

Giant nevus of Ota

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Key words: Hamartoma, melanoma, nevus of Ota, ocular melanocytosis, skin hyperpigmentation

Nevus of Ota is a hamartoma of dermal melanocytes presenting as blue hyperpigmented patches on the face usually within the distribution of ophthalmic and maxillary branches of the trigeminal nerve.^[1] A case of giant nevus of Ota involving all three branches of the trigeminal nerve is presented.

Case Report

An 18-year-old girl presented with skin hyperpigmentation over the left side of the face since childhood. External examination revealed bluish hyperpigmented patch involving the forehead, periorbital area, temple, cheek and mandibular area [Fig. 1]. Examination of the left eye showed conjunctival melanocytosis, anterior and intercalary staphyloma, leucomatous corneal opacity and pseudoproptosis [Fig. 2]. The right eye was essentially normal. Visual acuity was nil perception of light in the left eye and 20/20 in the right eye. Left eye ultrasound ruled out intraocular mass and showed optic nerve head cupping; the axial length was 31 mm [Fig. 3]. Magnetic resonance imaging of brain done elsewhere (images not available) was reported normal. She underwent enucleation of the left eye. Histopathological examination of enucleated eye revealed melanocytic proliferation in the conjunctiva, iris, ciliary body, choroid and sclera [Fig. 4].

Discussion

Nevus of Ota usually appears at birth and has a female preponderance (4.8:1).^[2] It may be associated with nevus of Ito, ocular melanoma (1 in 400) and glaucoma (10.3%).^[3] Hence, the need for detailed ophthalmic and systemic examination. Histologically it is classified into five types, which are superficial, superficial dominant, diffuse, deep dominant,

and deep. The more superficial lesions tend to be located on the cheeks while deeper lesions occur on periorbital areas, temple, and forehead. This classification bears significance in making a therapeutic prognosis of the disease.^[4] A lifelong follow-up with dermatologist and ophthalmologist



Figure 1: External photograph reveals blue-grey skin pigmentation over left face involving forehead, periorbital area, temple, cheek and eyelids. Hemifacial involvement is typical while the involvement of mandibular area is atypical. Left eye (left image in panel) shows pseudo-proptosis caused by increased axial length due to secondary glaucoma

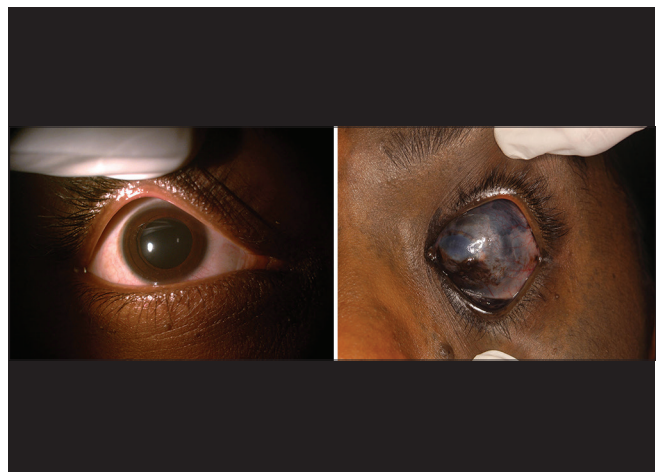


Figure 2: Right eye is normal (left). Left eye has ocular melanocytosis involving eyelids, conjunctiva, sclera, cornea and uvea. Anterior and intercalary staphyloma, leucomatous corneal opacity and buphthalmos are due to long-standing secondary glaucoma

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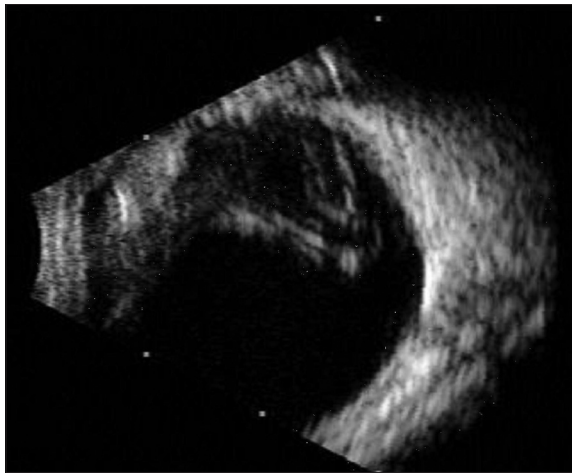


Figure 3: Ultrasound shows vitreous detachment; axial length is 31.5 mm. In the presence of media opacity, ultrasound helps rule out intraocular mass lesion

is required given the increased incidence of malignant melanoma.^[5]

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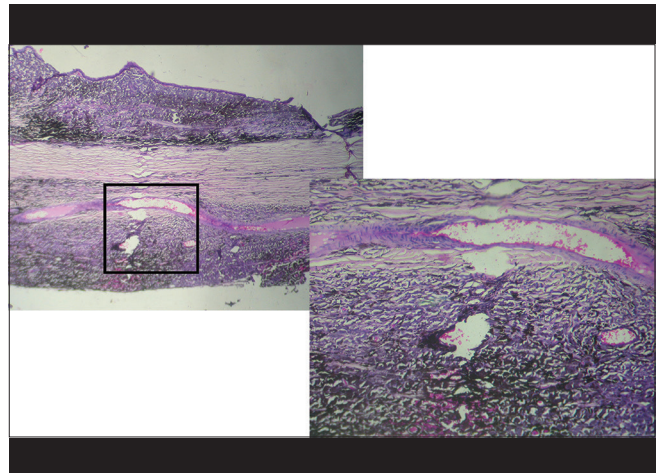


Figure 4: Histopathologic examination of enucleated eye reveals extensive melanin infiltration in the uveal tissue (H and E, x40)

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