Case report

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Focal choroidal excavation associated with nonmelanocytic iris tumor

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Abstract

Purpose: To describe a unique unilateral association between an iris stromal tumor and a macular focal choroidal excavation.

Case Description: A 40-year old patient presented with a small iris tumor associated with a unilateral macular lesion disclosed during a routine ophthalmologic examination. The patient was asymptomatic and visual function was not affected. After clinical and instrumental evaluation, a diagnosis of nonmelanocytic undefined stromal tumor of the iris associated with macular focal choroidal excavation was made. The size and shape of the two lesions remained stable during a 7-year follow-up and the patient did not develop other signs.

Conclusion: The concurrent presence of a stromal iris tumor associated with focal choroidal excavation has never been reported. Further reports of this association are required in order to understand its exact pathogenesis.

Keywords

iris, stromal tumor, focal choroidal excavation, macula

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Introduction

The most common iris lesions are iris nevi, iris melanomas and iris pigment epithelium cysts.[1] Nonmelanocytic tumors of the iris are relatively uncommon lesions, that have to be considered in the differential clinical diagnosis. Focal choroid excavation (FCE) is a rare localized area of intrachoroidal cavitation, mostly located in the macular region, normally seen in eyes with no other known pathology and it is typically asymptomatic.[2] The occurrence of FCE in conjunction with various retinal pathologies has already been described, but its association with a unilateral nonmelanocytic iris tumor has never been reported.

Case description

A 40-year old Caucasian male was referred for evaluation for a small iris tumor associated with a macular lesion disclosed during a routine ophthalmologic examination. The patient was asymptomatic and his previous medical history was unremarkable, with no personal or familiar history of ocular pathology. Best-correct visual acuity (BCVA) was 20/20 Snellen equivalent in both eyes. The anterior segment and fundus examination of the right eye was unremarkable.

In the left eye, a solid, translucent, slightly elevated triangular tumor was present on the nasal side of the iris, extending approximately 3mm from the pupillary margin (Figure 1, A and B). The pupil was round with no afferent pupillary defect; the anterior chamber angle was open with normal structures and absence of pigment dispersion. Intraocular pressure was 14 mmHg. The lens was regular-shaped, transparent, and clear.

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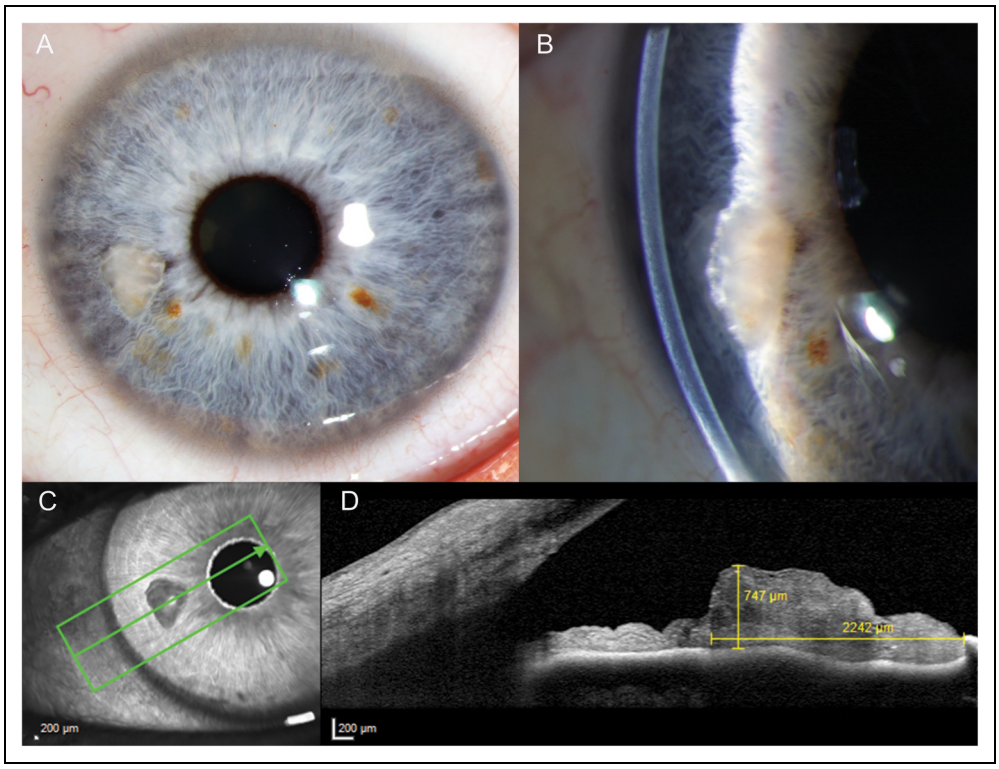

Figure 1. Slit-lamp photographs and anterior segment optical coherence tomography of the iris tumor.
Translucent, triangular-shaped, nonmelanocytic tumor located on the nasal side of the iris (A, B, magnification 10x and 16x, respectively). Anterior segment optical coherence tomography scan of the lesion shows a homogenous hyperreflective mass, presenting a larger diameter of 2242 µm and a thickness of 747 µm (C, D).

On fundus examination, a juxtafoveal round-shaped yellowish-grey, half-disk diameter in size, empty lesion with clear margins was present (Figure 2, E). Peripheral retina and optic disc were within normal limits. In the early and venous phase of ICGA a well-defined hypofluorescent area was present and the choroidal defect was larger than the retinal pigment epithelium (RPE) defect (Figure 2, F). A large choroidal vessel was visible through the RPE defect. Optical coherence tomography (OCT) scan over the lesion revealed the presence of a choroidal defect corresponding to a focal choroidal excavation (FCE). The neuroretina tissue collapsed through the RPE opening which was smaller than the choroidal defect. On the adjacent noninvolved choroidal area, a relative pachychoroid (373 µm) was observed and a large choroidal vessel was visible at the bottom of the FCE (Figure 2, G and H). On high-frequency ultrasound biomicroscopy (UBM) and anterior segment OCT the iris tumor appeared as a solid homogeneous mass with the same reflectivity as the stromal tissue surrounding the tumor. No extension or infiltration to the posterior iris surface, the iris root or the ciliary body was present (Figure 1, C and D). A clinical diagnosis of nonmelanocytic undefined stromal tumor of the iris was made and a periodic observation was recommended.

The patient was followed for seven years, and both the iris tumor and the focal choroidal excavation did not show any documented modification. The size and shape of the two lesions remained stable and the patient did not develop other signs.

Nonmelanocytic tumors of the iris are relatively uncommon, representing 11% of solid iris tumors.[3] Clinical differential diagnosis includes amelanotic melanomas or nevi, inflammatory lesions, and metastases.[4] FCE, first described by Jampol et al. in 2006,[5] is a rare localized area of intrachoroidal cavitation, mostly located in the macular region, normally seen in eyes with no other known pathology and are typically asymptomatic. FCE pathogenesis still remains unclear. According to some authors, these lesions may arise from a congenital development defect while others suggest that they could be related to subclinical choroidal inflammation or choroidal ischemia.[6] The occurrence of FCE in conjunction with various retinal pathologies such as central serous chorioretinopathy, choroidal neovascularization, age-related macular degeneration,

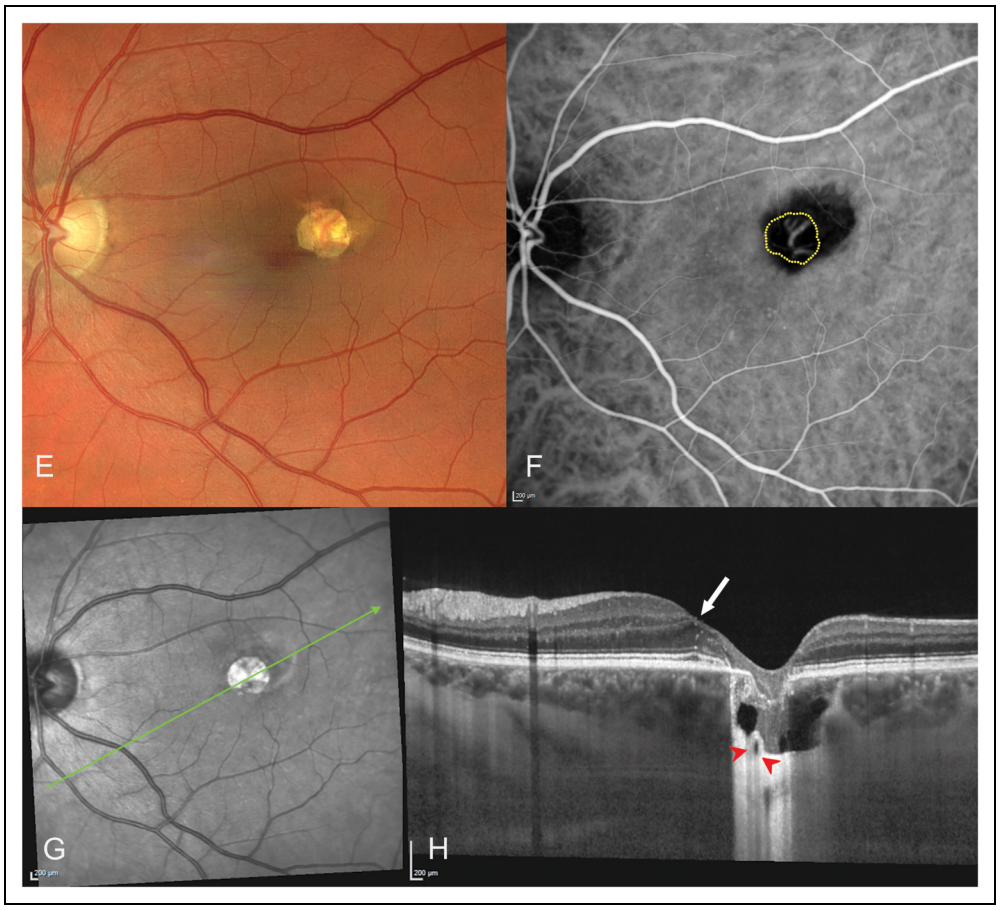

Figure 2. Multimodal imaging of the focal choroidal excavation.
(E) Fundus photography of the macular region. Round shaped yellowish, half-disc diameter retinal pigment epithelium (RPE) defect with clear margins corresponding to the focal choroidal excavation (FCE). (F) Venous phase of indocyanine green angiography (ICGA). Well-defined hypofluorescent choroidal area, larger compared to the RPE defect (yellow dot line). A large choroidal vessel is visible through the RPE defect. (G and H) Infrared (IR) and Enhanced-Depth Imaging spectral domain optical coherence tomography (EDI SD-OCT). Absence of choroidal vascular tissue in the focal choroidal excavation which is larger than the RPE defect. Collapse of the neuroretina tissue through the RPE defect. The same large choroidal vessel seen on ICGA is visible on EDI SD-OCT on the bottom of the FCE (red arrowhead). Absence of modifications of the external retinal layers on the fovea located next to the FCE (white arrow).

vitelliform macular dystrophy, and within the pachychoroid disease spectrum was described.[7]

Conclusion

To our knowledge, there is no previous report of a stromal iris tumor associated with unilateral focal choroidal excavation. Although the association between these two lesions may be considered as fortuities, we hypothesized that a specific insult to the developing structures during embryogenesis leading to the simultaneous development of these two lesions is not excluded. Moreover, particular attention to chorioretinal abnormalities in iris tumor patients should be increased. Further reports of this association, histopathological testing, and genetic counselling would be required in order to understand the exact pathogenesis.

Patient Consent

Written informed consent was obtained from the individuals for the publication of any potentially identifiable images or data included in this article.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of conflicting interests

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