intima or the media [9].

The objective of this case report is to propose the possibility of diagnosing temporal arteritis without the need for biopsy, considering that in the context of our patient, clinical and imaging findings led to the suspicion of this disease condition based on the finding on cranial CT of bilateral calcifications of the superficial temporal artery as well as of the ophthalmic and vertebral arteries. The additional use of Doppler ultrasound increases the odds of diagnosing the disease based on the presence of the hypoechoic halo and the compressibility signs described above.

In the case of this patient who refused temporal artery biopsy, based on the epidemiological characteristics, the reported symptoms, the physical examination, elevated acute phase reactants and the signs seen on cranial CT and Doppler ultrasound, the most probable diagnosis is temporal arteritis. Other potential etiologies such as calcifications found in chronic renal disease cannot be ruled out. However, so far there is no consensus to allow the appropriate distinction between vessel wall inflammation and atherosclerotic changes [4].

Patient consent

The tomography images such as the Doppler of the superficial temporal artery and the clinical history have been authorized by informed consent by both the patient and her daughter. Formal consent form available upon request.

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