CASE REPORT

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Giant cell arteritis with arteritic anterior ischemic optic neuropathy

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Abstract

Giant cell arteritis (GCA) is an inflammatory vasculitis of unknown etiology that mainly involves large and medium arteries, particularly the cranial branches of the aorta. GCA with consecutive arteritic-anterior ischemic optic neuropathy (A-AION) has rarely been diagnosed in Romania. Recently, we encountered an 83-year-old patient who presented with left eye visual impairment and corresponding optic disc diffusely swollen and pale. He also had typical manifestations of GCA, such as malaise, and temporal headache, and a highly elevated erythrocyte sedimentation rate and C-reactive protein level. Biopsy of his left superficial temporal artery revealed a granulomatous inflammation with multinucleated giant cell infiltration, so he was diagnosed with GCA with consecutive left A-AION. Because without treatment, this affection usually progresses very rapidly, the patient was promptly treated with an adequate dosage of steroids, which was essential to save the visual function of both eyes. Our case report confirms the potential of visual recovery after prompt corticosteroid treatment in GCA with eye involvement.

Keywords: giant cell arteritis, granulomatous inflammation, vasculitis, temporal artery biopsy.

Introduction

Giant cell arteritis (GCA) is an inflammatory vasculitis of unknown etiology that mainly involves large and medium arteries, particularly the cranial branches of the aorta. Superficial temporal artery (TA), internal maxillary, and orbital (retrobulbar) arteries [ophthalmic artery (OA), central retinal artery (CRA), and posterior ciliary arteries (PCAs)] are the most frequently affected vessels. Other names for GCA include Horton disease, temporal arteritis, granulomatous arteritis, arteritis cranialis, and arteritis of the aged [1–3].

Age is the most important risk factor for GCA. The disease is rare in patients younger than 50 years. In those 50 years and older, the incidence increases with age, peaking in the eighth decade. The age range in one series of 166 cases proven by temporal artery biopsy (TAB) was 55–92 years. The median age of onset is 75 years [1–3].

The annual incidence in northern European countries has been reported to be more than 20 cases per 100 000 people. A United Kingdom study reported an incidence of 22 per 100 000 [4]. Scandinavian countries have reported the highest incidence. The annual incidence in southern European countries has been reported to be less than 12 cases per 100 000 people. Countries with a lower life expectancy have a lower prevalence [1–3].

Ocular involvement might occur in up to 50–70% of patients with GCA [arteritic-anterior ischemic optic neuropathy (A-AION) or central retinal artery occlusion (CRAO)] and represents a true ophthalmic emergency because the possibility of visual loss is very high if it is not recognized and treated promptly [3].

Aim

The aim of our paper is to investigate the ophthalmologic and systemic features, the ultrasound findings of the orbital vessels and of the superficial temporal and the carotid arteries and the TAB data in a patient with left AION, which helped us to quickly differentiate newly diagnosed arteritic-AION from non-arteritic-AION. This is because A-AION requires immediate glucocorticoid treatment, in order to protect both eyes from going blind.

Case presentation

An 83-year-old man of Greek descent with sudden onset of painless visual decrease, and visual field loss in his left eye for one day was referred to our Department of Ophthalmology on September 27, 2015. He had been suffering malaise and temporal headache for two weeks. He had no medical history of systemic disease. No chest pain or breathing difficulty was noted. On ophthalmologic examination, his best-corrected visual acuity was 15/20 in the right eye and 6/20 in the left eye. The intraocular pressure of his right and left eye was 14 mmHg and 17 mmHg, respectively. Both eyes were freely movable and non-tender upon eye movement. A relative afferent pupillary defect of light reflex was noted in his left eye. He presented a severe altitudinal defect of his left eye during the visual field test. Although the patient did not

complain of visual disturbance in his right eye, the visual field test revealed unspecified right visual field defects, which suggested early concomitant involvement of his right eye (Figure 1).

Slit-lamp examination identified pseudo-phakic intraocular lens (IOL) in both eyes, asteroid hyalosis and peripapillary atrophy (post-glaucoma attack) in the right eye and a pale swelling of the left optic disc (Figure 2).

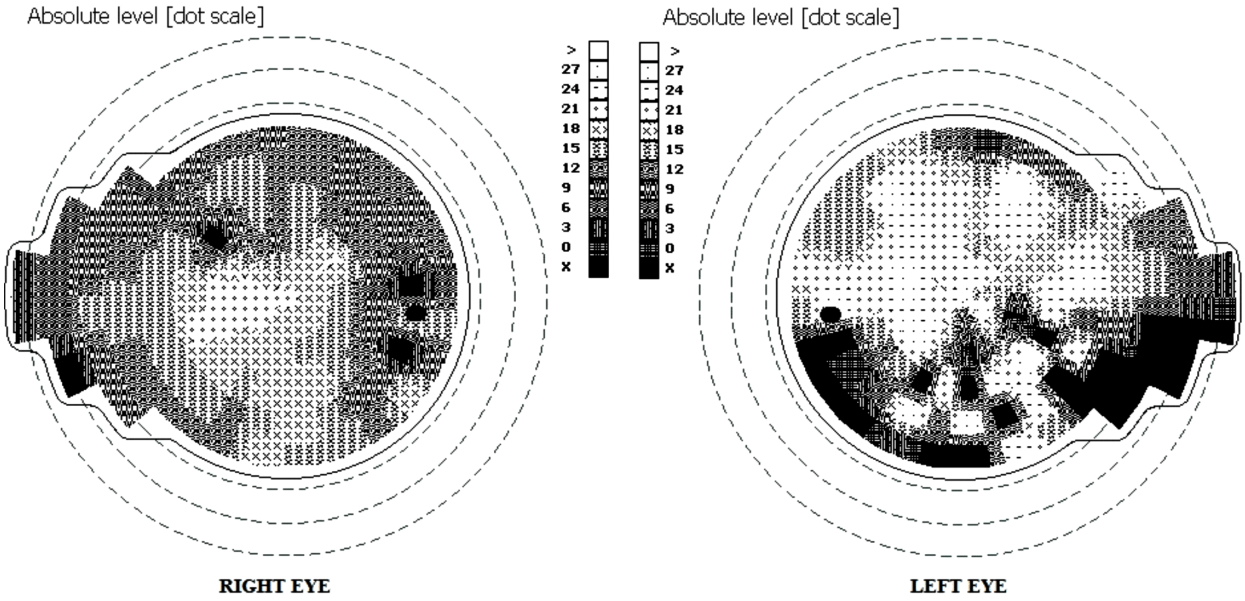

Figure 1 – ***An automatic visual field test showed unspecified visual field defects of the right eye and severe inferior altitudinal scotoma of the left eye.***

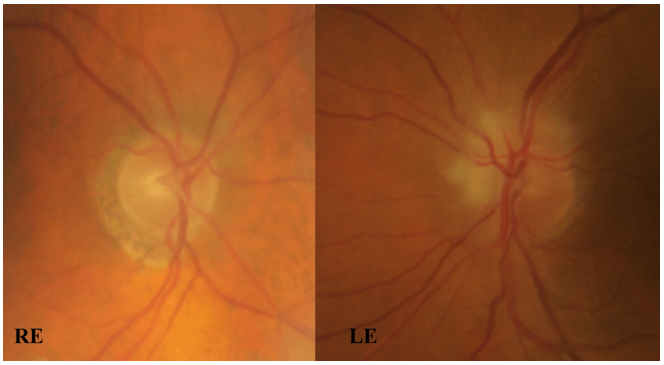

Figure 2 – ***Fundus color photography identified peripapillary atrophy of the right optic disc and marked pale swelling of the left optic disc. RE: Right eye; LE: Left eye.***

A mild color disturbance was noted on an Ishihara color plate test (6/15 errors, normal ≤4/15). Optical coherence tomography (OCT) of the optic nerve head revealed retinal nerve fibers layer (RNFL) swelling in the left eye (Figure 3).

Color Doppler imaging (CDI) of orbital vessels (OA, CRA, PCAs) was done at presentation in our Department. We noted an increased resistance index (RI) in all retrobulbar vessels, including both PCAs (both nasals and temporals).

Extracranial Duplex sonography (EDS) identified a typical sonographic feature for temporal arteritis as part of GCA: a "dark halo" sign at the level of a portion of the temporal ramus of the left TA. All other vessels, including the left internal carotid artery did not present any signs of stenoses or occlusions.

Head computed tomography excluded stroke.

Laboratory investigations showed an erythrocyte sedimentation rate (ESR) of 93 mm/h (normal value at his age <20 mm/h) and a C-reactive protein (CRP) level of 2.3 mg/dL (normal value <0.5 mg/dL). Hemoglobin was 13.2 g/dL and the blood cell profile was within normal limits. Renal and liver function tests, immunological factors such as antinuclear antibody were all normal. A biopsy of the left superficial TA of 3 cm section was performed. It was guided by the EDS of the TAs.

The histopathological examination revealed accumulation of histiocytes, epithelioid cells, and giant cells at the intima–media junction, followed by fragmentation, degeneration, and dissolution of the internal elastic lamina (Figure 4).

He was promptly diagnosed with left acute A-AION due to GCA, and was then immediately treated with 1 mg/kg/day oral Prednisone. The patient's symptoms (malaise, temporal headache) improved rapidly, ESR decreased to 18 mm/h and CRP level decreased to <0.1 mg/dL within four weeks of treatment. The visual acuity improved after treatment and recovered completely one month later, from 6/20 in the left eye to 6/6. Fundus color photography at left eye, after one month noted resolution of the optic disc edema (Figure 5).

The patient was treated with oral Prednisone on a careful tapering schedule, with monthly monitoring of ESR and CRP level for over five months. The patient presented stationary ophthalmologic evolution at five months, without classic clinical symptoms (temporal headache) of GCA and without any systemic manifestations (fever, malaise).

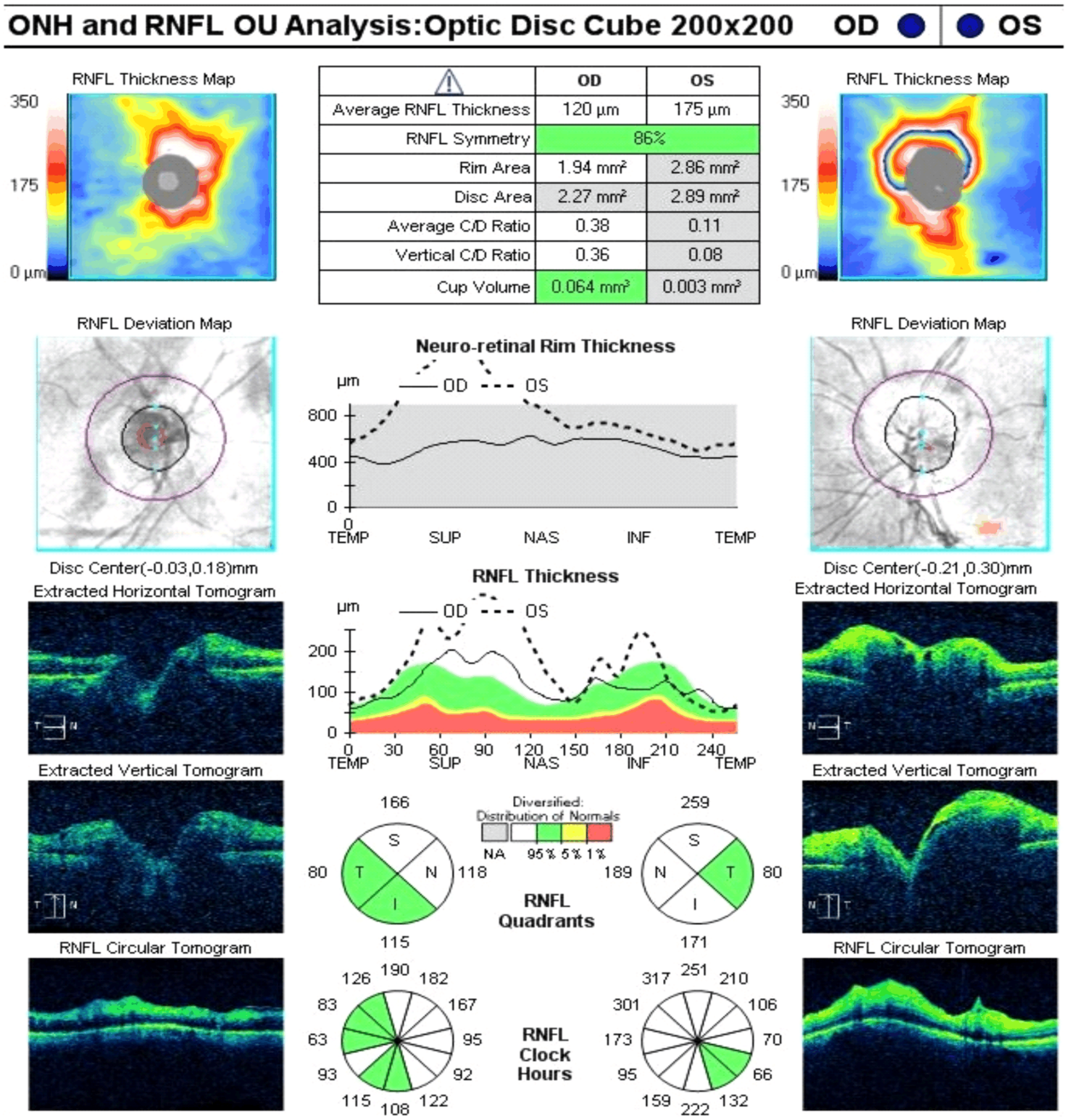

Figure 3 – ***Optical coherence tomography of the optic nerve head (ONH) revealed retinal nerve fibers layer (RNFL) swelling in the left eye. OU: Both eyes (oculi uterque); OD: Right eye (oculus dexter); OS: Left eye (oculus sinister); C/D: Cup-to-disc; TEMP (T): Temporal; SUP (S): Superior; NAS (N): Nasal; INF (I): Inferior.***

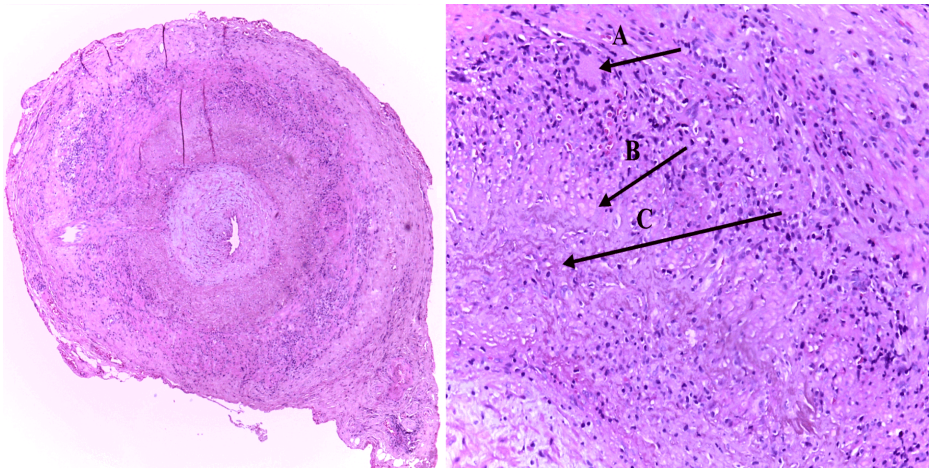

Figure 4 – ***The histopathological examination of the left superficial temporal artery biopsy (TAB) noted thickened vascular wall with inflammatory infiltration of multinucleated giant cells (A), epithelioid cells (B) and dissolution of the internal elastic lamina (C). HE staining: left, ×40; right, ×100.***

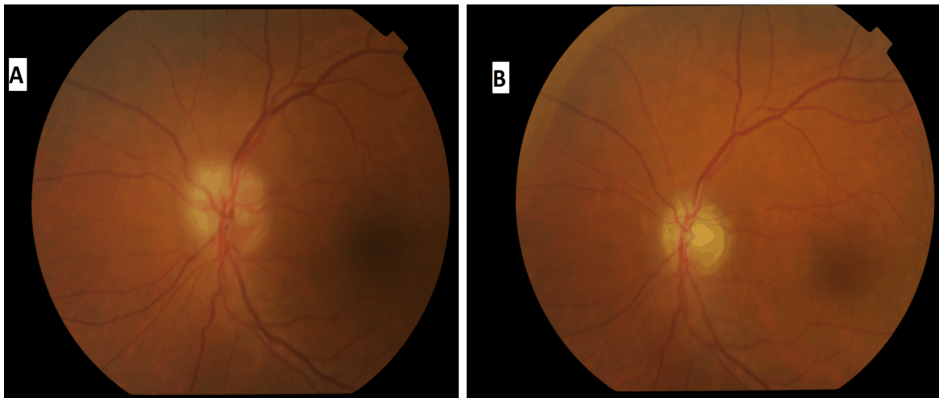

Figure 5 – ***Fundus color photography at left eye, on presentation (A) and after one month of treatment (B) showed resolution of the optic disc edema.***

Discussion

AIONs represent a segmental infarction of the optic nerve head supplied by the PCAs: nasals and temporals [5, 6]. There are two types of AIONs: arteritic-AION, and non-arteritic-AION, which is a multifactorial disease [5, 6]. GCA represents the most common cause of A-AION, although in rare cases, other vasculitides may cause it [5].

The clinical presentation of arteritic-AION is similar to that of non-arteritic-AION, but several aspects should raise clinical suspicion for A-AION [5].

Usually, the degree of visual loss is often more severe in arteritic-AION than in non-arteritic-AION. In one study [5], 54% of patients with A-AION due to GCA had initial visual acuity ranging from counting fingers to no light perception, as compared to 26% in the non-arteritic-AION group, and only light or no light perception in 29% and 4%, respectively. This result shows that sudden, painless, severe permanent loss of vision is extremely suggestive of A-AION [5, 6]. However, on ophthalmologic examination, the best corrected visual acuity of our patient was 6/20 in the left clinical affected eye, suggesting non-arteritic-AION.

Ophthalmoscopy indicated in his case a pale swelling of the left optic disc, in concordance with different studies that asserted that optic disc edema is associated with pallor (a chalky white color) in arteritic-AION patients, and more frequently with hyperemia in non-arteritic-AION cases; pallor is delayed in non-arteritic-AION [5–8].

The finding of associated retinal or choroidal ischemia in addition to AION is highly suggestive of GCA [5–13].

According to different studies [9–13], severe diminished blood flow velocities in the PCAs, especially on the affected side, and high RI in all retrobulbar vessels, in both orbits, like in our patient's case, represent characteristic signs of the CDI of the orbital vessels in A-AION. In non-arteritic-AION, blood velocities and RI in PCAs are relatively preserved [9–13].

Ultrasonography of the TAs in temporal arteritis is very important for GCA diagnosis [11–13]. Schmidt *et al.* demonstrated that the most specific (almost 100% Sp) and sensitive (73% Se) sign for GCA is a concentric hypoechogenic mural thickening "halo", which was interpreted as vessel wall edema [13]. Other positive findings for GCA are the presence of occlusions and stenoses at the level of different branches of external carotid artery, including TAs [11–13]. We detected a "dark halo" sign at the level of an inflamed portion of the temporal ramus of the left TA of our patient. Schmidt *et al.* compared the results of TAs ultrasound examinations with the occurrence of visual ischemic complications in 222 consecutive patients with newly diagnosed, active GCA. However, findings of TAs ultrasonography did not correlate with eye complications [11–13].

For this reason, just as in our case, additional ultrasound investigation of orbital vessels enabled prompt identification of A-AION caused by vasculitis from GCA [11–13].

According to the criteria of the *American College of Rheumatology* [14], GCA should be considered in all patients with A-AION who are older than 50 years of age, with new headache in the temporal area, TAs tenderness, and/or reduced pulse, jaw claudication, systemic symptoms (malaise, fever, etc.), ESR exceeding 50 mm/h, and typical histological findings (granulomatous involvement) in TAB.

Systemic symptoms of GCA may precede visual loss by weeks or months, like in our patient's case. However, about 25% of patients with biopsy-confirmed GCA present with isolated A-AION without any systemic symptoms (so-called occult GCA) [1–3].

ESR and CRP high levels (like in our patient's case) are considered the most highly predictive laboratory data of GCA, with a combined sensitivity of up to 99.2% [8, 15]. Normal values of these data in the context of low clinical suspicion are enough to safely rule out GCA and TAB is not necessary in these cases [5, 8, 15]. These laboratory data are also very useful for monitoring GCA activity and regulating steroid therapy (like in our patient's case) [5, 6, 8]. Most neuro-ophthalmologists prescribe high-dose intravenous Methylprednisolone to treat patients with acute A-AION, like in our case [5].

TAB represents the gold standard for diagnosis of GCA [1–3, 5–8, 16]. To establish the diagnosis and prevent treatment delay of this potentially blinding disease, TAB should be performed as soon as possible in every patient suspected of having GCA, or A-AION [1–3, 5–8, 16]. Because of segmental involvement of TAs in GCA, the TAB has to be guided in all cases with clinical suspicion of GCA by EDS of TAs (in our case the portion with "dark halo" sign of the temporal ramus of the left TA) and by typical TAs clinical signs (tender, swollen portions of TAs) [11–13]. To reduce the chance of false negative results caused by skip lesions, it is recommended that, in TAB, at least a 2.5 cm section (3 cm in our case) is obtained, together with more closely spaced sections (0.25 mm or 0.5 mm) for pathological examination [3].

In all cases with A-AION due to GCA the histopathological picture is represented by a granulomatous inflammation of the media layer (chronic inflammatory infiltrate with giant cells) with characteristic fragmentation of the internal limiting lamina and intimal thickening (like in our case) [1–3, 7, 8, 14].

Although the visual prognosis is very poor in acute unilateral A-AION, a study revealed some recovery of vision in one third of cases [3] and final visual acuity remained unchanged in most of the patients in other study [16]. The result of our case report confirms the potential of visual recovery after prompt corticosteroid treatment.

Conclusions

Our report presented a rare case of GCA with unilateral A-AION, were a rapid diagnostic work-up, and a prompt treatment with an adequate dosage of steroids led to a complete recovery of the visual function of both eyes.

Conflict of interests

The authors declare that they have no conflict of interests.

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