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Ocular melanocytosis and secondary glaucoma in a child

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Abstract

Glaucoma at young age is an unusual presentation of congenital ocular melanocytosis. We report a young patient with a unilateral splinter disc hemorrhage, mild asymmetric optic nerve cupping, and increased intraocular pressures, suggestive of glaucoma, associated with bilateral ocular melanocytosis. While glaucoma and/or increased intraocular pressures have been reported in oculodermal melanocytosis in the adult population, there are only rare reports of congenital ocular melanocytosis associated with secondary glaucoma in a child. Glaucoma is believed to result from melanocytic involvement of the anterior chamber angle and trabecular meshwork with resultant obstruction of outflow.

Introduction

Ocular melanocytosis, a melanocytic hyperpigmentation of the episclera and uvea, is a variant of oculodermal melanocystosis in which the melanocytosis is limited to the globe. When there is melanocytic skin involvement of the ophthalmic branch of the trigeminal nerve, it is referred to as oculodermal melanocytosis, or nevus of Ota, after the Japanese dermatologist Masao Ota. Dermal involvement may occur without ophthalmic involvement, and may occur in the distribution of the ophthalmic, maxillary, and occasionally mandibular branch of the trigeminal nerve. The melanocytosis is caused by increased fusiform dendritic melanocytes, which may be found in the uvea and episclera, and may, on occasion, extend to the anterior chamber angle and trabecular meshwork.[1] We present a case of ocular melanocytosis with glaucoma in a child.

Case Report

A The patient, a 6 year-old Malaysian-Indian boy, was referred to ophthalmology by his pediatrician after noting "dirty" appearing pigmentation of sclera bilaterally. He had a history of small stature and a ligated patent ductus arteriosus. On examination, the patient was able to fixate and follow with each eye. Extraocular movements were full and orthophoric and pupils with equally reactive without any afferent pupillary defect. There was no proptosis. Also, there was no hyperpigmentation of facial skin. However slit-lamp examination revealed patchy, slate grey discoloration of both temporal episclera that did not move with the conjunctiva and remained fixed to the globe (Figure 1). The conjunctivae of each eye was otherwise unremarkable. The corneas were clear bilaterally and the anterior chambers were deep and quiet in each eye. The iris in each eye was normal without any pigmentation. The lenses were clear bilaterally. The patient did not tolerate gonioscopy. Intraocular pressures were 23 and 32 mmHg in the right and left eyes, respectively. Dilated fundoscopy

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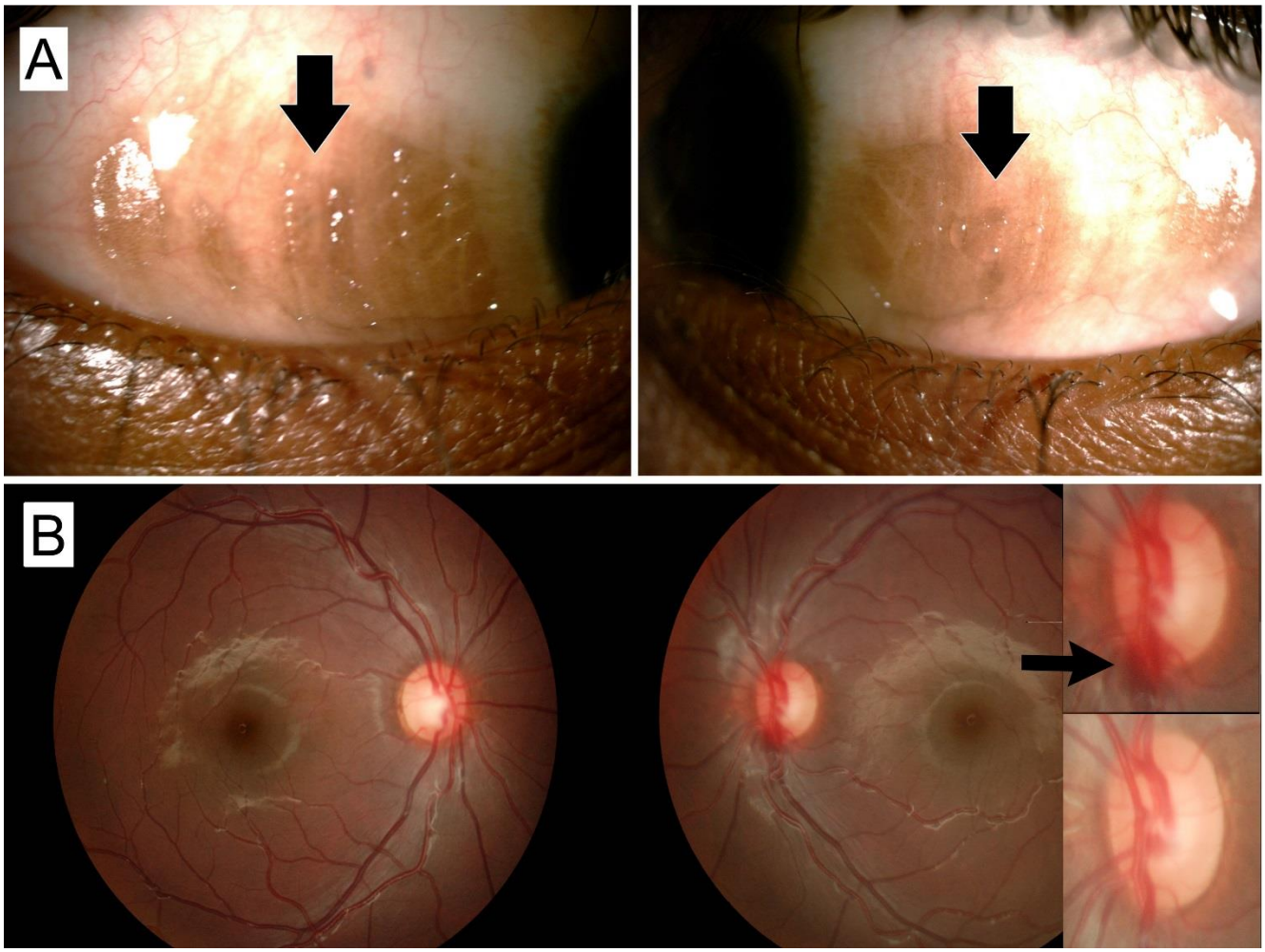

Figure 1

A. *Anterior segment photographs of the patient demonstrate the prominent discoloration (arrow) of the episclera temporal to the limbus in each eye (right eye on left and left eye on right side of image).*
B. *Fundus photographs of the patient demonstrate mild asymmetric cupping of the optic discs (right eye on left and left eye on right side of image). There is a splinter hemorrhage at the inferior margin of the left disc margin (arrow in inset to upper right) which is resolved on a follow up fundus photograph (inset to lower right).*

revealed a clear vitreous and sharp and pink disc in each eye. The cup-to-disc ratio was 0.4 and 0.5 in the right and left eyes, respectively. There was an inferior splinter disc hemorrhage on the left. The nerve fiber layer appeared full in each eye. The patient had sharp foveal reflexes with a normal macula and periphery in each eye.

A diagnosis of bilateral ocular melanocytosis with secondary glaucoma was made and the patient was started on topical dorzolamide followed by topical timolol and after 4 months his intraocular pressures had decreased to 15 mmHg in each eye. The patient was unable to cooperate with retinal nerve fiber layer / optic nerve head topographic imaging or with automated visual fields. The patient subsequently developed asthmatic symptoms and the timolol was changed to latanoprost, and his intraocular pressures remained in the mid-teens.

The left splinter disc hemorrhage resolved over time, and with over two years of follow-up, the discs remained unchanged with full-appearing nerve fiber layers in each eye.

Discussion

Ocular melanocytosis is a melanocytic hyperplasia that is believed to be a result of the incomplete migration of melanocytes from the neural crest to the surface layers. Familial and hormonal factors may have strong associations with the pathogenesis. Congenital ocular melanocytosis is relatively uncommon condition and very rarely associated with glaucoma in children. In a review of 194 patients with oculodermal melanocytosis, all patients demonstrated episcleral involvement and nearly all patients (87%) demonstrated iris involvement manifest as heterochromia iridis. Iris involvement was very frequently associated with anterior chamber involvement (83% of patients). A substantial percentage of the patients (79%) demonstrated choroidal hyperpigmentation, particularly when there was iris involvement. Less commonly, the authors found involvement of the conjunctiva, tarsus, cornea, lens, and optic nerve head.[1]

While glaucoma and/or increased intraocular pressures have been reported in oculodermal melanocytosis in approximately 10% of cases in the adult population,[1] there are only rare reports of congenital ocular melanocytosis associated with secondary glaucoma in a child. The mechanism of the glaucoma in ocular melanocytosis is believed to result from melanocytosis of the anterior chamber angle and trabecular meshwork, resulting in obstruction of outflow.[2] Goncalves, et al. reported a case of a boy with ocular melanocytosis and advanced glaucomatous damage.[3] Duke, et al. reported a case of a boy with oculodermal melanocytosis, a relative afferent pupillary defect, and glaucomatous optic disc changes.[2] Lee, et al. reported a case of a boy with oculodermal melanocytosis and concurrent Sturge-Weber syndrome with associated glaucoma.[4]

Ocular complications of oculodermal melanocytosis includes increased intraocular pressure with or without glaucoma was the most common (10.3%), followed by asymmetric cupping of the optic nerve head that was not associated with glaucoma (9.8%), uveitis (2.6%), cataracts (1%), and, rarely, orbital melanoma (0.5%).[1] The occurrence of glaucoma in patients with the oculodermal melanocytosis may be related to the obstruction of aqueous outflow by accumulated melanocytes; however, some studies have reported that the oculodermal melanocytosis alone cannot cause the secondary glaucoma.[5]

In our case, the patient had mild asymmetric cupping of the optic nerve head and a unilateral splinter disc hemorrhage. Splinter disc hemorrhages are typically a feature of glaucomatous discs, usually small flame or splinter shaped, and more common in the inferotemporal quadrant.[6] Furthermore, importantly, optic disc hemorrhage may occur even before neural rim notching, retinal nerve layer defects, or visual field defect.[7] Although optic disc hemorrhage has been reported as an important risk factor for glaucoma progression, the majority of eyes that developed a disc hemorrhage in the Ocular Hypertension Treatment Study have not developed a primary open angle glaucoma endpoint to date.[8] While disc hemorrhages may occur in a variety of ocular and systemic conditions, in our case, the combination of the splinter disc hemorrhage, optic nerve cupping, and increased intraocular pressures were suggestive of glaucoma.

Conclusion

Congenital ocular melanocytosis in children, may pose a significant diagnostic and therapeutic challenge even to the ophthalmologist. Both parents and physicians are often unaware of oculodermal melanocytosis and may mistake it for other benign pigmented lesions of the conjunctiva or sclera, such as benign epithelial melanosis, primary acquired melanosis, or conjunctival nevi. While parents are often mainly concerned about the cosmetic appearance of oculodermal melanocytosis, glaucoma and ocular melanoma are serious complications which may require lifelong ophthalmology monitoring.

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