REVIEW ARTICLE

Optic nerve gray crescent can confound neuroretinal rim interpretation: review of the literature

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ABSTRACT • RÉSUMÉ

The optic nerve gray crescent can be of clinical significance if unrecognized during assessment for glaucoma. It has a characteristic appearance of a slate gray area of pigmentation within the disc margins and commonly appears along the inferotemporal or temporal neuroretinal rim areas. This type of disc rim pigmentation can create the impression of neuroretinal rim thinning, and thus lead to the misdiagnosis of glaucoma or "glaucoma suspect" with attendant implications for overtreatment or unnecessary close monitoring of such patients. The gray crescent is more common in African Americans than whites (prevalence rate 27% vs 7%) and is bilateral in at least 58% of cases. It has been reported in association with Kjer optic atrophy type 1. Suggested causes of the gray crescent include an accumulation of melanocytes, or retinal pigment epithelium cells partially located in the optic nerve head region if Bruch's membrane extends internal to the peripapillary scleral ring. Other causes of pigmentation that may resemble gray crescent are conus pigmentosus and variations of peripapillary atrophy. When a gray crescent is present, clinicians should endeavour to identify the true anatomical disc margins via the scleral lip and, if necessary, evaluate the patient further with imaging and visual field studies.

Le croissant gris dans la tête du nerf optique peut avoir une importance clinique s'il n'est pas reconnu pendant l'évaluation du glaucome. Il a l'apparence caractéristique d'une zone de pigmentation grise-ardoise dans les marges de la papille et apparaît le plus souvent le long des zones de la jante neuro-rétinienne inférotemporales ou temporales. Ce type de pigmentation de la marge discale peut créer une impression d'amincissement de la jante neuro-rétinienne et mener ainsi à un diagnostic erroné du glaucome ou de « soupçon de glaucome » avec implications systématiques de surtraitement ou de surveillance étroite non nécessaire de tels patients. Le croissant gris est plus fréquent chez les noirs (prévalence de 27 % vs 7 %) et bilatéral dans au moins 58 % des cas. Il a été signalé en association avec l'atrophie optique Kjer de type 1. Les suggestions d'étiologie concernant le croissant gris comprennent une accumulation de mélanocytes ou de cellules d'épithélium pigmentaire de la rétine, située partiellement dans la région de la tête du nerf optique si la membrane Bruch s'étend à l'intérieur de l'anneau scléral péripapillaire. D'autres causes de pigmentation qui peuvent ressembler au croissant gris sont le « cônus pigmentosus » et une variété d'atrophies péripapillaires. Lorsqu'un croissant gris est présent, les cliniciens peuvent s'efforcer d'identifier les véritables marges discales anatomiques par la lèvre sclérale et, si nécessaire, évaluer davantage le patient avec des imageries et des études du champ visuel.

Various features of the optic nerve head are commonly considered in the context of glaucoma, including measurement of disc size and evaluation of the neuroretinal rim for colour, as well as focal changes such as thinning, notching, sloping, cup-to-disc ratio, and disc hemorrhages. The "gray crescent" (GC) is an important and underrecognized physiologic variant first described by Dr. Bruce Shields[1] in 1980; it can be defined as slate gray pigmentation on or within the neuroretinal rim. This type of disc rim pigmentation can create the impression of neuroretinal rim thinning, and thus lead to the misdiagnosis of glaucoma or "glaucoma suspect" with attendant implications for overtreatment or unnecessary close monitoring of such patients.[2,3] We present a case of GC detected through our teleglaucoma program, and review the literature surrounding GC and select entities involving pigmentation within or around the disc margin.

Case Report

A 37-year-old African male was referred from an optometrist for "abnormal optic nerves." He had a spherical equivalent refraction of –2.75 OD, –2.50 OS and best corrected vision of 20/30 OU. The IOP measured by noncontact tonometry was 13 mm Hg OD and 12 mm Hg OS. The vertical cup-to-disc ratio was 0.75 OU. No visual field information was provided. Based on this referral information, particularly the nerve description, we believed the patient could be at risk for glaucoma; thus, we assessed the patient through our hospital-based teleglaucoma system.[4]

The patient had no ocular symptoms, prior ocular history, or family history of glaucoma, and was taking no ocular or systemic medications. Central corneal thickness was 543 OD and 536 μm OS. Slit-lamp examination was unremarkable, with intraocular pressure (IOP) 16 OD and 15 mm Hg OS by Goldmann applanation tonometry. Stratus optical coherence tomography (OCT) was of high quality (signal strength = 10 OU) and revealed average retinal nerve fibre layer (RNFL) thickness of 113 μm OD and 111 μm OS, with no focal RNFL thinning evident. The disc diameters were assessed as large by OCT: 2.41 mm OD and 2.41 mm OS. Frequency doubling technology field testing was reliable and normal OU. Evaluation

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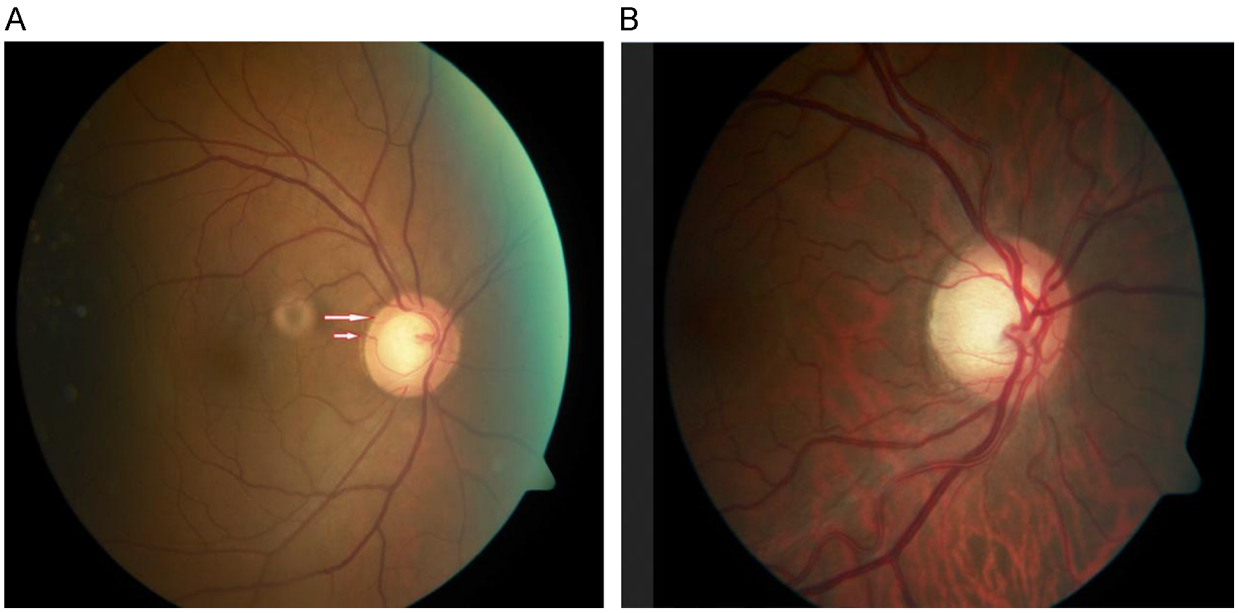

Fig. 1—A, OD with larger arrow pointing to underestimated disc margin and smaller arrow marking the actual disc margin with associated gray crescent temporally. B, Another case with a gray crescent OD covering the temporal disc margin.

of stereoscopic digital fundus photos of the right optic nerve revealed a large disc, vertical cup-disc ratio of 0.7, and a temporal GC with some adjacent peripapillary atrophy (PPA; Fig. 1). The left nerve was also large, with vertical cup-to-disc ratio of 0.7 and mild inferior thinning noted. There was a more prominent temporal GC OS with some adjacent PPA.

In light of the normal IOP, CCT, visual field test, RNFL findings on OCT, and GC temporally, the patient was thought to have physiologic rather than pathologic cupping, and hence the patient was considered unaffected for glaucoma. We recommended the patient continue to see an optometrist for annual evaluations, and if any concerns arose with regard to structure or function of the nerves, we could reassess.

Discussion and Literature Review

Originally described by Shields[1] in 1980, the GC is an often underrecognized anatomic optic nerve variant. Relevant to this discussion is a definition of the area of the optic nerve head, which Jonas[5,6] described as all areas inside the peripapillary scleral ring. Outside of the scleral ring is the peripapillary region, which is characterized by various other abnormalities discussed later.[7] The scleral ring is an anterior extension of sclera appearing as a thin white rim marking the disc margin.

According to Shields,[1,2] the GC is characterized by increased pigmentation of slate gray colour within the substance of the optic nerve head. Jonsson et al. extended the definition to the occurrence of a pigmented crescent that appeared to be located on or within the neuroretinal rim tissue, that is, inside the scleral ring.[3] In some instances, the true disc border can be seen, although partially obscured by the GC (Fig. 2).

The clinical significance of GC arises when it is mistaken for neuroretinal rim thinning and suspicious of glaucoma. Nonetheless, the diagnosis of glaucoma is often based on an integration of all available information including history, IOP, nerve examination, imaging, and visual field testing. In addition, taking a fundus photo to serve as a baseline may help for assessing progression/change.

In the initial report by Shields,[1] a consecutive series of 29 patients with GC were described with a mean age of 35

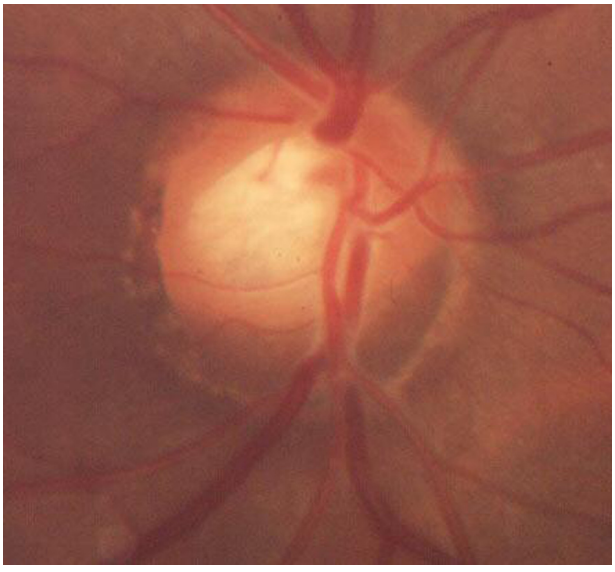

Fig. 2—Gray crescent OD with visible underlying disc margins.