Swept-source optical coherence tomography of an optic disc melanocytoma: The importance of the hyperreflective foci

Brijesh Takkar, Kabiruddin Molla, Pradeep Venkatesh

Optic disc melanocytoma (ODM) has been considered as a benign tumor with few reports of malignant transformation. We present swept-source optical coherence tomography (SSOCT) imaging of a case of ODM. As attaining histopathology is impossible in most cases, we discuss the possibility of using SSOCT as a tool for ruling out choroidal invasion or juxtapapillary melanoma.

Key words: Choroidal melanoma, optic disc melanocytoma, swept-source optical coherence tomography

Swept-source optical coherence tomography (SSOCT) imaging is advantageous over the conventional OCT systems in evaluating ocular tumors because of its better tissue penetration.[1] Optic disc melanocytoma (ODM) was initially considered to be malignant. In the past, enucleation was performed until evidence regarding the benign nature of the lesion was made clear. Reported cases of malignant melanocytomas were, in fact, primary peripapillary melanomas. The risk of malignant transformation of this lesion is quite low.[2]

We present the SSOCT features of an ODM. We discuss the role of SSOCT in diagnosing choroidal extension of the tumor and ruling out juxtapapillary melanoma of choroidal origin.

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/ijo.IJO_642_17

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Manuscript received: 26.07.17; Revision accepted: 18.09.17

Case Report

A 28-year-old male underwent a routine ophthalmic evaluation. Best-corrected visual acuity was 6/6 and N6 in both eyes. Pupil light reflexes, the anterior segments, and intraocular pressures were within normal limits in both eyes. While the right eye fundus examination was normal, ophthalmoscopy of the left eye revealed a densely pigmented brownish-black, dome-shaped nodular mass overlying the optic disc. The mass spared the superior and nasal margins of the optic disc [Fig. 1]. It measured to around 3 mm in its longest basal dimension and was well-defined. Overlying vitreous, macula, and surrounding retina appeared normal. A clinical diagnosis of ODM was made.

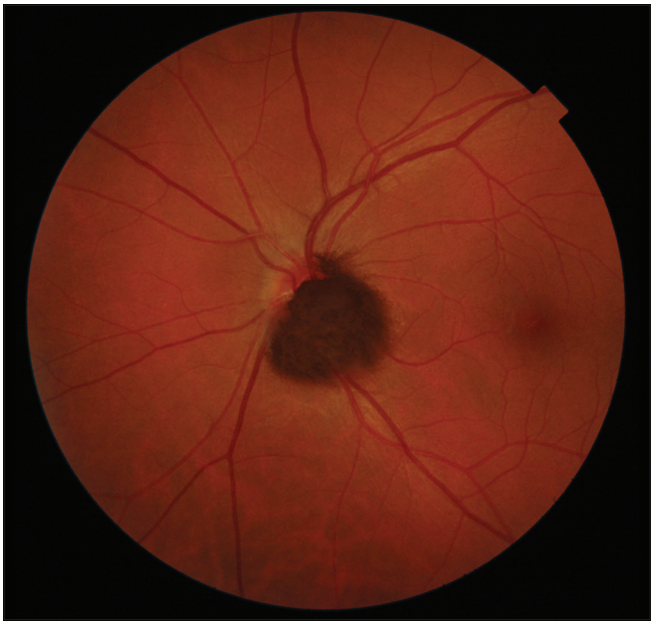

Figure 1: Fundus photograph of the left eye depicting a brownish-black mass overlying the optic disc and obscuring its inferior and temporal margins

Cite this article as: Takkar B, Molla K, Venkatesh P. Swept-source optical coherence tomography of an optic disc melanocytoma: The importance of the hyperreflective foci. Indian J Ophthalmol 2018;66:140-2.

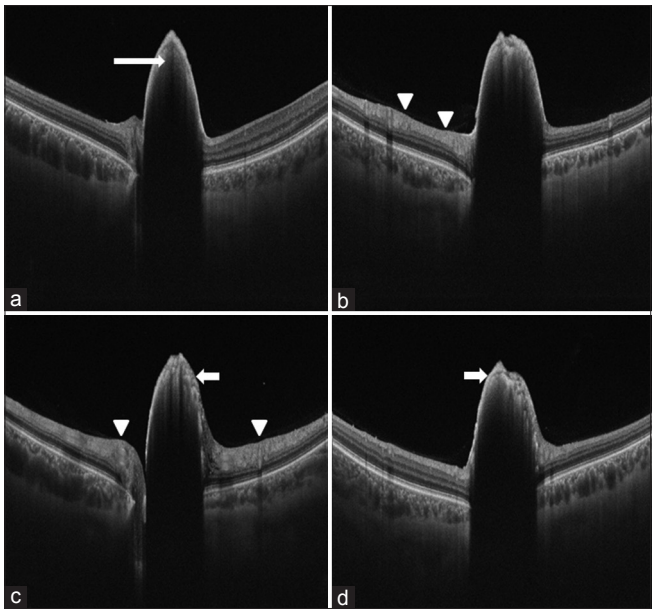

Figure 2: Swept-source optical coherence tomography radial line raster imaging of the mass. (a) The mass is seen as a thumb such as lesion overlying the optic disc with a hyperreflective vitreal surface, smooth moderately reflective internal architecture (arrow) followed by a dense optical shadow. (b) The mass has superficial nodules toward the vitreal side. Note that the choroidal and outer retinal architecture are well-maintained, but the retinal nerve fiber layer appears thickened (arrowheads). (c) Optical coherence tomography image showing scattered hyperreflective dots within the mass. The retinal nerve fiber layer appears thickened (arrowheads). (d) The junction between the mass and the overlying retinal layers is clearly visible (arrow)

SSOCT radial line raster imaging (Topcon DRI OCT-1, Topcon, Japan) revealed an oblong thumb-shaped lesion over the optic disc. The lesion had a densely hyperreflective vitreal border that was irregular and nodular [Fig. 2]. The overlying retinal nerve fiber layer appeared compressed with localized evidence of neuronal edema. However, the internal structure of the ODM appeared largely smooth and moderately reflective, marked by a posterior optical shadow that gave the lesion an appearance of a comet with a tail. There were a few hyperreflective foci within the tumor, and the retinal nerve fiber layer was thickened [Fig. 2]. There was no apparent vitreous seeding, and the surrounding choroidal structure was well-maintained. Specifically, there was no evidence of tumor spread or other choroidal lesions. The SSOCT angiography of the optic nerve head region revealed a superficial meshwork of vasculature over the mass. This could have been the dilated epipapillary vascular plexus overlying the tumor as they were imaged in the superficial OCT slab [Fig. 3]. On en-face OCT projection, there was a ladybug-like appearance of the mass with "black dots seen on the round body of the insect" [Fig. 3].

The patient was reassured of the usually benign nature of ODM but was advised follow-up imaging due to rare reports of enlargement and malignancy.[3]

Discussion

Before the observations of Zimmerman and Garron in 1962,[2] ODM was essentially treated as a malignant tumor.

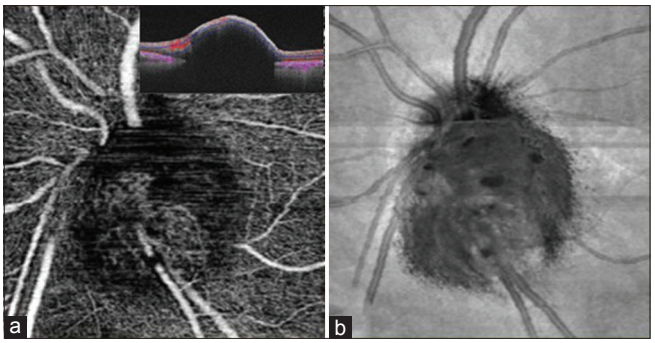

Figure 3: Swept-source optical coherence tomography angiography images of the optic nerve head region scanned over an area of 4.5 mm × 4.5 mm. (a) The imaging slab is set at 130 um below the surface. A meshwork of thin vessels, measuring 1,961,323 μm^2 in the area, can be seen at this level. (b) The en-face optical coherence tomography projection shows the mass simulating a ladybird due to its strategic relationship with retinal vessels and the optic disc

However, they proved the tumor to differ from melanoma because of its uniform internal microscopic structure. In addition, the cellular mass was described to contain "a maximum of melanin" along with other features of a benign tumor. They believed the malignant ODMs described before their evaluation to actually represent misdiagnosed peripapillary choroidal melanomas. This notwithstanding, there have been very rare reports of malignant transformation of ODMs in the current literature as well.[3,4] Although magnetic resonance imaging has been found to be useful in differentiating benign ODM from otherwise, the gold standard would be histopathology which unfortunately cannot be obtained easily. In this context, OCT is very useful as it offers a noninvasive optical biopsy of the ocular tissue.

ODM has been imaged with conventional OCT systems such as spectral domain, as well as the newer ones such as enhanced depth imaging scanning laser ophthalmoscopy.[5-8] Most of these descriptions mention a highly reflective surface of the mass, vitreous seeds, and choroidal/retinal invasion. Choroidal involvement is seen as hyperreflective structures within the choroid along with an upward displacement of the retinal pigment epithelium (RPE).[5] However, these reports have generally failed to comment on the internal structure due to dense optical shadowing and have labeled the tumor interna as an optically empty space.[5-8]

In our case, imaging with SSOCT could decipher some of the tumor's internal characteristics. The tumor was seen to have anterior nodularity and seemed to slope much more than described before on spectral domain OCT.[8] These findings have been noted previously with SSOCT.[9] The most interesting finding, however, is of the bright hyperreflective dots seen within the tumor [Fig. 2]. These could be due to either collections of densely aggregated tumor cells or due to melanophages[9] as noted previously. Zhang *et al*. have defined peripapillary choroidal tumor invasion on OCT as "thickening, hyperreflectivity, or hyporeflectivity of the choroid" (as discussed above). The real nature of these hyperreflective foci is doubtful at present. However, a mass noted predominantly in the choroid on OCT should alert the surgeon toward the possibility of the more dangerous juxtapapillary choroidal melanoma. Unfortunately,

autofluorescence imaging was not done for this case, as it may have helped in understanding the reason behind these foci.

Conclusion

SSOCT may be advantageous over the conventional systems in deciphering the internal structure of an ODM. With further documentation, it is likely to have a role in the follow-up of lesions where benign nature is uncertain.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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