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CASE REPORT

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Color Doppler imaging of a temporal artery – a new diagnostic tool in giant cell arteritis

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ABSTRACT

Giant cell arteritis (GCA) is the most common potentially sight-threatening vasculitis. The typical clinical image with headache or jaw claudication accompanied by erythrocyte sedimentation rate (ESR) over 50 mm/h, confirmed by inflammatory features in temporal artery biopsy (TAB), is essential for GCA diagnosis. About 30% of patients experience a sudden loss of vision in one eye. Delay in treatment may lead to bilateral blindness, which makes GCA an ophthalmological emergency.

An 80-year-old male patient was admitted to the ophthalmological emergency due to sudden and permanent vision loss of his left eye. He also complained of recurrent compressive headaches. However, he denied a history of general weakness, or jaw claudication. The distance visual acuity in the left eye had decreased to 0.1 and a blurred, elevated left optic disc in indirect ophthalmoscopy was observed. In palpation the temporal artery pulse was detectable. Interestingly, vascular smooth muscle thickening was detected in TAB. Despite the uncharacteristic TAB result and ESR equal to 27 mm/h, temporal artery color Doppler imaging (CDI) revealed the "halo" symptom confirmed by the pressure sign. According to typical features in CDI, GCA was diagnosed and the proper treatment was implemented. As a result, substantial resolution of inflammatory parameters in blood tests and CDI was observed. During the 24-month follow-up the good general and ophthalmological condition was maintained.

Despite the lack of typical symptoms in the presented case, CDI determined quick and accurate diagnosis for proper treatment implementation. Color Doppler imaging is a non-invasive and highly specific tool for both diagnostics and treatment monitoring in GCA.

KEY WORDS: asymptomatic, color doppler imaging, diagnostics, giant cell arteritis, temporal artery biopsy.

INTRODUCTION

Giant cell arteritis (GCA) is the most common vasculitis affecting large and medium arteries, especially the aorta and its major branches. The disease usually occurs in female patients older than 50 years. The GCA prevalence for Northern Europe is about 20 per 100 000 and much lower for Southern Europe – 1.1 to 11.1 [1]. Moreover, due to the aging population, the number of patients affected by GCA is expected to rise [2].

GCA's representative symptoms are headaches, scalp hypersensitivity, jaw claudication, and systemic symptoms, including malaise, weakness, subfebrile temperature, and weight loss [3]. About 1/3 of patients experience a visual loss, which almost always remains permanent. Consequently, it makes GCA an ophthalmological emergency. Early diagnosis and proper treatment are essential. Nevertheless, the mean estimated diagnostic delay stands at nine weeks [4].

In 1990 the American College of Rheumatology (ACR) established criteria for the classification of GCA including: age over 50 years, new headache, temporal artery abnormality, elevated erythrocyte sedimentation rate (ESR) – usually above 50 mm/h, and abnormal artery biopsy [5]. Accordingly, temporal artery biopsy (TAB) was for years considered the gold standard for GCA diagnosis. Currently, thanks to the development of high resolution imaging, biopsy is gradually being replaced by some noninvasive techniques [6]. Introduction of novel applications of diagnostic tools such as color Doppler imaging (CDI) in GCA may improve the diagnostic time, and shorten the time untill treatment implementation. The CDI is commonly used in rheumatology, but it still remains unfa-

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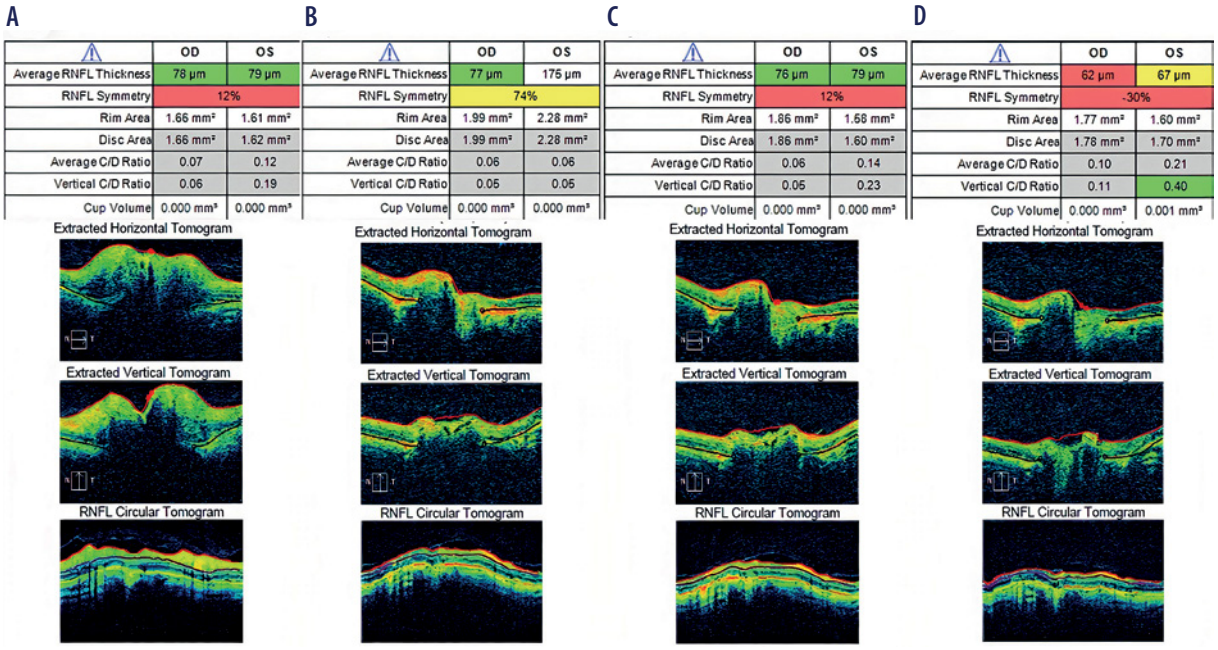

Figure 1. Optical coherence tomography results of the optic disc in patient with giant cell arthritis (A) at baseline, (B) 1 month after the first cycle of intravenous steroid treatment, and (C) 12 months after the ocular symptoms onset, and (D) 24 months after the ocular symptoms onset

miliar for a wide range of ophthalmologists. The aim of this case presentation is to report CDI efficiency in identifying GCA and implementation of the right treatment to the patient without pathognomonic symptoms.

CASE PRESENTATION

An 80-year-old hypertensive, dyslipidemic male patient sought medical attention in the ophthalmological emergency due to a permanent vision loss of his left eye that appeared the same day. This symptom occurred suddenly and painlessly. Interestingly, the day before, he had been examined while adjusting spectacle correction and there had been no abnormalities or permanently decreased visual acuity revealed. This development of the clinical condition confirms an acute onset of the symptom. The patient complained of intermittent doubled vision and a history of episcleritis of the left eye in the past.

His past medical history included recurrent, bilateral, compressive headaches, especially in the temporal and occipital areas. The patient estimated the pain intensity at 7-10 according to the Visual Analogue Scale (VAS). The pain was intensified by looking up and it was passing spontaneously or after pain reliever intake. The patient reported deterioration of vision and narrowing of the visual field during exacerbation of the headache. However, he did not report experiencing photophobia or sensitivity to sounds. Due to the recurrent character of the headache, neurological examination and brain CT scan were carried out prior to the ophthalmological incident. However, no abnormalities were detected. There was no history of systemic symptoms including weakness, weight loss, lack of appetite, subfebrile temperature, or malaise.

During the initial ophthalmological evaluation, best corrected visual acuity in the left eye was 0.1 and in the right eye was 1.0 measured on a Snellen letter chart. Furthermore, both pupils remained symmetrical with normal direct and indirect light reflex. Slit-lamp examination of the anterior segment showed a nuclear cataract in both eyes. In indirect ophthalmoscopy blurring and elevation of the optic discs were observed. The picture of asymmetrical papilledema, more prominent on the left side, was confirmed in optical coherence tomography (OCT, Figure 1A). Additionally, arteriovenous nicking in fundoscopy was detected. Moreover, kinetic perimetry results showed altitudinal visual field loss in the lower hemisphere of the left eye (Figure 2A).

Based on the medical history and clinical presentation the initial diagnosis of anterior ischemic optic neuropathy (AION) was established. Hyperlipidemia, hypertension, internal carotid artery obstruction, or GCA was considered as a primary cause. Due to the history of undetermined headaches, the GCA diagnostics was continued. However, in palpation of temporal arteries no abnormalities were revealed. Similarly, jaw claudication or scalp tenderness was not detected. Blood tests revealed ESR equal to 23 mm/h and CRP was 1.7 mg/l. Additionally, fibrinogen level was increased to 353.3 mg/dl. According to the findings unspecific for GCA and urgent need to start steroid therapy in case of disease confirmation, the decision of CDI of the superficial temporal artery was made. As a result, the periluminal halo sign (presence of hypoechoic, increased thickness of the vessel wall, resulting from inflammatory infiltration) and a positive compression sign (lack of disappearance of hypoechoic rim after closing vessel lumen by applying pressure with the ultrasound probe), characteristic for GCA, were detected (Figure 3A; Supplementary data 1). Consequently, IV monthly cycles of intravenous infusions of 500 mg methylprednisolone for 3 days

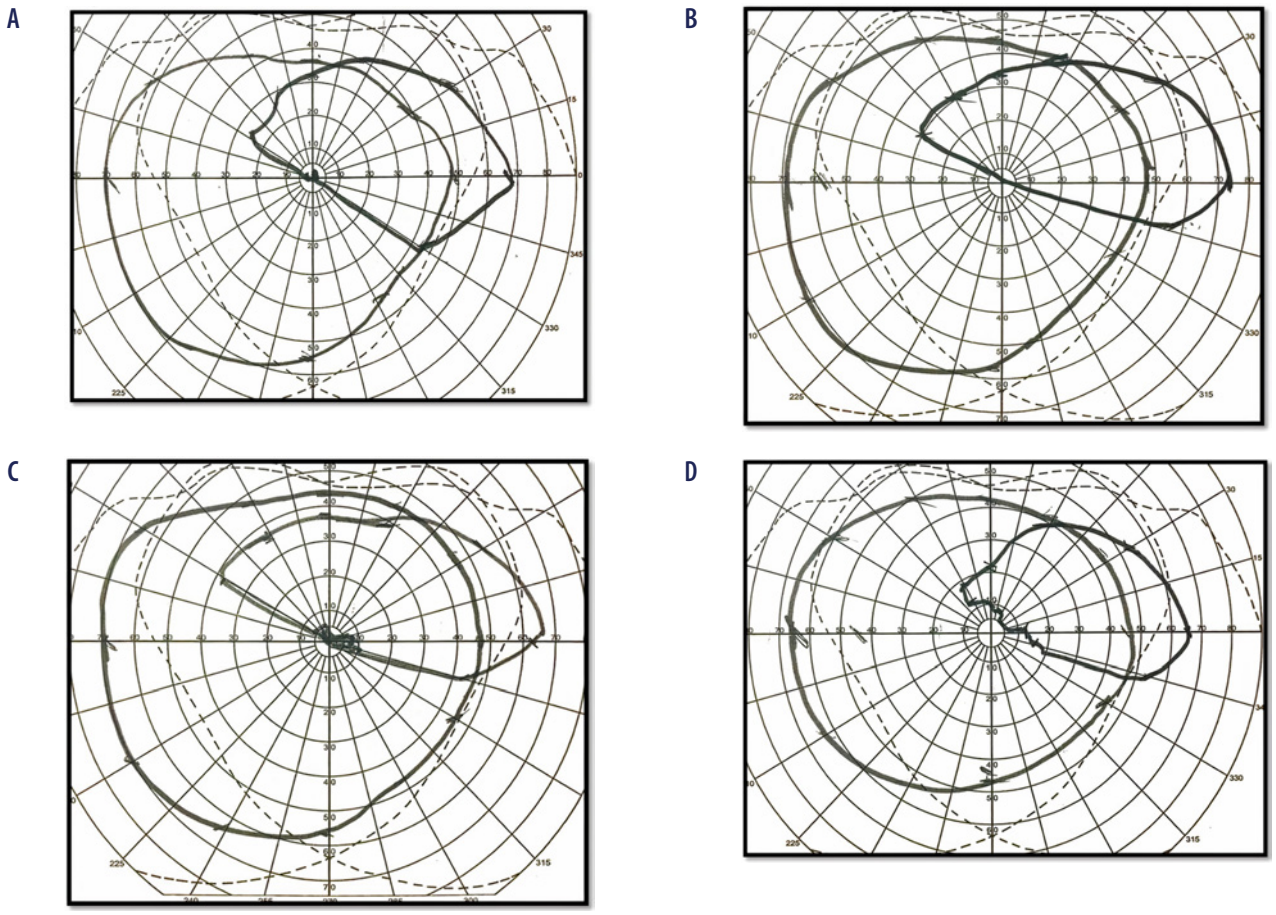

Figure 2. Visual field results in patient with giant cell arthritis (A) at baseline, (B) 1 month after the first cycle of intravenous steroid treatment, and (C) 12 months after the ocular symptoms onset, and (D) 24 months after the ocular symptoms onset

were implemented. Furthermore, oral prednisone in decreasing doses (40 mg – 40 mg – 20 mg – 20 mg) was administered afterwards. Since iatrogenic osteoporosis is a frequent complication of steroid therapy, 1000 IU of cholecalciferol and 1 g of calcium were administered. Additionally, citicoline as neuroprotection for optic nerve was implemented twice a day.

Although the TAB was performed immediately in the emergency the results were obtained with a 2-week delay. It revealed slight intimal thickening without inflammatory infiltrates. In summary, the TAB results did not provide sufficient evidence to diagnose GCA.

The patient was under strict medical supervision for 24 months. During this period, follow-up blood tests showed ESR reduction to 7 mm/h since the completion of the first cycle of intravenous infusions of the steroid. Additionally, in the corresponding time decrease of CRP to < 1.0 mg/l and fibrinogen to 207.3 mg/dl was detected. Visual acuity of the left eye stabilized at 0.04 and OCT revealed decreased papilledema (Figure 1B-D). Kinetic perimetry showed constant visual field restriction in the left eye, while the right eye remained unaffected (Figure 2B-D). Interestingly, CDI performed directly before the second cycle of steroid infusion disclosed reduction of halo thickness (Figure 3B), which confirmed the substantial resolution of inflammatory changes. The patient declared significant relief of the headache and improved subjective quality of life. It is worth highlighting that the fellow eye parameters remained intact during the whole follow-up.

DISCUSSION

The GCA is an ophthalmological sight-threatening emergency that requires urgent treatment. Nowadays, there is a rising probability of facing a patient suffering from GCA due to society's senescent model [2]. What is more, in patients with large-vessel GCA, classic GCA symptoms or those fulfilling ACR diagnostic criteria are less frequent [7]. It may pose difficulties in quick diagnosis required for immediate treatment and inhibition of disease progression. The clinical picture of the reported patient significantly differs from standard GCA cases. A typical GCA patient is an older woman suffering from headaches, vision loss, scalp hypersensitivity, jaw claudication, and systemic symptoms [3]. Contrary to the pathognomonic pattern, the presented patient was an older man with intense, unusual headaches and vision loss. Additionally, the patient presented just slightly elevated ESR. Temporal artery palpation and TAB showed no abnormalities, in contrast to the typically expected results.

According to ACR guidelines the current gold standard for GCA diagnosis is TAB. Nevertheless, it produces up to 60% of false-negative results [8]. In the present case report

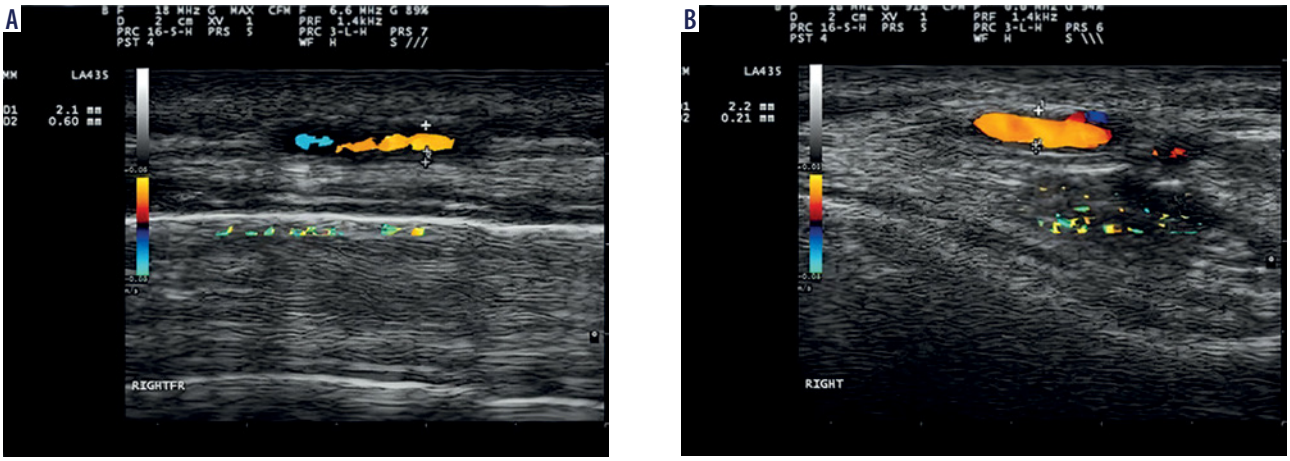

Figure 3. Color doppler imaging results in patient with giant cell arthritis (A) at baseline, and (B) 1 month after the first cycle of intravenous steroid treatment

the TAB findings were not characteristic for GCA. It should be noted that vasculitis is a discontinuous phenomenon, and biopsy material often comprises an unaffected lenght of blood vessel. In this situation GCA could remain undiagnosed. What is more, TAB is an invasive procedure demanding violation of the skin integrity. That could lead to some further complications such as hematoma, scalp necrosis, or infections. Moreover, the classic hematoxylin and eosin staining might not differentiate well the affected tissues in GCA. Although Masson's trichrome staining is recommended for inflammation process in collagen and muscular tissue differentiation it increases the costs of diagnostics and has limited availability. Additionally, TAB results are not immediate, which causes significant diagnostic delay [8].

On the other hand, CDI of the superficial temporal arteries is a quick, low-cost, and non-invasive tool for GCA confirmation. Moreover, it can be easily repeated. According to studies, CDI's sensitivity and specificity may be respectively 69% and 82% compared to 55% and 95% in the case of the TAB [9]. The meta-analysis revealed the presence of unilateral halo sign associated with a sensitivity of 68% and specificity of 91%, rising to a specificity of 100% in the presence of bilateral halos [10]. Interestingly, additional confirmation of the halo sign by the compression sign can increase the sensitivity to 75-79% and the specificity to 100% in GCA diagnosis [11]. Moreover, CDI can be applied as a stand-alone diagnostic tool or assistance to choose an accurate inflamed part of the vessel for biopsy to improve its efficacy. Finally, CDI could be used to monitor the patients' condition in follow-up. After appropriate treatment, the halo sign is reduced after 16 days since therapy induction [12]. This is consistent with our observation where the halo sign was not detected directly before the second cycle of steroid intravenous infusion. Moreover, the absence of the halo sign proves the correct patient's response to treatment and well-diagnosed GCA. The TABUL study showed significantly a smaller halo sign in patients who had received over 4 days of steroid treatment, compared with those receiving up to 4 days of treatment [8, 13]. Nevertheless, the standards for interpreting the CDI results still need to be defined [13]. In the present case, the halo sign was significantly reduced and the CDI was the exact cause of fast identification of GCA, despite negative TAB results.

However, CDI is not free from disadvantages. The examination is mainly dependent on the ultrasonographer's knowledge and experience in conducting the examination or the quality of equipment the physicians work with. Studies suggest that after rereading by an experienced specialist, 7 from 43 negative CDI results turned out to be positive [14]. It might pose a problem with the accessibility of ultrasonographers familiar with GCA images. On the other hand, the halo sign can occasionally be found also in granulomatosis with polyangitis or polyarteritis nodosa and infections with secondary vasculitis [8, 15]. Nevertheless, several advantages over TAB make CDI a valuable tool in GCA emergency diagnostics.

CONCLUSIONS

The presented case report proves that GCA is potentially sight-threatening urgent condition in ophthalmology. Moreover, this disease demands immediate intervention to implement the proper treatment and prevent vision loss in fellow eye. Considering the fact of several adverse effects of steroid treatment the quick, non-invasive, and highly specific tool, allowing rapid GCA differentiation and prompt treatment implementation is needed. It is worth to highlight, that the presented case confirmed an essential role of the CDI in early GCA diagnostics. In comparison with TAB this tool is accurate and easily available in case of ophthalmological emergency. It might be especially beneficial in less symptomatic patients. To sum up, the CDI is a cost-effective tool for both diagnostics and treatment monitoring in GCA.

DISCLOSURE

The authors declare no conflict of interest.

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