Niger J Paed 2013; 40 (2): 198 –200

CASE REPORT

Duke R
David R
Utam U

Secondary open angle glaucoma in a child with congenital oculodermal melanocytosis

DOI:http://dx.doi.org/10.4314/njp.v40i2,20

Accepted: 25th September 2012

Duke R (✉)
David R, Utam U
Department of Ophthalmology, Pediatric Ophthalmology & Strabismus Unit, University of Calabar College of Medical Sciences, Faculty of Clinical Sciences

POBox 910 Calabar, Post code 540001, Cross River State, Nigeria
Email: dr.roselineduke@gmail.com
Tel: +2348061302925

Abstract: Congenital oculodermal melanocytosis, a pigmented ocular surface lesion in children, may pose a significant diagnostic and therapeutic challenge even to the eye physician. Further, parents, care givers and non-eye care physicians are often unaware of this diagnosis and its associated potential ocular morbid conditions or the potential for malignancy thereby causing a delay in examination of such affected eyes. This article highlights an uncommon pigmented ocular surface lesion encountered in childhood. Further, a call to attention, that congenital oculodermal melanosis in childhood may harbor and coexist with an ocular morbid condition of glaucoma which develops in childhood. We report a case of congenital oculodermal melanosis with secondary open angle glaucoma in childhood. Special emphasis is placed on early identification and referral by non eye physicians. Accurate diagnosis and management of congenital oculodermal melanosis and the associated condition of secondary open angle glaucoma in childhood by the eye care physician is important. An understanding of the natural history of pigmented eye lesions in children by non eye physicians is essential for successful outcomes and prevention of uniocular blindness and its attendant consequences.

Key words: oculodermal melanocytosis; unilateral; secondary glaucoma; blindness

Introduction

Ocular surface lesions of which pigmented or melanocytic lesions including congenital oculodermal melanocytosis, are uncommon, even in dark pigmented patients.[1,2] When uncomplicated, they rarely are associated with symptoms, therefore, may not be given adequate and due attention by parents, care givers or physicians.

Pigmented lesions of the conjunctiva, episclera and the sclera arise from either melanocytes or nonmelanocytes and have a diverse differential diagnosis. These lesions can be classified into congenital melanosis or melanocytosis, conjunctival nevi, acquire melanosis (secondary or primary), and conjunctival melanomas. In secondary acquired melanosis, the increased conjunctival pigmentation may be caused by irradiation, hormonal changes, chemical irritation or chronic inflammatory conjunctival disorders such a severe vernal conjunctivitis in children.[4,5,6]

While some pigmented lesions remain innocuous others have malignant potentials or may be associated with ocular morbid conditions. Some of these pigmented ocular surface lesions are congenital in origin and as such present from birth; however, may become more prominent at or after puberty.[7]

We report a case of congenital oculodermal melanocytosis associated with secondary open angle glaucoma, starting in childhood.[8] Further, we seek to highlight the need for early referral to the eye care physicians from non-eye physicians, when apparently benign pigmented lesions are seen in children.

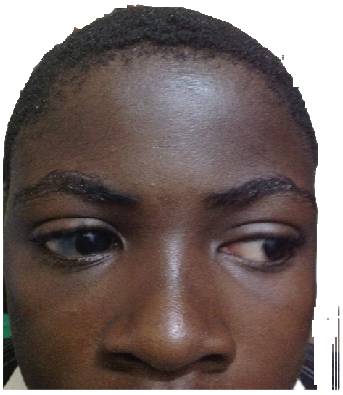
Case report, site of involvement and clinical features:

IK an 11 year old male student, presented to the eye clinic of our institution with a complaint of blurring of distant vision in both eyes in the last two weeks. A non-progressive grey discoloration of the white part of his right eye was noticed from birth, with associated gradual increase in discoloration of the lower eyelid. However, he had been admitted into the children emergency clinic of the hospital on two occasions, the first on account of malaria and the second on account of pneumonia when he was two years and seven years old respectively. There is no family history of ocular or dermal pigmentation.

On general examination, a healthy looking young boy was seen. On ocular examination, visual acuities were normal with 6/6 in both eyes. Objective automatic refraction in the right and left eye revealed hyperopic astigmatism with the subjective best correction resulting in visual acuity of 6/5 and N5 at 33cm in both eyes.

The left eye and eye lid was normal. In the right eye lid, there was a grey pigmented area involving the lower lid, about 5cm from the medial angle. This was associated with a flat, slate grey, avascular, multifocal lesion in the white part of the external eye,which did not move, measuring 7-10mm and extending inferiorly and superiorly. See figure 1. The anterior segment including the aqueous drainage angle and structures were normal, however with increased hyper pigmentation of the iris and trabecular meshwork . Pupillary examination showed a subtle relative afferent pupillary defect in the right eye. Stereoscopic examination of the right optic disc revealed a cup disc ratio of 0.5 cupping, pallor, notching with a disc symmetry of 0.2 decrease in the left optic disc which appeared normal in size and contour, compared to the right. All other posterior segment structures were normal. Intraocular pressures on the right and left eyes were 13 and 11 millimeters of mercury (mmHg) respectively taken at the first visit. Axial length measurement of both eyes, with an A scan using IOL master revealed in the right 24.31mm and on the left 24.21mm respectively. Using the Zeiss Frequency Doubling Technology, his first visual fields revealed a mean standard deviation of -16.12 dB P<0.5 on the right and -7.75 dB P<25 on the left, however with 20% fixation errors. Optical coherence tomography confirmed a disc asymmetry with loss of nerve fibers, larger on the right with contour neuroretinal rim changes in the right eye. Optic disc pictures were taken for documentation and monitoring.

Fig 1: A child with right oculodermal melanocytosis



A diagnosis of a mild astigmatic refractive error, right oculodermal melanocytosis associated with right secondary open angle glaucoma was made.
The patient was given a pair of corrective spectacles. A CT scan of the brain and orbits were requested as well as a follow up visit.

Clinical progress of patient since diagnosis:

With the use of the spectacles, the patient was satisfied with his class work. The CT scan result of the brain and orbits was normal. Intraocular pressures was 12 and 10mmhg respectively at the second visit.
Right laser trabeculoplasty was recommended, the parents however, declined laser surgery. He was placed on Timolo.5%, a beta blocker 12 hourly, in the mean time and will be monitored for life.

Discussion

Congenital oculodermal melanocytosis also known as Naevus of Ota, is a relatively uncommon condition, however, the most frequent form of ocular melanocytosis seen. It is characterized by unilateral patchy but extensive slate grey or bluish discoloration of the sclera with associated pigmentation of the periocular skin.[2,8] This condition is seen more commonly in Blacks and Asians. Histologically a focal proliferation and increase in number, size and pigmentation of melanocytes is seen in affected eyes and skin. From a prognostic point of view, it is important to examine and follow up the posterior segment of the eye of these patients with nevus of Ota, as the incidence of uveal melanoma is 0.4% (1/400 cases) in adult caucasians.[9] Further, these patients have a slightly higher risk to develop orbital, and meningeal melanoma which is seen most commonly in whites. Oculodermal melanocytosis may pose a risk for the developmental and progression of glaucoma. Secondary glaucoma in this patients, is associated with the condition in less than 10% of cases and usually presents in adulthood with blindness.[3,8] Glaucoma is an optic neuropathy, that leads to characteristic central visual field loss and optic never damage resulting in blindness if left unidentified and treated. The pathogenesis of glaucoma in congenital oculodermal melanocytosis is not well understood, but it is suggested that hyper pigmentation of the trabecular meshwork as well as clogging of the meshwork outflow may be responsible for secondary glaucoma.[8,10] Glaucoma management in childhood is a challenging condition which requires referral to a tertiary eye care center and more recently sub specialist care.[11] The goal of preserving a lifetime of a vision for such children involves early, prompt control of the intraocular pressure, correction of refractive error and rigorous amblyopia therapy depending on the age of the child. Control of intraocular pressure is primarily surgical with medical therapy playing a supportive role or as an adjunct in blacks, ensuring the prevention of uniocular loss of visual fields in adolescent and blindness in adulthood.[11, 12,13]

Opportunities to identify patients with congenital oculodermal melanocytosis possibly lies primarily with the non eye physician. The need to be aware of such high risk patients, health educate and encourage referrals for documentation, monitoring as well as intervention when necessary, cannot be over emphasized.

Conclusion

Congenital oculodermal melanocytosis is uncommon and appears harmless, but may be associated with the development of secondary open angle glaucoma in childhood. A practical approach to managing children with this condition by the non eye physicians, would entail early identification and referral to the eye physicians.

Authors contribution

Duke R: Conceived the study, performed the literature search , prepared and reviewed the manuscript.

David R: Involved in the management of the patient and manuscript review.

Utam U: Involved in the management of the patient and manuscript review.

Conflict of interest: None

Funding: None

References

1. Teekhasaenee C, Ritch R, Rutnin U, Leelawongs N. Ocular findings in oculodermal melanocytosis. *Arch Ophthalmol 1990;108:1114-20*
2. Kanski J, Bowling B. Clinical ophthalmology: a systematic approach. 7th ed. Edinburgh: *Elsevier limited; 2011.p.263-5*
3. Chodosh J, Burris R. Ocular Melanocytosis. *N Engl J Med 2004;350:809*
4. Liesegang T J. Pigmented conjunctival and scleral lesions. *Mayo Clin Proc1994;69(2):151-61*
5. Othman S I. Ocular surface tumors. *Oman J Ophthamol 2009;2 (1):3-14*
6. Dahan E, Appel R. Vernal conjunctivitis in the black child and its response to therapy. *Br J Ophthalmol 1983;67(10):688-92.*
7. Hidano A, Kajima H, Ikeda S, et al. Natural history of nevus of Ota. *Arch Dermatol 1967;95:187–95.*
8. Gonclaves V, Sandler T,O'Daonnell E. Open angle glaucoma in melanosis oculi: a response to laser trabeculoplasty. *Annals of Ophthalmology 1985;17 (1):33-6.*
9. Gonder JR, Shields JA, Albert DM, et al. Uveal malignant melanoma associated with ocular and oculodermal melanocytosis. *Ophthalmology 1982;89:953–60.*
10. Papadopoulos M, Khaw P T. Childhood glaucoma. In: taylor D, Hoyt CS(ed) Pediatric Ophthalmology Strabismus. Elsevier Saunders: Philadelphia, 2005,pp 458-471.
11. Papadopoulos M, Khaw P T.Advances in the management of paediatric glaucoma. *Eye 2007;21:1319-1325*
12. Yeung H H, Walton S D. Clinical classification of childhood glaucomas. *Arch Ophthamlo 2010;128 (6):680-684*
13. Ellong A, Ebana Mvogo C, Nyouma Moune E, Bella-Hiag A. Juvenile glaucoma in Cameroon. *Bull Soc Belge Ophtalmol 2007; (305):69-77.*