



## OCULODERMAL MELANOCYTOSIS

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Received for publication: August 13, 2013; Accepted: September 16, 2013

**Abstract:** Oculodermal Melanocytosis (ODM) or naevus fuscocoeruleus ophthalmomaxillaris or Nevus of Ota, a melanocytic hyper pigmentation of the globe and periorbital skin, was described in 1939 by Ota and Tanino. Melanocytic hyperpigmentation involving the globe alone is called ocular melanocytosis, and similar hyperpigmentation involvin the skin alone is called dermal melanocytosis. In Caucasians, there is an association between ODM and uveal melanoma. Most cases of the nevus of Ota are unilateral (90%), although pigmentation is present bilaterally in (5%–10%). Ocular abnormalities included pigmentation of the sclera, cornea, retina, and optic disc and cavernous hemangiomas of the optic disc, elevated intraocular pressure, glaucoma, and ocular melanoma. Malignant melanoma of the uveal tract has been reported to occur in patients with ocular or oculodermal melanocytosis. Therefore, all patients with Oculodermal melanocytosis should have periodic ocular examination to allow early detection of glaucoma and uveal melanoma.

**Keywords:** Oculodermal melanosis, melanoma, Nevus of Ota

## INTRODUCTION

Oculodermal melanocytosis (ODM) or Nevus of Ota is a congenital condition characterized by benign dermal melanosis of the skin in the area innervated by first, second, and rarely third division of the trigeminal nerve. Patients develop ipsilateral increase in pigmentation of the episclera, conjunctiva, uveal tract, and occasionally optic nerve head. Oculodermal melanocytosis was described in 1939 by Ota and Tanino<sup>1</sup>. In Caucasians, there is an association between ODM and uveal melanoma.<sup>2,3</sup> Malignant melanoma of the uveal tract has been reported to occur in patients with ocular or oculodermal melanocytosis.<sup>4</sup> The nevus of Ota primarily affects Asian populations. Other ethnic groups with increased prevalence include Africans, Afro-Americans, and East Indians. It is very uncommon among Caucasian patients. It has been rarely studied, although some studies report an estimated rate of about 0.038% of all the dermatological outpatient cases<sup>5</sup>.

## MATERIALS AND METHODS

### Case report:

A 68-year-old white woman was referred to the Department of ophthalmology for assessment of a increased pigmentation of the left eye and periorbital skin. The patient had been aware of pigmentation in left eye and skin around left eye since early childhood. The family history was unremarkable.

## RESULTS

On examination the patient's visual acuity with correction was 6/60 in both the eyes. External

examination revealed left sided oculodermal melanocytosis (Fig.1). Intraocular pressure by applanation was 14mmHg in both eyes. Slit-lamp examination revealed conjunctival and episcleral pigmentation in left eye and bilateral senile immature nuclear cataract (Fig. 2). Gonioscopy and dilated fundus examination were normal in both eyes.



Figure.1

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**Figure.1:** Photograph of patient with Oculodermal melanocytosis.

\*Note the pigmentation on left globe and dermal pigmentation.



**Figure.2:** Photograph of patient with Oculodermal melanocytosis.

\*Note conjunctival and episcleral pigmentation in left eye.

## DISCUSSION

In patients with ocular and oculodermal melanocytosis the pigmentation may involve the conjunctiva, episclera, the skin of the cheek as illustrated by our case. Other tissues that had been reported to be involved in ocular or oculodermal melanocytosis included the skin of the eyelid, temple, forehead, the orbit, optic disc, uveal tract, tympanic membrane, the orbital and cranial bones, meninges, the pharynx, nasal mucosa, and hard palate.<sup>6</sup> Glaucoma has also been associated with the nevus of Ota in 10% of patients.<sup>7</sup> Oculodermal melanocytosis is 4 times more prevalent in women than men. The pigmentation is congenital in 60% of persons, while others developed the pigmentation at puberty or during pregnancy.<sup>6</sup> In addition ocular and oculodermal melanocytosis have been associated with choroidal malignant melanoma<sup>3</sup> melanomas of the iris, orbit<sup>8</sup> central nervous system,<sup>9</sup> and skin.<sup>10</sup> It is estimated that one in 400 patients with ODM followed for life develop uveal melanoma, as compared to one of 13 000 in the general population.<sup>11</sup>

## CONCLUSIONS

On the basis of accumulated reports in the literature ocular and oculodermal melanocytosis have been stated to have an increased malignant potential. An association between ODM and uveal melanoma is well recognized. Therefore, all patients with Oculodermal melanocytosis should have periodic ocular examination to allow early detection of glaucoma and uveal melanoma.

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**Source of support:** Nil  
**Conflict of interest:** None Declared