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INVITED REVIEW

# Ophthalmologic Assessment in Giant Cell Arteritis

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Note:
This manuscript was reviewed by Özlem Dikmetaş.

**Introduction**

Ocular examination starts with the measurement of visual acuity. In a patient with decreased vision, color vision, light reaction, and relative afferent pupillary defect should also be assessed. After evaluating ocular motility, especially in cases complaining of diplopia, a slit lamp examination should be done to determine anterior segment structures. Intraocular pressures are measured afterward. If the preliminary diagnosis necessitates a visual field exam, it should be done before pupillary dilatation for funduscopy. Possible funduscopic findings in the acute stage of giant cell arteritis (GCA) include pallid edema of the optic nerve head and signs of retinal ischemia, including cotton wool spots (Figure 1) [1]. Besides visual field testing, angiography and optical coherence tomography (OCT) is also employed in diagnosing and monitoring patients with GCA-associated ocular involvement.

**Ancillary Ocular Examination**

*Visual field testing*

In patients with GCA, evaluating the visual field is as important as measuring the central visual acuity. Visual field defects develop in GCA due to retinal, optic nerve, or cortical pathologies. Visual field exams can be done with static automated perimetry or, less commonly, with kinetic (dynamic) perimetry, which is mostly manual. In nonarteritic anterior ischemic optic neuropathy (NA-AION) patients, absolute inferior nasal defect (22%) and relative inferior altitudinal defects are more frequently detected with kinetic perimetry. If only static automated perimetry is available, patterns including central and peripheral points (e.g., FF 120, 165) should be employed to evaluate the periphery. Central scotomas are also a feature of AION. The visual field defect is usually the same type but more extensive and pronounced in arteritic-AION than NA-AION (Figure 2). In posterior ischemic optic neuropathy, central visual field defects are more frequent [2].

*Fundus fluorescein angiography (FFA)*

In this imaging modality, sodium fluorescein is given intravenously from an antecubital vein. Its passage into the choroidal circulation and then into the retinal circulation can be monitored by a fundus camera with special filters [3]. Suspected GCA may be evaluated with FFA, which can demonstrate delay of perfusion and hypofluorescence of either choroid, retina, or both (Figure 3) [1]. A large area of choroidal hypoperfusion is highly suggestive of GCA. Prolonged arm-to-choroid and choroidal filling times are observed in patients with A-AION, GCA's most common ocular finding [4]. The affected PCA will have peripapillary choroidal hypofluorescence on the same side. Depending on the degree of involvement, the optic disc may be stained in the late phases [5]. In NA-AION, early stages of FFA disclose filling defect or delay, usually at the prelaminar and peripapillary region and the watershed zone (the area between the medial and lateral PCA) [2].

In cases with central retinal artery occlusion, FFA is important to rule out GCA. Choroidal non-filling, in addition to the delayed retinal filling, will be a sign of accompanying PCA occlusion. In the case of cilioretinal artery (CLRA) occlusion, which may be together with A-AION, hypoperfusion is seen in the area supplied by the CLRA extending to the macula.

In GCA, FFA is an extremely important imaging modality as it shows choroidal and retinal hypoperfusion, especially in the acute phase. When collaterals develop over time, the delay in choroidal filling may not be detected [3].

*Indocyanine green angiography (ICGA)*

Indocyanine green angiography is a less commonly employed imaging modality in daily practice, as the dye is not widely available. The fundus camera should have specific filters according to the absorption and emission wavelengths of the dye. It shows choroidal circulation better than FFA as the molecule is highly protein-bound and does not leak from fenestrated choriocapillaris. In GCA, ICGA discloses severe ischemia of the choroid and staining of several peripheral choroidal vessels [3].

*Optical Coherence Tomography (OCT)*

Optical Coherence Tomography (OCT) is a non-invasive, non-contact imaging system providing high-resolution cross-sectional images of the posterior segment of the eye. It is analogous to B scan ultrasonography but uses light instead of sound.

In patients with optic neuropathies, OCT shows retinal nerve fiber layer (RNFL) edema in the acute phase [6]. With the development of optic atrophy, RNFL gets thinner (Figure 4). With OCT, it is possible to measure peripapillary RNFL thickness.

Cotton wool spots are focal hyperreflectivity in inner retinal layers (Figure 5). Deep capillary ischemia is seen as paracentral acute middle maculopathy, characterized by a hyperreflective band-like lesion in the inner nuclear layer and outer plexiform layer on OCT.

In patients with central retinal artery occlusion, OCT-A shows increased retinal thickness and reflectivity of the inner retinal layers at the acute stage. At later stages, retinal thinning becomes evident.

*Optical coherence tomographic angiography (OCT-A)*

OCT-A is a new, non-invasive imaging method that allows visualizing blood flow in the retina and choroid without employing a dye. With OCT-A, it is possible to evaluate the retina's superficial and deep capillary plexi separately. Peripapillary capillary plexus can also be examined (Figure 6). In cases with AION, the flow deficits in retinal capillaries were compatible with visual fields [7].

## Conclusion

The ocular examination is essential in the timely diagnosis of ocular complications of GCA. Visual field testing is critical in the diagnosis of optic neuropathy. To assess ocular ischemia, fluorescein and/or indocyanine green angiography should be employed. OCT is used for detecting retinal and peripapillary changes. The role of the newest technique, OCT-A, in monitoring is currently being studied.

## Figures

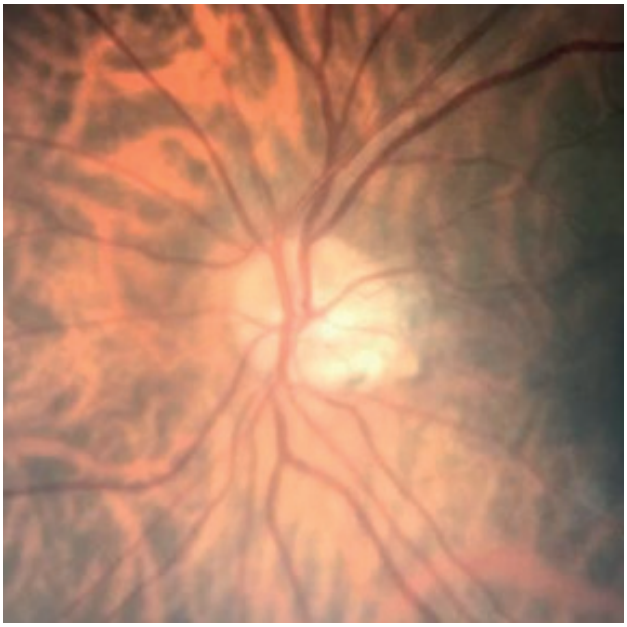

**Figure 1.** Left optic disc of a patient with A-AION due to GCA

**Figure 2.** Visual Field of the eye in Figure 1: Inferior altitudinal defect

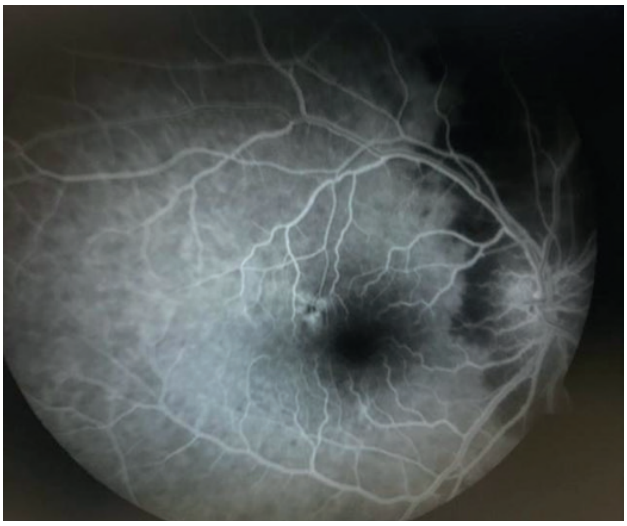

**Figure 3.** FFA: Peripapillary choroidal filling defect more marked superiorly in a case with A-AION

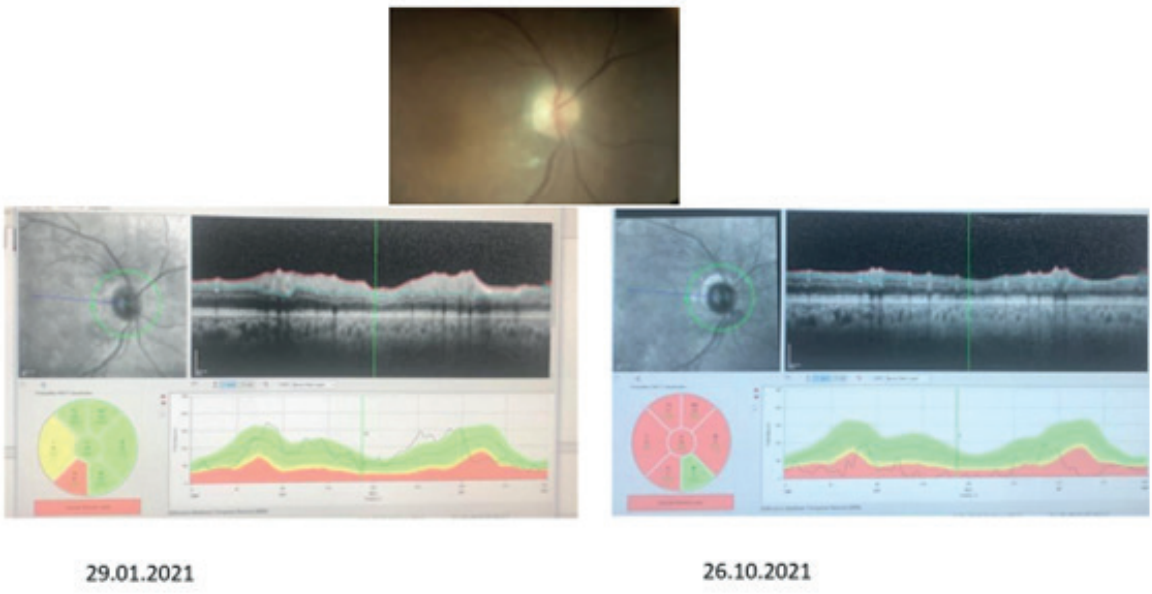


**Figure 4.** OCT: Development of optic atrophy and thinning of peripapillary nerve fiber layer in 9 months in a case with A-AION secondary to GCA

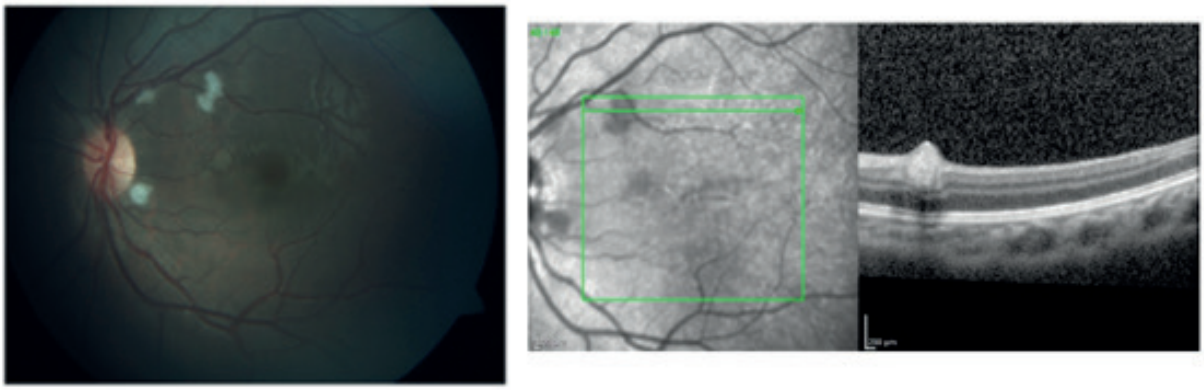

**Figure 5.** OCT: Increased reflectivity in inner layers of the retina corresponding to the cotton wool spots

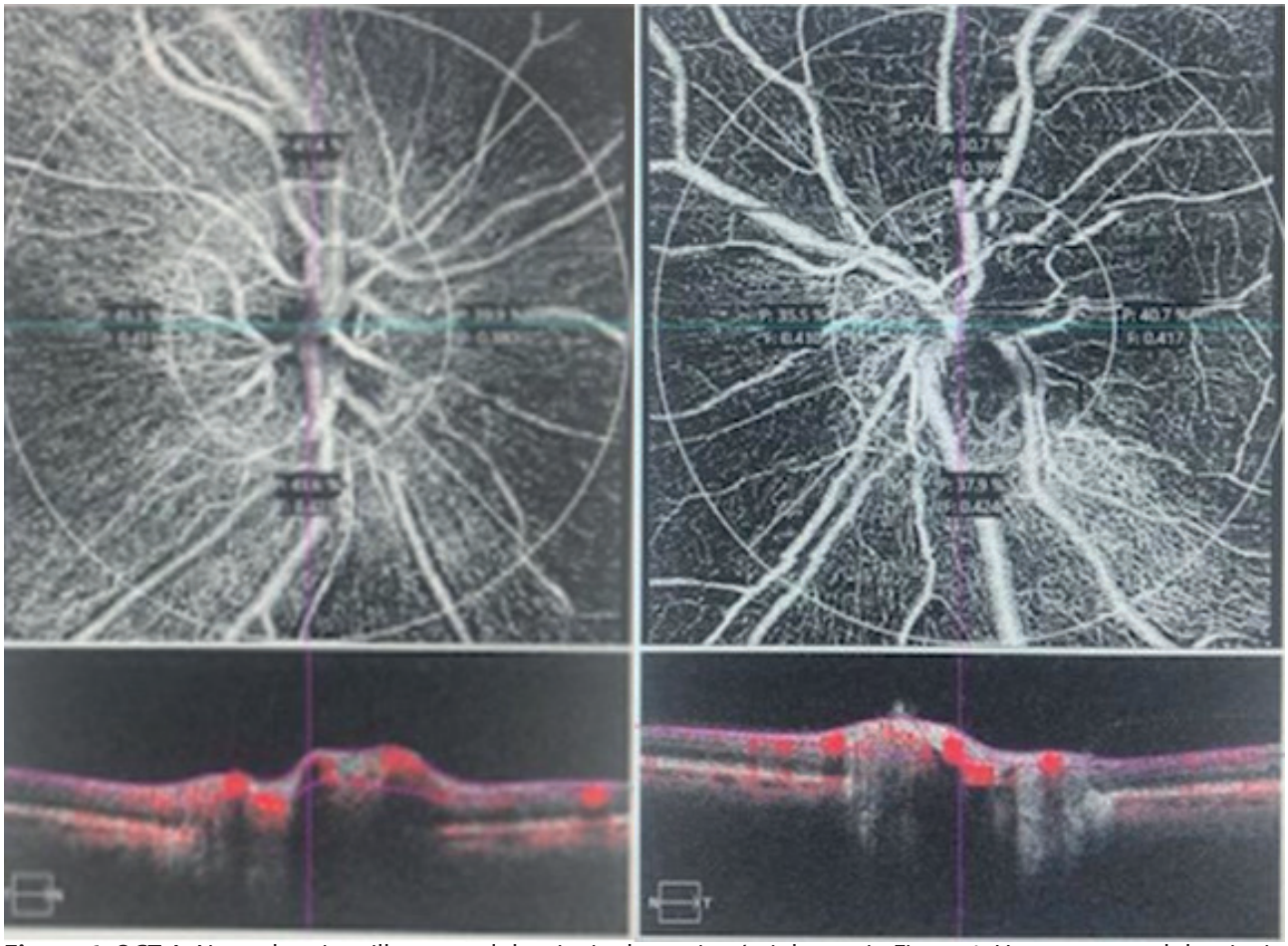

**Figure 6.** OCT-A: Normal peripapillary vessel density in the patient's right eye in Figure 1. However, vessel density in the left eye decreases, which has ischemic optic neuropathy

## REFERENCES


[1] Vodopivec I, Rizzo III JF. Ophthalmic manifestations of giant cell arteritis. Rheumatology. 2018;57(suppl_2):ii63-ii72.

[2] Hayreh SS. Ischemic optic neuropathy. Prog Retin Eye Res. 2009;28(1):34-62.

[3] Patil P, Karia N, Jain S, et al. Giant cell arteritis: a review. Eye Brain. 2013;5:23-33.

[4] Siatkowski RM, Gass JDM, Glaser JS, et al. Fluorescein angiography in diagnosing giant cell arteritis. Am J Ophthalmol. 1993;115(1):57-63.

[5] Hayreh SS. Anterior ischaemic optic neuropathy. II. Fundus on ophthalmoscopy and fluorescein angiography. Br J Ophthalmol. 1974;58(12):964-980.

[6] Iorga RE, Moraru A, Ozturk MR, Costin D. The role of Optical Coherence Tomography in optic neuropathies. Rom J Ophthalmol. 2018;62(1):3-14.

[7] Gaier ED, Gilbert AL, Cestari DM, et al. Optical coherence tomographic angiography identifies peripapillary microvascular dilation and focal non-perfusion in giant cell arteritis. Br J Ophthalmol. 2018;102(8):1141-1146.