Bilateral Nevus of Ota with Secondary Glaucoma: A Rare Case Report

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Abstract

Purpose: To describe a rare case of Nevus of Ota with secondary Glaucoma in Ispahani Islamia Eye Institute and Hospital (IIEH &H). A 5 years 7 months old boy came to this hospital for dimness of vision in left eye. On examination, there was hyperpigmentation of both eyes and surrounding adnexa (L/E > R/E). His BCVA was 6/6 in R/E and 6/24 in L/E. Colour vision was normal in B/E. Iris pigment on anterior lens capsule B/E was found (L/E > R/E). IOP was 12mmHg in R/E and 16mmHg in L/E with anti-glaucoma medication (Timolol and Dorzolamide combination). B-scan report showed axial length and CRC thickness was normal in B/E. In CFP B/E fundus is hyperpigmented(L/E>R/E) and C:D ratio is 0.5:1 in R/E and 0.85:1 in L/E. After adequate counselling, patient was advised for Trabeculectomy with Mitomycin C under GA in L/E. We follow-up the patient for 1st , 7th and 1 month after surgery.

Conclusion: Although bilateral Nevus of Ota is a rare disease, there is a 10% chance for developing Glaucoma. Trabeculectomy with Mitomycin C can decrease the risk of progression of Glaucoma.

Keywords: Nevus of Ota, Secondary Glaucoma.

Introduction

Nevus of Ota is a form of dermal melanocytosis that produces hyperpigmentation of the eye and surrounding adnexa along the V1 and V2 trigeminal nerve distribution. Hyperpigmentation is seen as a bluish or brownish pigmentation of the eyes (conjunctiva, episclera, sclera, cornea, iris, choroid, retina, extraocular muscle, retrobulbar fat and periosteum) and/or in the face skin and lids³. Nevus of Ota extending into the eye have an increased risk of developing secondary glaucoma (10% of patients) as invasion of melanocytes can block drainage of aqueous, leading to elevated IOP³.

Case Summary

A 5.7 years old boy came to the Glaucoma department of IIEI &H on 23rd October, 2023.On examination, there was bluish grey skin

pigmentation on the eye (sclera, conjunctiva) and surrounding adnexa (cheeks and lids) in the both side of the face. ¹BCVA was 6/6 in R/E and 6/24 in L/E. There was compound myopic astigmatism in L/E. His IOP was 12mmHg in R/E and 16mmHg in L/E using Goldmann applanation tonometer. Patient was using 0.5% Timolol and Dorzolamide combination anti glaucoma medication in B/E for last 2 years. Pupil was round, regular and reacting in R/E and sluggish reaction in L/E.On slit lamp examination, there was patchy episcleral pigmentation in B/E1.Iris showed increased pigmentation and loss of stromal crypts. 1 By gonioscopy, all angle structures of B/E were obscured by a heavy amount of pigment.1 CFP shows relative choroidal hyperpigmentation in B/E(L/E>R/E) and C:D ratio was 0.5:1 in R/E and 0.85:1 in L/E.On confrontation test ,there was loss of peripheral vision in L/E.Our impression was Nevus of Ota(B/E) with secondary glaucoma

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(L/E>R/E) with left anisometropic amblyopia. We have advised the patient for Trabeculectomy with Mitomycin C under G/A in L/E and continue 0.5% timolol and dorzolamide combination anti glaucoma medication in R/E.Trab+MMC was done on 11th November, 2023.After 7th POD IOP was 4mmHg, after 1-month IOP was 9mmHg. Subsequent follow up will be 4 monthly.



Fig:1 Bilateral Nevus of Ota



Fig:2 Hyperpigmentation of eye and surrounding adnexa



Fig:3 Scleral hyperpigmentation



Fig: 4 CFP shows relative choroidal hyperpigmentation(L/E>R/E)

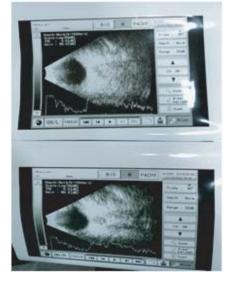


Fig:5 B-Scan (L/E and R/E)



Fig:6 1st POD of Trab+MMC in L/E.

Discussion

Nevus of Ota is a non-hereditary pigmentation disorder which is more frequent in females than male (5:1). It is caused by melanocytes that have not migrated completely from the neural crest to the epidermis during the embryonic phase. Consequently, the melanocytes enter the ophthalmic and maxillary branches of the trigeminal nerve creating spots on the nervous regions.³ It can be associated with various ocular abnormalities such as congenital glaucoma, Duane's syndrome and melanoma.³ When the pigmentation of the chamber angle is intensive there is an increased risk of glaucoma associated with Ota's Nevus and the patient should be referred for periodic measurements of IOP and a complete ophthalmic examination.²

Conclusion

Nevus of Ota is a non-hereditary disorder which may cause secondary glaucoma. Early diagnosis and treatment can prevent blindness. Patient requires periodic examinations by ophthalmologists and dermatologists to detect the complications earlier.

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