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Is there an association between optic disc drusen and angioid streaks?

A.M. Mansour

Department of Ophthalmology, University of Texas Medical Branch, Galveston, TX 77550, USA

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Abstract. We reviewed the color photographs and fluorescein angiograms of 110 subjects with angioid streaks. Evidence is presented for an association between drusen of the optic disc and angioid streaks.

Introduction

Drusen of the optic disc have been associated with small optic discs [6] and with tapetoretinal degeneration [3]. We present evidence for an association between drusen of the optic disc and angioid streaks.

Patients and methods

We reviewed stereoscopically the color photographs and fluorescein angiograms of 110 consecutive subjects (from 110 families) with angioid streaks referred to the Retina Service of Wills Eye Hospital between March 1967 and June 1983. Prominent drusen of the optic disc were detected in 10 eyes of 5 subjects. No familial cases of disc drusen were included because family members were excluded from the study. The clinical profile of the 5 subjects with disc drusen was as follows: 4 males and 1 female, 4 whites and 1 black, and 2 out of 5 having pseudoxanthoma elasticum. In the present series, 4.5% of subjects with angioid streaks had drusen of the optic disc in both eyes. Likewise, Meislik et al. [7] detected optic disc drusen in 5.8% of 43 cases with pseudoxanthoma elasticum. The above observations contrast with the low incidence of disc drusen in clinical settings (0.34%), as found by Lorentzen in a study of 3200 subjects.

To analyze the relationship between optic disc drusen and angioid streaks further, we reviewed the photographic files of patients with optic nerve drusen seen between 1981 and 1988 at the Retina Service of the University of Texas Medical Branch. Out of 21 patients with established drusen of the optic disc, 1 patient (5%) had angioid streaks. This contrasts with the low prevalence of angioid streaks in the general population (estimated at 1/80,000) [5]. To check for selection bias (i.e., whether the occurrence of optic nerve head drusen or angioid streaks is unusually high among patients referred for fundus photography), we reviewed the fundus photographs and fluorescein angiograms of 100 consecutive patients who had fundus photographs. Forty patients had fluorescein angiograms and 1 patient underwent red-free photography without fluorescein dye injection. There was one case of bilateral optic disc drusen (1%) and no case of angioid streaks among the 100 patients reviewed. Moreover, a review of the fundus photographs and clinical records of 175 consecutive HIV-infected individuals in the same medical center disclosed one case of optic disc drusen (0.57%) and no case of angioid streaks.

Results and discussion

A possible association between angioid streaks and optic disc drusen has been alluded to in several case reports [1, 2, 3, 7]. Our observations point to the presence of such an association.

We propose the following hypothesis to account for the above association. Elastin densely lines the insertion of the lamina cribrosa into the sclera and is prominent in the laminar beams [8]. Elastin mineralization (as seen in Bruch's membrane in eyes with angioid streaks [5]) and adherence of abnormal glycosaminoglycans to elastic fibers (as seen in the dermis of patients with pseudoxanthoma elasticum [10]) of the lamina cribrosa can lead to marked thickening of the laminar beams of the optic nerve. This thickening in a tight compartment leads to crowding of the laminar portion of the optic nerve, secondary alteration of axoplasmic transport [9], and formation of optic nerve head drusen. This hypothesis awaits pathologic documentation of laminar beam mineralization and thickening.

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