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Original Article

Macular star formation in diabetic patients with non-arteritic anterior ischemic optic neuropathy (NA-AION)

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Abstract

Background: NA-AION is a condition that exhibits a number of unique characteristics in diabetics compared with the rest of the population. In some diabetic patients with NA-AION, lipid deposits can be observed around the macula forming an incomplete macular star.
Methods: We describe 12 case studies of patients with NA-AION observing the development of lipid deposits around the macula forming an incomplete macular star.
Results: All our patients developed some level of lipid deposits around the macula in the form of a macular hemistar in the course of their illness.
Conclusion: Some authors have suggested that the macular star is formed by transudation from capillaries deep in the optic disk through the intermediary tissue of Kuhnt, which is located between the retina and the anterior portion of the lamina retinalis. However, the development of the macular star is currently understood not as a simple transudation but as a multifactorial process involving the presence of vascular damage around the optic disk, which is considered one of the most important factors leading to its occurrence.
Although some studies mention the presence of a macular star in patients with NA-AION, we believe that this phenomenon may be significantly more common than the current literature suggests.

Keywords: Non-arteritic anterior ischemic optic neuropathy, Lipid deposits, Macular hemistar, Diabetes mellitus, Neuroretinitis, Macular edema

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Introduction

One of the risk factors for the development of non-arteritic anterior ischemic optic neuropathy (NA-AION) is the presence of diabetes mellitus (DM).[1–3]

NA-AION does not appear to have a worse prognosis in diabetic patients compared with non-diabetics in terms of visual acuity (VA) and visual field impairment at 6 months after onset.[1]

However, NA-AION does exhibit some peculiar characteristics in diabetic patients:

- involvement or recurrence of NA-AION in the contralateral eye is more common in diabetics.[1]
- clinical detection of incipient NA-AION (i.e., in the asymptomatic phase for the patient) is more common in diabetics.[1]
- in diabetics, optic disk edema takes longer to resolve than in non-diabetics.[1]
- optic disk edema in diabetics with NA-AION is characteristically associated with the presence of prominent telangiectasias and more abundant retinal hemorrhages compared with non-diabetics.[1]

All of these features lead us to the conclusion that NA-AION is a condition that manifests unique characteristics in diabetics relative to the rest of the population.

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In some diabetic patients with NA-AION, lipid deposits can be observed around the macula forming an incomplete macular star. Usually, these exudates appear when the optic disk swelling begins to resolve, and they are not typically present during the period of acute VA loss.[4–6] Although some studies mention the presence of a macular star in patients with NA-AION, we believe that this phenomenon may be more common than has been previously reported in the literature.

We present a series of 12 clinical case studies of patients with NA-AION in whom the presence of an incomplete macular star was detected.

Clinical case studies

We present 12 patients with NA-AION who developed lipid deposits around the macula in the form of a macular hemistar over the course of their illness. Table 1 describes the following clinical characteristics for all patients: age, gender, the presence of unilateral or bilateral NA-AION, the existence of vascular risk factors, such as diabetes mellitus or hypertension, VA in the acute phase and after patient follow-up, color vision, visual field testing results, the onset of macular star formation, the presence of diabetic retinopathy, and cranial and orbital MRI findings.

Mean age of patients in our series was 55 years, with an age range of 43–73 years. 5 patients out of 12 presented with bilateral NA-NAION, calling attention to the fact that in two cases the clinical presentation was simultaneous in both eyes.

It was striking to find cases of bilateral NA-AION in which the macular star was only observed in one eye (patients 2, 11 and 12). If the development of lipid deposits is related to vascular involvement around the optic disk (as will be argued in the discussion), then it would be assumed *a priori* that the microvascular involvement would be symmetrical in both eyes, with the symmetrical development of macular lipid deposits.

All of the patients were diabetic, except patient 10, who was only hypertensive. In all patients, the macular star was incomplete (hemistar). 6 patients in our study had NPDR (non-proliferative diabetic retinopathy) and 6 other patients had no DR (diabetic retinopathy). Regarding the presence of vascular risk factors, 7 patients out of 12 had hypertension.

The initial mean VA of our patients was 20/300 (considering only the affected eye). The VA improved significantly in 4 patients after several months of development, remaining without significant changes in the rest of patients. The average follow-up time for our series was 7.4 months with a range of 4–12 months. Regarding the color vision measured with the Ishihara test, there was a great variability in the eyes with NA-NAION with a range from 0/15 to 15/15 and an average of 2/15.

It was also observed a large variability in the visual field defects in the affected eyes: 2 patients had superior altitudinal defect; 5 patients, inferior altitudinal defect; 2 patients, central scotoma; 4 patients with inferior-nasal defect and two patients with residual islet.

The time required for the formation of the macular star in the series presented varied between 1 week and 8 weeks. In this regard, one important limitation of our study should be mentioned: we considered the time of the macular star's appearance to be the first visit at which it is detected. However, it is possible that lipid deposits may have already been present prior to the visit. In fact, there were cases in which the macular star was already disappearing when it was first observed (patients 2, 5, 6 and 8).

Fig. 1 shows the fundi in patients 1–12.

Only one patient experienced neurosensory elevation or ophthalmoscopically observed detachment due to the accumulation of subfoveal fluid (patient 12). These findings were confirmed by performing optical coherence tomography (OCT) (Fig. 2).

Discussion

In this paper, we presented a clinical series of 12 patients with NA-AION in whom lipid deposits were detected around the macula during the clinical course of their disease (usually coinciding with the resolution of the optic disk edema) forming an incomplete macular star or a hemistar.

To start with this discussion we will comment the frequency of the possible etiologies that may be responsible for the pattern of optic disk edema with macular star, Chang et al.[7] published a report of a series of 173 eyes presenting with optic neuropathy of different etiologies. Only 23 had macular lipid exudates accompanying optic disk edema. Of these 23 eyes, 15 had neuroretinitis, 6 had NA-AION, and 2 were secondary to papilledema. Particularly for the NA-AION group, these authors found a clear correlation between macular star appearance and the age of patients as well as the existence of diabetes mellitus (DM) and arterial hypertension. It is also noteworthy that there was no worsening of VA secondary to the presence of lipid exudation in this study. This result suggests that decreased VA is due to the presence of optic nerve involvement but not to lipid exudates.

In our series, it was difficult to establish a relationship between the macular star and the deteriorating VA, mainly because VA varies greatly in NA-AION, and in many cases, VA is not recovered. The most common visual field defect in patients with NA-AION is an absolute inferonasal defect followed by a relative inferior altitudinal defect.[8] We suggest that if parafoveal lipid deposits cause visual field changes, these would have to consist of a central defect. In this regard, of the cases presented, 9 eyes (patients 1, 2 left eye, 3 right eye, 4, 5, 7, 8, 11 and 12) had visual field defects typical of NA-AION, and only 4 eyes (patient 2 right eye, 3 left eye, 6 and 10) had absolute central defects. Thus, we can conclude that only 4 eyes in our series (30.7%) could hypothetically have visual field defects attributable to the presence of lipid deposits.

Between the different etiologies mentioned before, the association of optic disk edema and lipid deposits in the form of a macular star occurs mainly in a condition called neuroretinitis. This condition may be idiopathic or secondary to syphilis, cat-scratch disease (CSD), Lyme disease and toxoplasmosis (although there is a long list of infectious agents listed as potential pathogenetic agents in different studies).[9–11]

In neuroretinitis, the primary process involves optic nerve inflammation causing a secondary lipid exudate in the macula (although there are cases of genuine retinitis, especially in CSD).[9,12] Lipid deposits are often not evident initially but develop approximately 9–12 days after the appearance of the optic disk edema.[9,12]