Arteritic Anterior Ischemic Optic Neuropathy in the Course of Giant Cell Arteritis After COVID-19

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Patient: Female, 69-year-old
Final Diagnosis: Anterior ischemic neuropathy • giant cell arteritis
Symptoms: Headache • tenderness • vision loss
Medication: —
Clinical Procedure: —
Specialty: Ophthalmology

Objective: Rare disease
Background: Giant cell arteritis (GCA) is an inflammation of large vessels that affects the lining of the arteries and leads to vessel swelling and the eventual reduction of blood flow. This can result in ischemia of the optic nerve, which is known as arteritic anterior ischemic optic neuropathy (AAION). The present case seems noteworthy because the patient developed GCA with the ocular manifestation of AAION shortly after having COVID-19.

Case Report: A 69-year-old woman was admitted to the Clinic of Ophthalmology after having COVID-19. She reported vision loss in the left eye, which appeared 2.5 weeks after a positive SARS-CoV-2 test. While in the hospital, she was diagnosed with AAION and GCA. The patient was treated with enoxaparin sodium, prednisone, and methotrexate. Three months after the hospitalization, the visual acuity of the left eye was limited to light perception, and optic nerve atrophy was reported.

Conclusions: We would like to emphasize the role of SARS-CoV-2 infection as a possible risk factor for the onset of GCA and its ocular manifestations, such as AAION. However, further research is needed to determine the relationship between SARS-CoV-2 infection and GCA. Because some symptoms of the 2 diseases are similar, the diagnosing process might be long and challenging. The diagnosis of GCA should be made as soon as possible to avoid serious complications, such as bilateral vision loss.

Keywords: COVID-19 • Giant Cell Arteritis • Optic Neuropathy, Ischemic • Vision Disorders

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Giant cell arteritis (GCA) is an inflammation of the large vessels. Most commonly, it affects the temporal arteries; hence, it is also called temporal arteritis. The exact causes of the disease have not yet been discovered; however, an autoimmune and infectious background is considered relevant. Patients with GCA develop an inflammatory reaction which affects the lining of the arteries, leading to vessel swelling and the eventual reduction of blood flow [1]. This can result in ischemia of the optic nerve, a condition known as arteritic anterior ischemic optic neuropathy (AAION), which accounts for 5% to 10% of anterior ischemic optic neuropathies (AION). The disease results in unilateral or bilateral blindness if left untreated. In about 20% of patients with AAION, there are no other symptoms of GCA, and the ocular manifestations might be the only symptoms present [2].

The case we are presenting seems noteworthy because the patient developed GCA with the ocular manifestations of AAION shortly after having COVID-19. It is not unlikely that SARS-CoV-2 infection may have triggered the onset of GCA.

Case Report

A 69-year-old woman was admitted to the Clinic of Ophthalmology at the beginning of January 2021 after having COVID-19, which started at the beginning of December 2020. While having the SARS-CoV-2 infection, which was confirmed by a RT-PCR test (performed in an authorized laboratory in Wrocław, Poland), the patient had fever, fatigue, and cough. The patient did not receive any treatment before hospitalization. She reported vision loss in the left eye (OS), which initially covered only the superior half of the visual field (described by the patient as a falling curtain) and proceeded into complete vision loss. The vision loss appeared 2.5 weeks after the positive SARS-CoV-2 test. Additionally, the patient had severe headaches, which were near the eyeballs and the occiput, and scalp tenderness. Unfortunately, the patient did not remember the exact time when the symptoms appeared. She claimed that all of them began around 2.5 weeks after the onset of COVID-19. On admission, the patient provided documentation concerning arterial hypertension and type II diabetes. Both conditions were controlled with appropriate oral treatment. Her glycosylated hemoglobin levels were unremarkable, and her blood pressure was within normal limits.

The patient’s ocular examination on admission showed the following: The visual acuity of the right eye (OD) was 0.9 cc, and in the OS, the patient had light perception from the nasal and superior side. The intraocular pressure was 10 mm Hg in the OD and 9 mm Hg in the OS. The pupil of the OD showed no abnormalities. The pupil of the OS was wider than that of the OD and showed no direct response and a slow indirect response to light. In both eyes, cortical cataracts were present. The dilated fundus examination of the OD revealed cotton wool spots near the major superior artery and vein (Figure 1). In the OS, the blurring of the optic margins with flame hemorrhages

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**Figure 1.** Fundus of right eye: cotton wool spots in the area of watershed zones seen in fluorescein angiography.

**Figure 2.** Fundus of left eye: blurring of the optic margins with flame hemorrhages.
were found (Figures 2, 3). Fluorescein angiography indicated
the following abnormalities: delayed choroidal filling (Figure 4)
and watershed zones (Figures 5, 6). The result of the C-reactive
protein test was unremarkable, and the erythrocyte sedimenta-
tion rate (ESR) was elevated (63 mm/h; range, 3-15 mm/h).
The results of the complete blood count (including platelet
count), renal and liver function, activated partial thromboplas-
tin time, prothrombin time, and antithrombin tests were unre-
markable. The result of the D-dimer test was only slightly ele-
vated (0.511 ug/mL; range, <0.5 ug/mL). The patient was also
tested for the presence of anti-neutrophil cytoplasmic antibod-
ies, anti-cyclic citrullinated peptide antibodies, and rheuma-
toid factor. The results of these tests were unremarkable. The
electrocardiogram was within the normal range.

Additionally, the patient reported some typical symptoms of
GCA on admission, including tenderness of the scalp and head-
aches near the occiput. The patient’s overall image suggest-
ed the onset of GCA; therefore, an ultrasound of the tempo-
al arteries was performed. The result revealed the signs of
an inflammation: wall thickening and a “halo”. The patient
also underwent ultrasound of the retrobulbar arteries, which
suggested thrombosis in the ciliary arteries, as no flow was
detected in them. Flow was detected in the ophthalmic ar-
tery and in the central retinal artery. The American College
of Rheumatology does not require biopsy for the diagnosis
of GCA. Our patient satisfied the required minimum of 3 out
of 5 criteria with the following 4 satisfied criteria: patient age
>50 years, new-onset headache, temporal artery abnormality, and elevated ESR > 50 mm/h [3]. We therefore diagnosed the patient with GCA and AAION.

The patient was treated with enoxaparin sodium 0.4 mL/day for 10 days and prednisone 80 mg/day for 4 days. At the end of the hospital stay, she was prescribed methotrexate 15 mg/week and prednisone, with a reduction of doses until reaching 60 mg.

At the follow-up visit at the end March 2021, the visual acuity of the OD was 0.8 cc, and there was light perception in OS. The intraocular pressure was 12 mm Hg in the OD and 10 mm Hg in the OS. Optical coherence tomography demonstrated macular traction in both eyes, and retinal nerve fiber layer analysis of the OS revealed optic nerve atrophy.

After treatment, the condition of the OS showed no improvement. Fortunately, the OD was not affected by the disease. The visual acuity of the OS was limited to light perception, and optical coherence tomography and retinal nerve fiber layer analysis revealed optic nerve atrophy.

**Discussion**

In our case, the clinical presentation (scalp tenderness, headaches), laboratory tests (elevated ESD) and imaging exam ("halo" and thickening of the wall in the ultrasound of the temporal arteries) suggested AAION caused by GCA. The comorbidities of the patient included diabetes type II and hypertension, which were both controlled with appropriate oral treatment. As the overall image pointed to the inflammatory character of the disease, and the patient’s past medical history had not suggested any other pathological background, we did not conduct additional tests for thrombophilia or arteriosclerosis.

Reports on the coexistence of GCA and COVID-19 seem scarce. Lecler et al observed an increased incidence of GCA by 70% during the pandemic, compared with 2019 [4]. Luther et al stated that while 28 patients were diagnosed with GCA in 2019, between April 2020 and June 2020, the number of new GCA cases was 24, suggesting that viral etiopathogenesis of GCA might exist. Higher rates of ocular manifestations of GCA were observed during the pandemic; however, none of the patients diagnosed with GCA had any symptoms suggesting COVID-19 [5]. Mulhearn et al observed the excess of 33 cases of GCA in 2020 in comparison with 2019, which they reported might have been caused by the damage to the endothelium and activation of T-helper cell type 1 cellular immunity and the monocyte-macrophage system [6]. Monti et al, on the other hand, noticed fewer cases of GCA during the past year, which might have been caused by the reluctance of patients to visit hospitals and more complicated access to medical care during the pandemic [7]. There are some reports concerning non-arteritic anterior ischemic optic neuropathy (NAION) during or after a course of COVID-19. Rho et al described a case of a 43-year-old man who developed NAION in the setting of COVID-19 [8]. Garcia Briones et al presented a case of a 55-year-old man who developed simultaneous NAION and concluded that COVID-19 significantly contributed to the disease [9]. However, AAION during the course of COVID-19 has not yet been described.

Unfortunately, we failed to find pathogenetic similarities between COVID-19 and AAION. However, we did find more references relevant to the topic of our case report. Riera-Marti et al described a case of a 50-year-old man who had had GCA with spontaneous resolution, most likely triggered by SARS-CoV-2, as the virus was claimed to have affinity for vascular endothelium [10]. According to Ostrowski et al, the hypothesis that the pathogenesis of GCA might include viral infection (VZV infection) should be treated with caution but not dismissed [11]. Therefore, it does not seem unlikely that other viruses may also trigger the disease. Xu et al suggested a link between SARS-CoV-2 infection and Kawasaki disease in children [12]. Kawasaki disease, just like GCA, is a type of vasculitis. Hanafi et al reported a case of a central nervous system-vasculitis-like pattern as a complication of SARS-CoV-2 infection and hypothesized about the presence of endothelitis in the course of the viral infection [13].

What also needs to be emphasized is that some symptoms of COVID-19 and GCA overlap, including headache, fatigue, elevated inflammatory markers, and fever, which can make diagnosing GCA challenging [14]. In both conditions, the vascular endothelium is affected, which can lead to thrombosis of the blood vessels [10,15]. Having analyzed our case, we suspect that COVID-19 might have triggered the sudden onset of GCA and the following AAION.

**Conclusions**

We would like to emphasize the role of SARS-CoV-2 infection as a possible risk factor for the onset of GCA and its ocular manifestation, such as AAION. However, further research is needed to determine the relation between COVID-19 and GCA. Because a few symptoms of the 2 diseases are similar, the diagnosing process might be long and challenging. The diagnosis of GCA should be made as soon as possible to avoid serious complications of the disease, such as bilateral vision loss.

**Declaration of Figures’ Authenticity**

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.
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