INTRODUCTION
Oculodermal melanocytosis was described by Ota and Tanino in 1939. Nevus of Ota is the other name of this rare disorder which affects 0.014 - 0.034% of population in South East Asia. It is more prevalent in Japan, where the incidence is about 0.2 - 0.3%. Unilateral involvement is the rule; however, 10% of cases may be bilateral. The condition manifests as a bluish grey patch in the distribution of 1st and 2nd branches of trigeminal nerve. Ocular manifestation include episcleral melanosis, heterochromia, glaucoma, iris mammillations and iris nevii. The aim to report this case is to familiarise the ophthalmologists and healthcare professionals about the long-term ocular complications that can be associated with this rare disorder.

CASE REPORT
A 26-year male presented with blurred distant vision in left eye for the last 15 days. He had a dark patch over left side of his face, since early childhood, which was noticed by the parents at the age of 2 years. The lesion was initially bluish in colour and gradually became darker over time. There was no history of redness, photophobia, watering, floaters, flashes of light, trauma, ocular medication, eye rubbing or allergies. Family, drug, personal, and socioeconomic history were not contributory.

On general physical examination, multiple mottled bluish grey patches were seen over the left cheek extending to the left temporal area (Figure 1). Visual acuity was 6/6 in right eye while 6/12 improving to 6/6 with -1.00 DS in left eye. Anterior segment examination of left eye revealed episcleral pigmentation and heterochromia secondary to hyperpigmented iris of left eye (Figure 2). Gonioscopy of left eye revealed hyperpigmentation of trabeculum; however, anterior chamber angle was wide open and intraocular pressure was normal in both the eyes. Posterior segment examination of left eye revealed retinal and sub-retinal hyperpigmentation involving posterior pole and extending to mid periphery, while the examination of the right eye did not reveal any abnormality (Figures 3 and 4). Based on the distinct color, morphology, and location of the lesion, the condition was diagnosed as Nevus of Ota.

Figure 1: Left sided Nevus of Ota.
Figure 2: Episcleral hyper pigmentation.
Figure 3: Right fundus.
Figure 4: Left fundus.
Relevant ophthalmologic investigations, like ultrasound B-scan and optical coherence tomography (OCT) were performed which did not reveal any abnormality. Dermatological examination by dermatologist confirmed the diagnosis of Nevus of Ota. Auditory examination by otolaryngologist was also unremarkable. Patient was counselled about the nature of disease, risk of long-term complications, and was advised regular follow-up.

DISCUSSION
Nevus of Ota is rare disorder that affects Asians and blacks more commonly than Whites. It is a unilateral disorder in 90% cases and more common in females with female to male ratio of 5:1. Pathophysiology is still unconfirmed. It is postulated that Nevus of Ota results from defective migration of neural crest cells to the epidermis during embryonic stage. The two peak ages of onset in early infancy and in early adolescence suggest that hormones play a role in the development of this condition.4,5

After its onset, Nevus of Ota may enlarge slowly and progressively and darken in color. Its appearance usually remains stable once adulthood is reached. The color and perception of Nevus of Ota may fluctuate depending on personal and environmental factors such as fatigue, menstruation, insomnia, and cloudy cold and hot weather conditions.6

Ocular associations are present and include involvement of sclera, cornea, anterior chamber angle, iris, retina and optic disc. The two vision-threatening complications are glaucoma and uveal melanoma. The metastasis from uveal melanoma can risk the life of the patient.7

The risk of glaucoma is 10 - 15%. In cases of uveal melanoma, 3% have evidence of Nevus of Ota. Moreover, the risk of metastasis from uveal melanoma is also higher in cases of Nevus of Ota than without preexisting nevus.8,9

The patient we reported here was a young male who had ocular involvement including episcleral pigmentation, hyperpigmentation of anterior chamber angle, hypochromic iris and pigmentation of retina and choroid. There was no involvement of tympanic membrane or oral mucosa. Various topical and laser treatments have been used successfully in treating the cosmetic blemish associated with this condition; however, the risk of ocular complications is always there, thus regular and close follow-up is required.

In summary, although Nevus of Ota is a benign and rare disorder, the ocular association may be vision-threatening and, therefore, a regular and long-term follow-up by an ophthalmologist is necessary.

REFERENCES

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