

## Case report

# Optical coherence tomography angiography of a pigmented Fuchs' adenoma (age-related hyperplasia of the nonpigmented ciliary body epithelium) masquerading as a ciliary body melanoma



Nisha Nagarkatti-Gude, Yan Li, David Huang, David J. Wilson, Alison H. Skalet\*

Casey Eye Institute, Oregon Health and Science University, Portland, OR, USA

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## ABSTRACT

**Purpose:** To report a case of age-related hypertrophy of the nonpigmented ciliary epithelium (ARH-NPCE) clinically resembling a ciliary body melanoma and report the optical coherence tomography angiography (OCTA) findings associated with this lesion.

**Observations:** A 51-year-old male was referred for evaluation of a deeply pigmented ciliary body mass with extension through the iris root. Iridocyclectomy was performed due to concern for ciliary body melanoma. Histopathologic analysis was instead consistent with ARH-NPCE, also known as Fuchs' adenoma. Prior to surgery, OCTA images revealed abnormal vasculature in the area of the lesion. Vessels in the peripheral iris approaching the lesion appeared more tortuous and were non-radial as compared with normal iris vessels. The ciliary body mass itself could not be penetrated using an OCTA system operating at 1050 nm.

**Conclusions and importance:** ARH-NPCE may clinically resemble a pigmented ciliary body melanoma. This is the second case describing this clinical scenario, which may be more common than previously thought. Non-invasive imaging with OCTA revealed an abnormal peripheral iris vasculature pattern in the area of the iridociliary mass characterized by disorganized, tortuous, and non-radial vessels. Despite advances in longer wavelength OCTA systems, poor penetration of the ciliary body lesion precluded imaging of the intratumoral vessels in this location.

## 1. Introduction

Ernst Fuchs first described benign tumors of the non-pigmented ciliary epithelium (coronal adenomas) in 1883<sup>1</sup> and hence, this lesion was termed Fuchs' adenoma. Some authors prefer the term age-related proliferation of the nonpigmented ciliary epithelium (ARH-NPCE) as the lesions are believed to be reactive proliferation of the nonpigmented ciliary epithelium associated with aging.<sup>2</sup> ARH-NPCE have been found as a common incidental finding on enucleated eyes but are less commonly detected clinically due to their small size and relatively occult location. Timm and Fritsch examined 200 enucleated eyes from children and found no Fuchs' adenomas<sup>3</sup>; in contrast, a study by Illiff et al. examined 100 enucleated eyes from adults and found 14% had the incidental finding of Fuchs' adenomas confirmed by histopathology.<sup>2</sup> Moreover, Illiff et al. noted that more than 50% of these tumors were found in patients older than 80 years of age.<sup>2</sup> Bateman and Foos also performed histopathologic evaluation of 500 adult eyes obtained at the time of autopsy and found these tumors present in 31% of patients.<sup>4</sup> Among eyes with Fuchs' adenoma in this study, sectoral cataract was

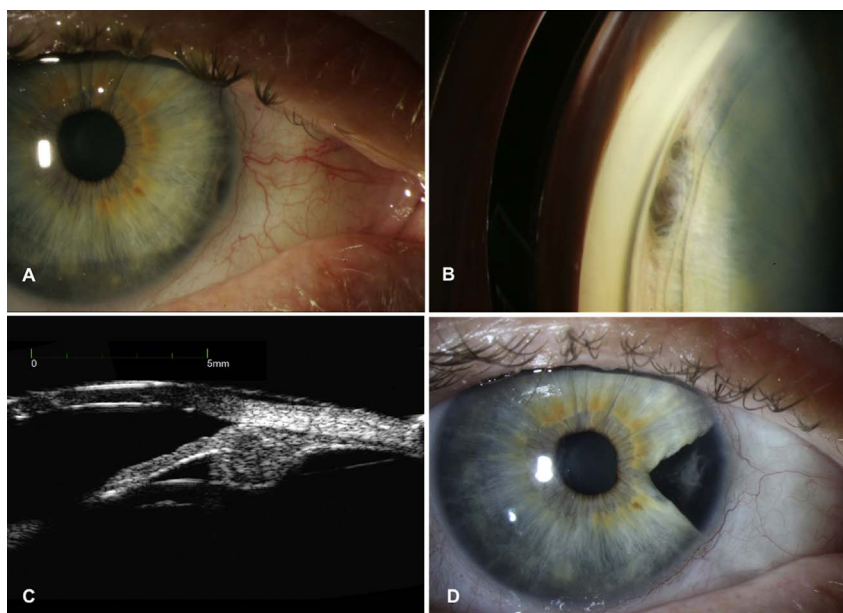
present corresponding to tumor location in 6% of cases.<sup>4</sup>

While it has been reported that these lesions may simulate melanoma, diagnosis of ARH-NPCE in a patient with ciliary body neoplasm is rare. To date there are only two cases in the literature describing lesions of the peripheral iris or ciliary body excised due to concern for malignant melanoma that were subsequently found to be Fuchs' adenoma on histopathologic examination.<sup>5,6</sup> Here we describe an additional case in which an ARH-NPCE was diagnosed after surgical excision of a ciliary body mass with extension through the iris root. The lesion was particularly unusual due to its deep pigmentation and relatively large size. We also describe the optical coherence tomography angiography (OCTA) findings associated with this mass. OCTA is a non-invasive vascular imaging technique with an emerging role in the imaging of intraocular and conjunctival tumors.<sup>7–9</sup>

## 2. Case report

A 51-year-old male with a past medical history of hypertension, hyperlipidemia, and mitral valve prolapse was referred to the ocular

\* Corresponding author. Casey Eye Institute, Oregon Health and Science University, 3375 SW Terwilliger Blvd, Portland, OR, 97239, USA.  
E-mail address: [skalet@ohsu.edu](mailto:skalet@ohsu.edu) (A.H. Skalet).



**Fig. 1.** Slit lamp, gonioscopy, and ultrasound biomicroscopy pictures of the lesion. A, Slit lamp colored picture with pigmented tumor present at ~3:30 eroding through the iris root. B, Gonioscopy demonstrating two foci of pigmented tumor extending through ciliary body band and iris root. C, Ultrasound biomicroscopy (longitudinal scan at 3 o'clock, 40 MegaHertz) showing ciliary body tumor with scale from 0 to 5 mm. D, Appearance of the eye 2 years post-operatively.

oncology service at Casey Eye Institute for the evaluation of a lesion involving the angle that was not visualized on prior eye exams. The patient had no ocular complaints at the time of presentation.

On presentation, visual acuity was 20/30-2 in the right eye and 20/20-2 in the left eye. Intraocular pressure was measured as 15 and 18 mm Hg in the right and left eyes, respectively. There was a small, flat, pigmented lesion in the temporal periphery consistent with a choroidal nevus present in the left eye. Examination of the left eye was otherwise unremarkable. Examination of the right eye revealed a pigmented ciliary body lesion eroding through the iris root at 3:30 with prominent intrinsic vascularity (Fig. 1A and B). There was a sectoral cortical cataract present nasally in the area of the ciliary body lesion. The posterior segment exam was unremarkable. Ultrasound biomicroscopy (UBM) was obtained to further evaluate the lesion. UBM showed a ciliary body mass measuring 1.8 mm in height, 2.7 mm longitudinally, and 2.8 mm in transverse length (Fig. 1C).

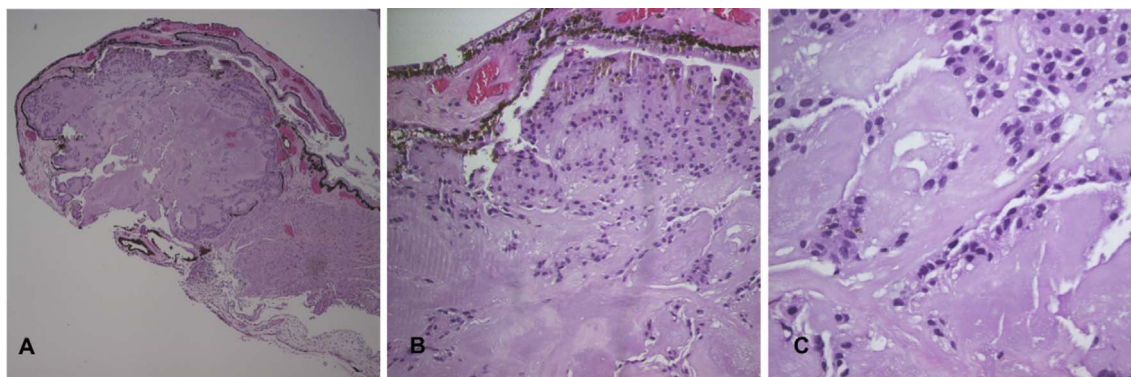
The differential diagnosis included ciliary body melanoma, leiomyoma, and tumor of the pigment epithelium. ARH-NPCE was not considered due to the pigmentation of the mass, the patient's relatively young age, and the size of the lesion. The patient elected to proceed with iridocyclectomy. Gross examination of the excised lesion disclosed a smooth, dark brown mass with whitish-gray associated tissue measuring 4 × 5 × 2.5 mm. Hematoxylin and eosin stained sections (Fig. 2A–C) revealed a proliferation of chords of non-pigmented ciliary epithelium with abundant acellular eosinophilic basement membrane

material. No cellular atypia was found. This pathologic evaluation was consistent with ARH-NPCE. The patient is now two years out from surgery and is 20/25 in the eye, with stable sectoral cataract (Fig. 1D).

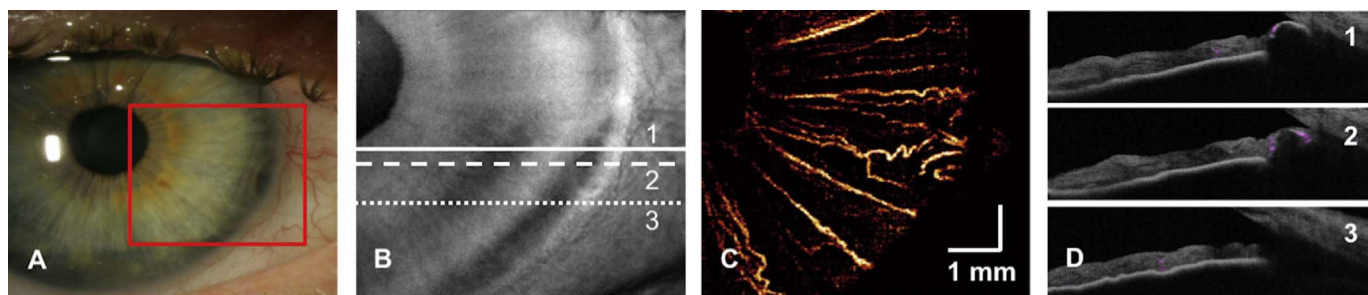
OCTA was performed on the tumor prior to surgical excision of the lesion. The subject's right eye was evaluated using a swept-source, anterior segment OCT operating at a wavelength of 1050 nm with an axial scan repetition rate of 100 kilohertz using a technique that has been previously described.<sup>7</sup> The anterior iris surface and the anterior boundary of the iris pigmented epithelial layer were segmented in cross-sectional OCT images then the *en face* iris angiogram was constructed by projecting the maximal flow signal between the two iris boundaries. OCTA revealed an abnormal vascular pattern in the peripheral iris in the area of the iridociliary mass (Fig. 3A–D). The vessels in this location were disorganized and non-radial. They appeared more tortuous than typical iris vessels.<sup>7</sup> The ciliary body mass itself could not be visualized with OCTA due to limited penetration.

### 3. Discussion

ARH-NPCE are common benign growths found in the eyes of older individuals, but rarely grow to a size in which they are clinically apparent or likely to be mistaken for malignant ciliary body melanoma. Ours is the third reported case of ARH-NPCE diagnosed after surgical excision of a mass concerning for melanoma involving the ciliary body and/or peripheral iris. In each of the previously reported cases, the



**Fig. 2.** Histopathologic hematoxylin-eosin stained images of mass. A, Hematoxylin and eosin stained section of tumor at x4 magnification. B and C, Hematoxylin and eosin stained sections at x20 and x40 magnification respectively showing cords of non-pigmented ciliary epithelium cells separated by eosinophilic basement membrane material.



**Fig. 3.** Slit lamp, structural optical coherence tomography (OCT), OCT Angiography (OCTA), and cross-sectional OCT images of tumor. A, Slit lamp colored picture revealing OCTA scanning area. B, Structural OCT through lesion. C, *En face* iris angiogram demonstrating abnormal tortuosity of tumor vessels compared to normal, radially-oriented iris vessels. D, Cross-sectional OCT images through iris surface and anterior extent of iris pigmented epithelium utilized to construct the *en face* angiogram by projection of the maximal flow signal between these boundaries.

tumor caused sectoral lenticular changes much like the patient described in this report. One case describes an elevated pigmented peripheral iris lesion in a 73 year old woman,<sup>5</sup> and the other, quite similar to this case, describes a “tan colored” lesion in a 53 year old woman that had eroded through the iris root.<sup>6</sup> Collectively, our case and the Shields’ case<sup>6</sup> are remarkable as ARH-NPCE are normally described as amelanotic, non-pigmented lesions that are quite small in size and occur in older patients. These cases of larger, variably pigmented ARH-NPCE suggest that while rare, this presentation may be more common than previously recognized. It is possible that clinicians empirically treating pigmented ciliary body neoplasms with radiation therapy without diagnostic biopsy may occasionally be treating these benign lesions in individuals suspected of having melanoma.

In this case, the patient elected for iridocyclectomy due to concern for melanoma given clinical findings. An alternative diagnostic approach discussed with the patient was fine needle aspiration (FNA) for cytologic evaluation of the lesion. Glasgow characterized the cytologic findings from intraocular biopsies of coronal adenomas from three enucleated eyes.<sup>10</sup> In this study, the coronal tumors had a more classic gross appearance and cytologic studies revealed cohesive sheets of epithelial cells with abundant cytoplasm and bland, round nuclei without atypia or mitotic figures.<sup>10</sup> Risks of FNA for iridociliary lesions include low yield and sampling error as well as a small risk for tumor seeding; however, this procedure is routinely performed with good diagnostic yield and safety profile.<sup>11,12</sup>

Our case is the first to our knowledge evaluating a ciliary body neoplasm with OCTA. Our findings demonstrate that OCTA can successfully image the vasculature of tumors involving the iris root at an operating wavelength of 1050 nm; however, poor penetration of the ciliary body itself precludes imaging of the intratumoral vessels of ciliary body lesions at the present time. Evaluation of vascular patterns of anterior segment tumors may provide a non-invasive means of differentiating tumors and assessing tumor activity in the future.

#### Patient consent

The subject was recruited from a tertiary ocular oncology clinic based at the Casey Eye Institute in accordance with HIPAA regulations. After written informed consent was obtained, as part of an IRB-approved study, the subject’s right eye was evaluated using OCT angiography. The patient’s oral consent was also obtained for the publication of this case report.

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**Conflict of Interest:** Oregon Health & Science University (OHSU), Y.L. and D.H. have a significant financial interest in Optovue, Inc, a company that may have a commercial interest in the results of this research and technology. These potential conflicts of interest have been reviewed and managed by OHSU.

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

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