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CASE REPORT

Optical coherence tomography angiography findings of an optic disc melanocytoma in a glaucoma eye

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Abstract
Purpose To determine the optical coherence tomography angiography (OCTA) characteristics of a case of optic disc melanocytoma (ODM) associated with glaucomatous visual field and retinal nerve fiber layer (RNFL) defects in normal tension glaucoma.
Methods The left eye of a 37-year-old female patient followed for a stable ODM for 10 years was investigated with OCT, OCTA, fluorescein (FA), and indocyanine green (ICGA) angiography. The ODM was unchanged, but a previously unknown inferotemporal neuroretinal rim loss and inferotemporal and superotemporal wedge shape glaucomatous RNFL thinning were seen with corresponding glaucomatous visual field defects. The intraocular pressure was 12 mmHg without treatment.
Results In the area of the ODM, FA showed minimal vasculature, and week staining in the late phase, while ICGA showed no signal. In contrast, OCTA showed a dense vasculature in both the superficial and deep layers of the melanocytoma, which was clearly separated from the capillaries of the peripapillary retina. OCTA also showed reduced peripapillary perfusion in the areas of the glaucomatous RNFL bundle defects.
Conclusions In the presented case of a stable ODM and newly detected normal tension glaucoma, OCTA provided more information on perfusion than FA and ICGA which are limited by the heavy pigmentation of the ODM. OCTA also showed a similarly decreased capillary perfusion in both RNFL bundle defects suggesting that the structural damage was related to glaucoma and not compression by ODM. These results suggest that OCTA may be a method preferred over conventional angiography in ODM cases.

Keywords Optic disc melanocytoma · OCT angiography · Glaucoma · Retinal nerve fiber layer · Optical coherence tomography

Introduction

Optic disc melanocytoma (ODM) is a heavily pigmented non-malignant melanocytic nevus, which typically occurs in the area of the optic disc [1]. In some cases, it is contiguous to the adjacent retina or choroid [1]. Regarding the clinical course, ODMs typically remain unchanged over time and cause few

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complications. However, subtle enlargement over the span of several years has been found in 10–15% of cases, and malignant transformation is estimated in 1–2% [2, 3]. Any part of the optic nerve head can be involved [1]. Compression by ODM may result in ischemic and compressive axonal loss which may cause clinically significant retinal nerve fiber layer (RNFL) damage and visual field defect [2, 4, 5]. The compressive RNFL loss visual field defect can be similar to those caused by glaucoma, a common cause of nerve fiber type visual field deterioration [2, 4]. ODM also needs to be differentiated from malignant melanoma of the optic nerve head and choroidal nevus. Malignant melanoma of the optic nerve head, in contrast to ODM, may propagate along the optic nerve without any obvious change detectable with funduscopy. ODMs demonstrate low signal intensity during all phases of fluorescein angiography (FA) and indocyanine green angiography (ICGA) [6]. These invasive angiography methods, however, have recently been increasingly substituted by noninvasive optical coherence tomography angiography (OCTA) in various diseases [7].

As far as we know, there is only one case report, in which OCTA characteristics of an ODM were investigated [7]. In that single case report an otherwise healthy eye was investigated with optical coherence tomography (OCT) and OCTA, thus investigation of any RNFL bundle type structural and functional defect was not possible. In the current case report, we present the clinical appearance, visual field, OCT, OCTA, FA, and ICGA findings of a newly detected normal tension glaucoma eye with a large and stable superior ODM, where the RNFL losses and visual field defects reflect both disc areas involved and uninvolved in the ODM.

Case presentation

A 37-year-old Japanese woman was referred to Kyorin university hospital for evaluation of her known left eye ODM in February 2017. The patient was diagnosed with ODM more than 10 years earlier. At presentation, her visual acuity was 20/20 in both eyes, and the refractive error was − 1.5 D OD and − 1.75 D OS. Her intraocular pressure was 12 mmHg on both eyes with no medication. No relative afferent pupillary defect was seen, and the pupillary light reflex was normal. Both eyes were normal on slit lamp examination. On dilated fundus examination, the right eye showed no abnormality. On the left eye, the ODM appeared as a prominent and heavily pigmented tissue covering the superior, superotemporal, and superonasal sectors of the disc (Fig. 1a). While the size and shape of the melanocytoma did not change compared to the those seen on the disc photos taken 10 years earlier, an inferior neuroretinal rim loss and a wedge shape superotemporal and inferotemporal RNFL defect suggestive for open-angle glaucoma and not visible on the 10-year old photos were detected. Ultrasonography of the orbit revealed no optic nerve involvement, thus conversion of the ODM into a melanoma was excluded. Since in our case the pigmented lesion was located in the area of the optic disc, the choroidal nevus was excluded.

The OCT examination (Cirrus high definition-OCT, Carl Zeiss Meditec Inc., Dublin, CA, USA) revealed significant superotemporal and inferotemporal RNFL thinning (Fig. 1b), and an extreme tissue thickening corresponding to the location of the melanocytoma (Fig. 1b, c). A spatially corresponding macular ganglion cell-inner plexiform layer (GC-IPL) thinning was also found (Fig. 1d). The Humphrey 30-2 visual field showed an inferior nasal step corresponding to the superior RNFL defect, and a mild superior arcuate sensitivity loss corresponding to the inferotemporal RNFL defect (Fig. 1e). The visual field defects were further confirmed with the result of the Humphrey 10-2 test. Based on the visual field results and the structural alterations, and the normal intraocular pressure the diagnosis of normal tension glaucoma of the left eye was set.

FA showed hypofluorescence in the total area of the melanocytoma in the early phase and a mild leakage in the late phase (Fig. 2a, b). ICGA showed no vessels and leakage in the area of the ODM (Fig. 2c). The OCTA was made using a swept source (SS)-OCTA (PLEX Elite, Carl Zeiss Meditec Inc., Dublin, CA, USA), which utilizes a wavelength of 1050 nm (1000–1100 nm full bandwidth), operates with a speed of 100,000 A-lines per second and an axial and lateral resolutions of approximately 5 µm in tissues and approximately 14 µm on the retinal surface [8]. Visualization of the retinal and choroidal vasculature from the volumetric datasets is achieved using a method known as optical microangiography, based on the complex OCT signal ($OMAG^C$). The $OMAG^C$ algorithm incorporates variations in both the intensity

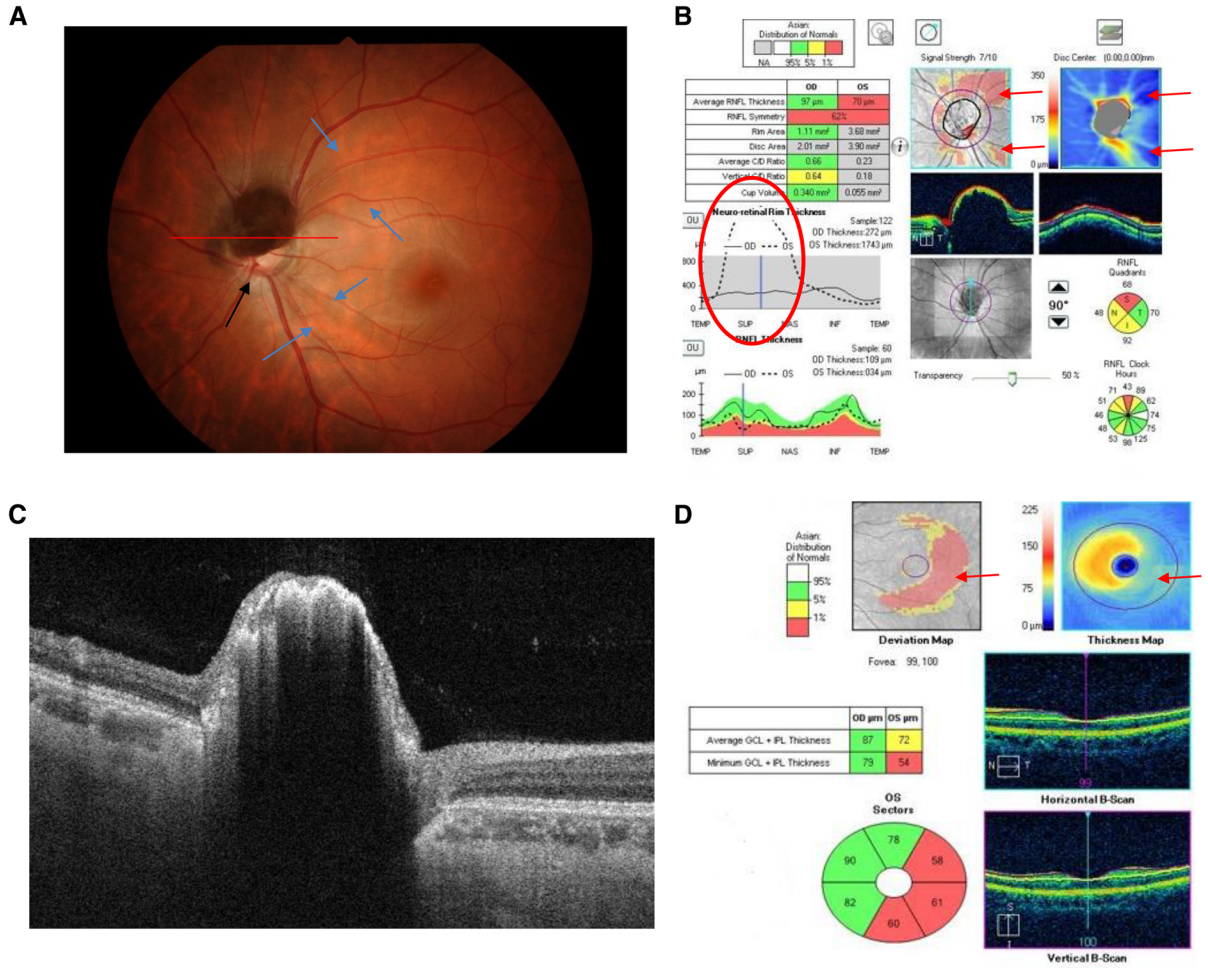

Fig. 1 Fundus photograph (**a**), optical coherence tomography findings (**b**–**d**) and visual field alterations (**e**) of the left eye. **a** The fundus photograph shows an extensive optic disc melanocytoma covering the superonasal, superior, and superotemporal areas; an inferior neuroretinal rim loss (black arrow); and wedge shape superotemporal and inferotemporal retinal nerve fiber layer defects (blue arrows). Swept-source optical coherence tomography scan direction for **c** is shown with the red line. **b** Increased tissue thickness corresponding to the location of the optic disc melanocytoma (red circle); retinal nerve fiber layer defects (red arrows). **c** The optic disc melanocytoma appears as an oval mass adjacent to the optic nerve. **d** Decreased macular ganglion cell-inner plexiform layer thickness (red arrows). **e** Inferior nasal step and superior paracentral defect on Humphrey 30-2 visual field

and phase information between sequential B-scans at the same position to generate the flow information. To detect information on vascular flow, the algorithm generates images based on the motion signals between the repeated OCT B-scans at the same positions. These signals, which reflect subtle differences between B-scans at the same positions, are mostly due to the movement of erythrocytes within blood vessels [9]. The ODM showed an extensive vasculature both in the superficial layer close the vitreous body (Fig. 2d) and the deep tissue layer (Fig. 2e). The vasculature (perfusion) of the tumor was clearly separated from the radial peripapillary capillaries of the RNFL. A clearly visible reduction of vasculature in the areas of the wedge shape RNFL defects was also seen.

Discussion

In the current case report, we presented a newly detected normal tension glaucoma eye in which an extensive ODM covering the superior part of the optic

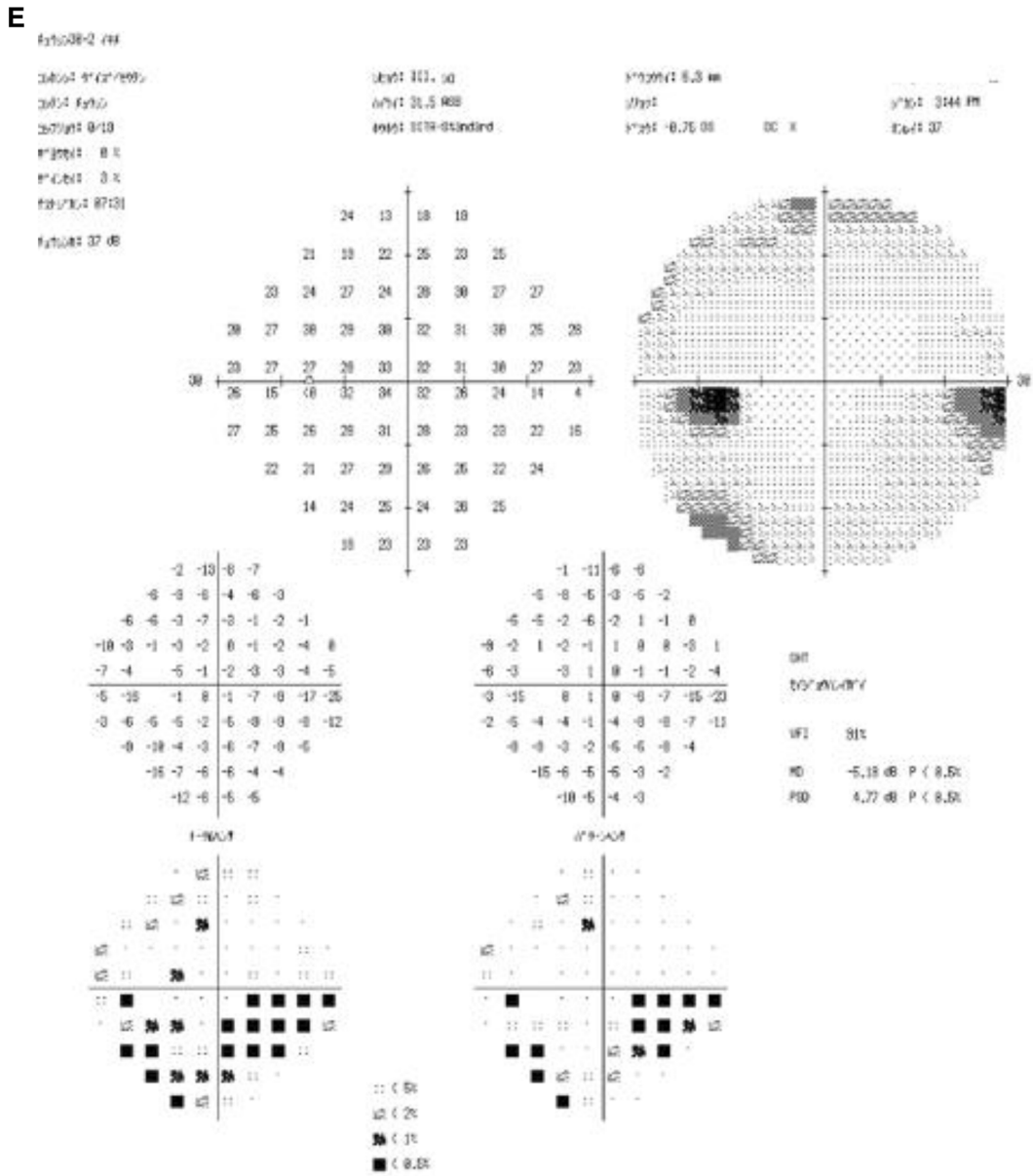

Fig. 1 continued

nerve head was found 10 years prior to the current investigation. While the ODM remained unchanged based on disc photography, normal tension glaucoma with typical superotemporal and inferotemporal wedge shape RNFL damages and spatially corresponding inferior and superior glaucomatous visual field defects developed. The normal tension glaucoma remained unrecognized prior to the current detailed investigation.

Since ODM is able to cause compressive RNFL defects and visual field deterioration [4, 5] which may be similar to those caused open-angle glaucoma, in the current case we had to answer two questions. First, is there any sign of malignant transformation of the melanocytoma; second, can the RNFL loss and visual field deterioration be attributed to the effect of the ODM. As expected FA and ICGA failed to provide clear information on the vasculature of the ODM due

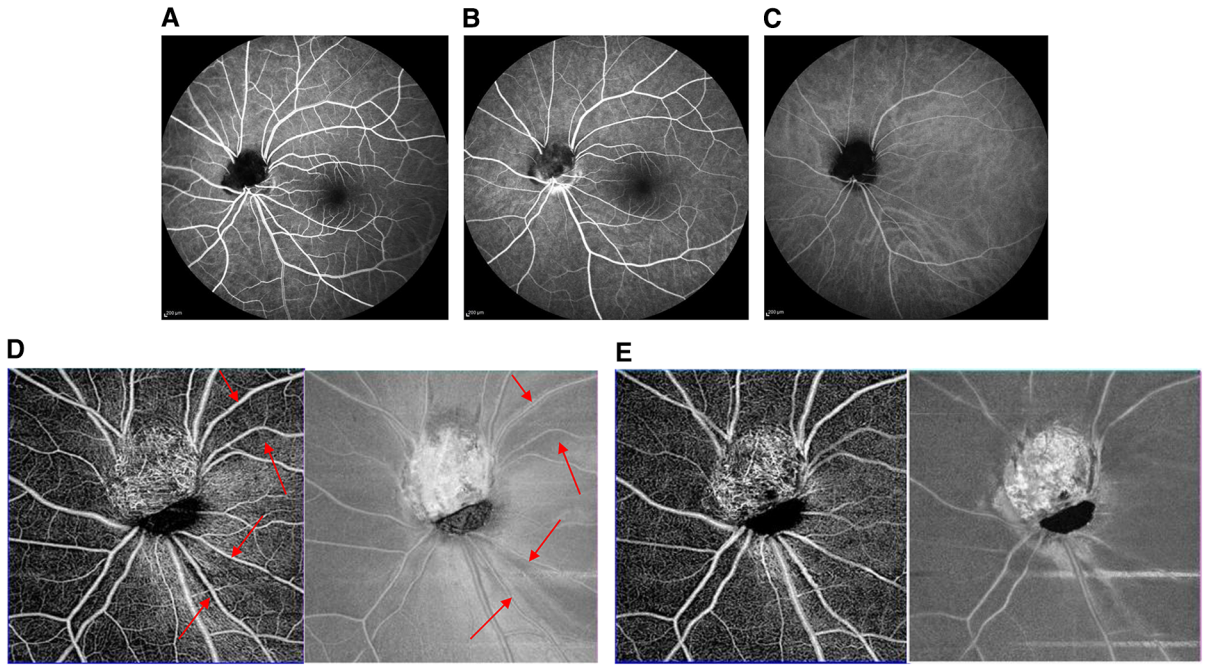

Fig. 2 Fluorescein angiography findings (**a** early, **b** late phase), indocyanine green angiography findings (**c** late phase), optical coherence tomography angiography (OCTA) image and the corresponding en-face structural image from the retinal nerve fiber layer (**d**) and the deep retinal layer (**e**). In contrast to the limited information provided by fluorescein and indocyanine green angiography, on the OCTA image, the retinal nerve fiber defects and the corresponding areas of decreased perfusion are clearly visible (red arrows) in the retinal nerve fiber layer

to its heavy pigmentation which blocks light penetration. OCTA, in contrast, is not influenced by pigmentation since the signal is derived from the movement of the red blood cells [9]. In fact, the vasculature of the melanocytoma was clearly shown both in the superficial and deep layers, and its separation from that of the surrounding tissues and uniformity suggested no malignant changes [10]. These findings were similar to those shown in a recent ODM case reported on an otherwise healthy eye [7]. It is important to note that in the current case report we used a different OCTA method (SS-OCTA) compared to the other case report in which the Angiovue OCTA (Optovue, Fermont, CA, USA) was used. As far as we know, no further publication on OCTA in ODM was published. The fact that different OCTA methods show similar information suggests that various OCTA systems may be usefully used in this condition, which is of clinical significance.

Regarding the cause of the RNFL bundle defects, OCTA also provided useful information. The normal capillary structure seen in the RNFL around the ODM suggested no compression. Further, the similarity of the decrease of capillary perfusion in the wedge shape areas of the superotemporal RNFL bundle defect (spatially corresponding to a disc area involved in the ODM) and inferotemporal RNFL bundle defect (unrelated to the ODM but spatially corresponding to a typical glaucomatous neuroretinal rim loss) was suggestive for open-angle glaucoma. This made it possible to introduce a careful follow-up with regular visual field examinations and the potential of introduction of intraocular pressure lowering treatment if glaucomatous progression is detected. However, RNFL bundle defects may also occur in superior segmental optic hypoplasia and optic disc pit, thus these entities must be considered for differential diagnosis [11, 12].

In conclusion, OCTA, as a part of the complex examination, seems to be a promising method for the investigation of ODMs and may substitute FA and ICGA, which are invasive and at the same time provide only limited information on non-malignant and heavily pigmented alterations.

Compliance with ethical standards

Conflict of interest Gábor Holló is a consultant of Optovue Inc and Zeiss. The other authors declare no conflict of interest.

Research involving human participants and/or animals The Kyorin University Hospital Institutional Review Board for Human Research approved the study protocol, and the study conduct adhered to the tenets of the Declaration of Helsinki.

Informed consent Informed consent was obtained from all individual participants included in the study.

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