• Letter to the Editor •

Optical coherence tomography-angiography of juxtapapillary hamartoma

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Received: 2016-09-16 Accepted: 2016-12-16

DOI:10.18240/ijo.2017.08.23

Citation: Cennamo G, Romano MR, Breve MA, Velotti N, de Crecchio G, Cennamo G. Optical coherence tomography-angiography of juxtapapillary hamartoma. *Int J Ophthalmol* 2017;10(8):1328-1330

Dear Editor,

I'm Dr. Gilda Cennamo from the Eye Clinic of Department of Neurosciences, Reproductive Sciences and Dentistry, University of Naples Federico II, Naples, Italy. I write to present four cases of juxtapapillary hamartoma evaluated with non-invasive optical coherence tomography-angiography (OCT-A).

The term "hamartoma" was coined by Albrecht[1] in 1904 to describe a group of benign tumor-like malformations arising from deranged mixing or deranged development of normal tissue in an organ. Hamartomas of the retina, the retinal pigment epithelium (RPE) are characterized by a mound of disorganized glial, vascular and melanocytic tissue; these alterations are also found in the papilla[2].

Spectral-domain optical coherence tomography (SD-OCT) of combined hamartoma of the retina and RPE shows an elevated pigmented mass that is commonly connected to the epiretinal membrane[3] but does not adequately image the vascular compartment. In this scenario, we evaluated the vascular features of juxtapapillary hamartomas in four patients using non-invasive OCT-A.

In this prospective study we evaluated four eyes of four patients affected by juxtapapillary hamartoma seen in the Eye Clinic of the University of Naples Federico II between September 2015 and December 2015. The study protocol was approved by the Institutional Review Board of the University of Naples Federico II, and adhered to the tenets of the Declaration of Helsinki. No patient had coexisting systemic neurofibromatosis type 1 or 2.

Each patient underwent evaluation of best corrected visual acuity (BCVA) according to the Early Treatment of Diabetic Retinopathy Study (ETDRS), A-scan and B-scan bulbar echography (Quantel Medical, Clermont-Ferrand, France), multicolor imaging, SD-OCT and fluorescein and indocyanine angiography (Spectralis HRA+, Heidelberg Engineering, Heidelberg, Germany), widefield en-face OCT and OCT-A (Optovue AngioVue System, Optovue Inc., Fremont, CA, USA).

OCT-A has an A-scan rate of 70 000 scans per second with a tissue axial resolution of 5 μm and a 15-μm beam width, each B-scan contained 304 A-scans. This new technique analyzes blood flow through the "split spectrum amplitude decorrelation" algorithm: blood flowing through vessels changes reflectance over time and localized areas of flow decorrelation between frames. The spectrum of the light source was split in multiple component parts to decrease the noise of images and then decorrelation was carried out to obtain an image of the contained blood flow[4].

The tumor area was measured with a 3×3 scan centered on the optic disc. We evaluated, simultaneously, the superficial vascular plexus, the deep vascular plexus, the outer retina and the RPE area (choroidal cup)

The median age of our four patients at diagnosis was 13.7y, and three were male. The BCVA of affected eyes was 0.1 logMAR to counting finger secondary to severe retinal disorganization and loss of photoreceptors in the macular region. All tumors involved the optic nerve. In three of the four patients, the tumor was located in the left eye. Multicolor images showed a green lesion at the level of the optic disc (Figures 1A, 2A). At standardized A-scan echography, the mean tumor basal dimension was 2.98 mm. SD-OCT examination showed retinal fluid in three affected eyes, retinal striae overlying the tumor in two eyes, retinal schisis in two eyes and vitreo-retinal traction in all eyes (Figures 1E, 2H).

Widefield en-face OCT revealed epiretinal membranes in all four patients: in two with foveal involvement, in one with extrafoveal involvement, and in two with both foveal and extrafoveal involvement. Vitreo-retinal traction was horizontal (tangential) in one patient, vertical (anteroposterior) in one

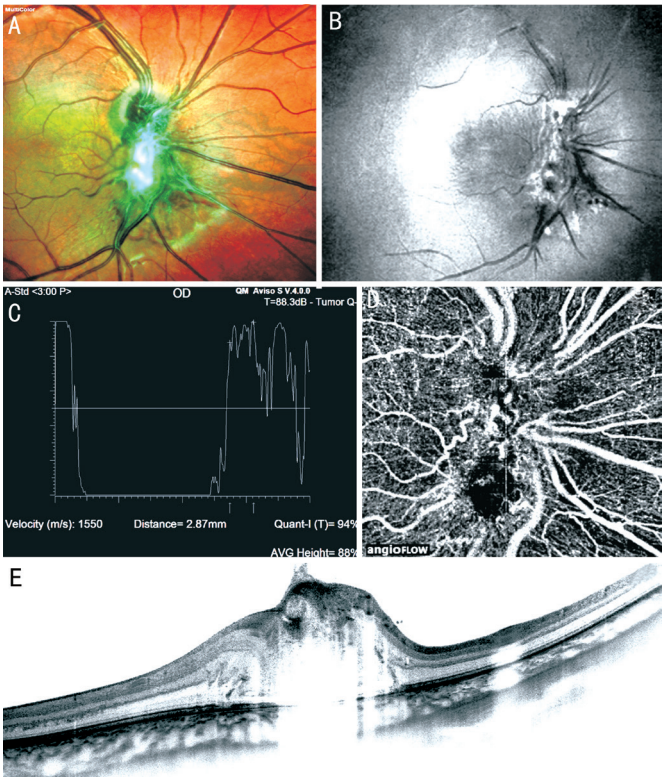

Figure 1 Juxtapapillary hamartoma in the left eye of a 8-year-old boy A: Multicolor image of the lesion; B: En-face widefield OCT revealed retinal folds emanating from the tumor; C: A-scan echography showing eye reflectivity in the internal part of the lesion; D: OCT angiography revealed vascular tortuosity and rarefied capillaries; E: SD-OCT scans through the tumor show retinal thickness, horizontal and vertical traction.

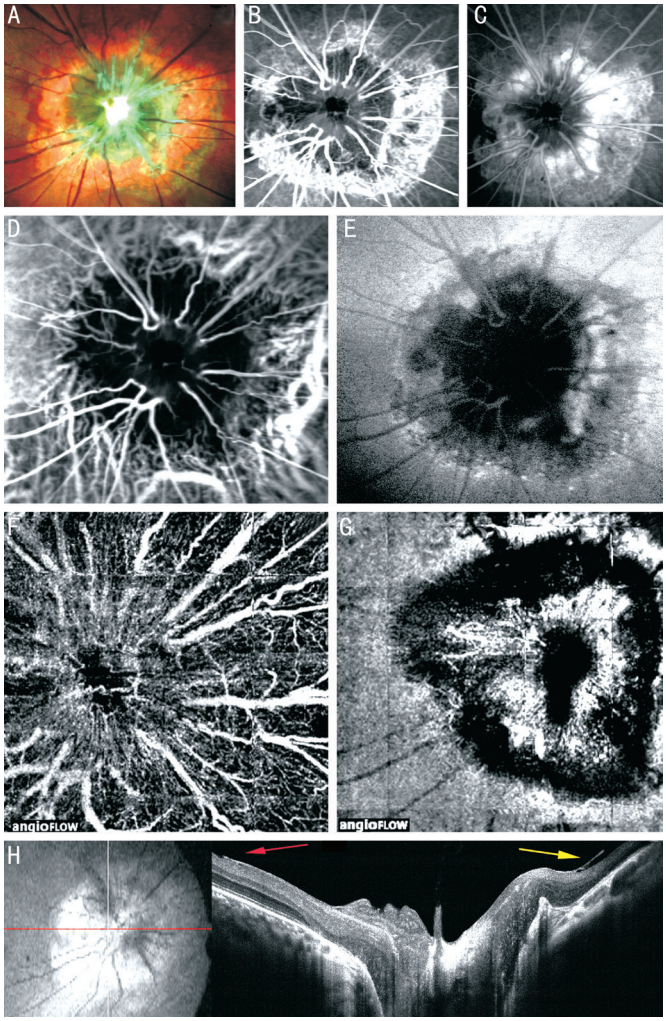

Figure 2 Juxtapapillary hamartoma in the left eye of a 22-year-old man A: Multicolor image of the tumor; B, C: Early and late phase fluorescein angiography showing fine vascularization in the center of the neoformation and numerous anastomotic vessels; D, E: Early and late phase indocyanine angiography show vascular tortuosity; F, G: OCT angiography of the superficial and deep capillary network reveals vascular irregularities within the tumor; H: SD-OCT scans through the tumor shows horizontal traction (red arrow) and vertical traction (yellow arrow).

patient, and both horizontal and vertical in two patients (Figure 1B). Early phase fluorescein and indocyanine angiography images showed fine vascularization in the center of the tumor and numerous anastomotic vessels, which were not visible in the later phase due to die leakage (Figure 2). Moreover, fluorescein angiography revealed ophthalmoscopic modifications of vessels and of the optic disc, characterized by an increase of fluorescein in the centre of the lesion due to the permeability of capillaries (Figures 2B-2E). OCT-A showed a series of vascular irregularities, namely, dilatation and vascular tortuosity and rarified capillaries (Figures 1D, 2F and 2G).

Clinically, the differential diagnosis of juxtapapillary hamartoma is with choroidal melanoma, retinoblastoma and neoplasm of the optic nerve with endobulbar manifestation. Gass[6] reported the histopathological features of these lesions as follows: thickening of the retina and optic nerve due to replacement of the normal architecture of the retina and optic nerve by dysplastic glial vascular tissue infiltrated by cords, strands and sheets of pigment epithelial cells; a sheet of fibrous tissue proliferation bridging the folded anterior surface of the lesion; and an unusual pattern of dilated capillaries. Hamartomas gradually lead to visual deficiency, but they never evolve to malignancy[7].

In our study, standardized A-scan echography showed a lesion with high reflectivity (about 85%) without acoustic shadowing, thereby excluding both malignant melanoma and retinoblastoma, and indicating an angiomatous malformation. Fluorescein angiography images showed modifications of vessels in the center of the tumor which were not visible in the later phase due to die leakage. Instead, OCT-A showed a series of vascular irregularities, namely, dilatation and vascular tortuosity and rarefied capillaries. The deformation of retinal vessels toward the center of tumor caused stretching of capillaries in the circumpapillary region which was clearly visible on OCT images. This technique also showed that the superficial retinal vessels had lost most of their collateral branches and presented many loops. At deep plexus level, the OCT images showed alterations of vessel size and morphology.

These findings confirm the histopathological features of the tumor reported by Gass[6].

In conclusion, OCT-A imaging provides an accurate picture of the vascularization of juxtapapillary hamartomas. This non-invasive technique may improve the diagnosis and follow-up of patients affected by this benign tumor.

ACKNOWLEDGEMENTS

We thank Jean Ann Gilder (Scientific Communication srl., Naples, Italy) for editing the manuscript.

Conflicts of Interest: Cennamo G, None; **Romano MR**, None; **Breve MA**, None; **Velotti N**, None; **de Crecchio G**, None; **Cennamo G**, None.

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