

Nevus of Ota with ipsilateral optic disc pigmentation and pigmentary glaucoma

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Key words: Melanocytic Nevi, open-angle glaucoma, trabecular meshwork

A 17-year-old girl from rural Bihar presented with left-sided discoloration of periocular skin since birth [Fig. 1a and b], occasional headache, and rapidly progressive diminution of vision in the left eye (LE) for past 20 days. Uncorrected visual acuity was 6/18 in the right eye (RE) and 6/600 in LE. On examination, there was mild left-sided periocular hyperpigmentation (Tanino's type 1A), LE temporal and inferior scleral pigmentation [Fig. 1c], Krukenberg spindle [Fig. 1d], iris heterochromia [Fig. 2a and b], anisocoria, grade 3 relative afferent pupillary defect, and raised intraocular pressure (IOP) of 38 mmHg. Gonioscopy showed open angles, Sampaolesi's line [Fig. 2c], and increased pigmentation of trabecular meshwork [Fig. 2d]. Anterior segment optical coherence tomography showed central corneal thickness of 496 μ m [Fig. 2e], open angles [Fig. 2f], and flattened iris contour in the region of hyperpigmentation [Fig. 2g]. Fundus showed marked cupping of ipsilateral optic disc with intense pigmentation [Fig. 2h]. Automated perimetry could not be performed due to poor visual acuity. The RE was normal.

IOP was controlled to 14 mmHg by intravenous mannitol followed by maximal topical antiglaucoma therapy. She declined consent for trabeculectomy.

Discussion

Nevus of Ota (oculodermal melanocytosis) is a rare benign developmental disorder of the neural crest cells characterized by melanocytic hamartomas, which can affect all ocular tissues derived from this embryonic structure.^[1] It is most frequent in Asians with a prevalence of 0.014%–0.034%, with women

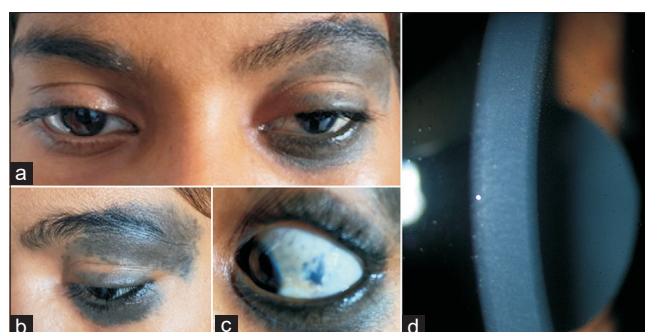


Figure 1: (a) Left-sided periocular hyperpigmentation (Tanino's type 1A). (b) Temporal extent of periocular pigmentation. (c) Temporal scleral pigmentation. (d) Krukenberg spindle

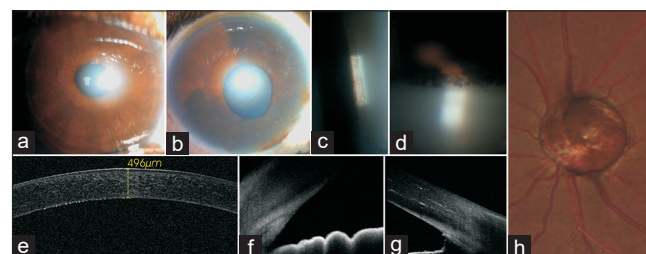


Figure 2: (a) Right eye showing normal iris. (b) Left eye showing iris hyperpigmentation across 12 to 8:30 o'clock (heterochromia). (c) Sampaolesi's line as seen on left eye gonioscopy. (d) Intense trabecular pigmentation as seen on left eye gonioscopy. (e) Anterior segment optical coherence tomograph of the left eye showing central corneal thickness of 496 μ m. (f) Anterior segment optical coherence tomograph of the left eye angle at 9 o'clock showing open angle and normal iris contour. (g) Anterior segment optical coherence tomograph of the left eye angle at 3 o'clock showing open angle with flattened iris contour. (h) Fundus photograph left eye showing intense pigmentation of optic disc and cup-disc ratio of 0.9:1

five times more likely to be affected.^[2-4] Raised IOP has been associated in 10.3% and optic disc pigmentation in 18.1% cases with ocular involvement.^[5] Such patients may require early glaucoma filtering surgery and lifelong follow-up with ophthalmologists and dermatologists.^[4]

Our case highlights the importance of screening for open-angle glaucoma in children with seemingly innocuous skin pigmentation and the need to sensitize primary care physicians regarding this condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other

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Cite this article as: Rungta N, Ranjan A, Nishant P, Sinha S. Nevus of Ota with ipsilateral optic disc pigmentation and pigmentary glaucoma. Indian J Ophthalmol 2022;70:2656-7.

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/ijo.IJO_2893_21

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Received: 15-Nov-2021

Revision: 06-Feb-2022

Accepted: 10-Feb-2022

Published: 30-Jun-2022

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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