HYPERPIGMENTATION OF MALAR AREA, PERIORBITAL SKIN ALONG WITH INVOLVEMENT OF SCLERA.

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CASE REPORT

A 18 year old male presented with complaint of asymptomatic, hyper-pigmentation of right side of cheek and both sides of periorbital skin since birth. The pigmentation appeared over both sides of eyelids simultaneously and get extended insidiously to the present size. Bluish pigmentation of sclera was noticed at the age of 6 years. Examination revealed confluent, slate gray hyperpigmented macules over right malar area. Periorbital skin is greenish-blue along with mottled, deep blue pigmentation of sclera of both eyes (Fig 1). There was no significant personal or family history. A biopsy was done from malar area and send for H&E staining (Fig 2).

Legends for figures
Fig 1: Confluent hyperpigmented macules on right cheek with greenish-blue disoloration of periorbital area. Mottled deep blue pigmented sclera also visible.
Fig 2: Elongated and spindle shaped melanocytes scattered among collagen bundles of upper reticular and papillary dermis.

What's your diagnosis?
Diagnosis: Nevus fuscoceruleus ophthalmomaxillaris/ Nevus of Ota.

Histopathology revealed infiltration of papillary and reticular dermis with elongated and spindle shaped melanocytes containing abundant melanin pigment.

DISCUSSION

The nevus of Ota consists of hyperpigmentation of facial skin and mucous membranes in the distribution of the ophthalmic, maxillary and rarely, the mandibular divisions of the trigeminal nerve. In 1939, Ota and Tanino described several cases of pigmented nevus of the skin and eye and named then “nevus fuscoceruleus ophthalmomaxillaris of Ota.” Tanino has classified the nevus of Ota into four types.1

Type I.
1A. Mild orbital type: Distribution over the upper and lower eyelids, periocular and temple region.
1B. Mild zygomatic type: Pigmentation is found in the infrapalpebral fold, nasolabial fold and the zygomatic region.
1C. Mild forehead type: Involvement of the forehead alone.
1D. Involvement of ala nasi alone.

Type II. Moderate type: Distribution over the upper and lower eyelids, periocular, zygomatic, cheek and temple regions.
Type III. The lesion involves the scalp, forehead, eyebrow and nose.

Type IV. Bilateral type: Both sides are involved.

Nevus of Ota occurs predominantly in more darkly pigmented individuals, especially in Asians and blacks. It is non hereditary entity showing two peaks of onset: the first (~50-60% of all cases) during infancy with the majority presenting at birth, and the second (40-50%) around puberty. About 80% of the reported cases with this nevus are females. It is mostly unilateral and bilateral involvement is seen only in 4% cases. Our patient is of rare type i.e Type IV of Tanino classification of nevus of Ota.

Classical presentation is presence of confluent individual macules varying from pinhead sized to several millimeters in diameter. Pigmentation is often speckled and is composed of deeper bluish and more superficial brownish element, due to Tyndal effect. In about 2/3 rd of patients it involves ipsilateral sclera whereas cornea, iris, fundus oculi, retrobulbar fat, periostium, retina and optic nerve are rarely involved. The melanocytosis also affects the oral cavity, nasal mucosa, external auditory canal, tympanic membrane, orbital fissures, meninges and the brain. It should be differentiated from Mongolian spot, blue nevus, melasma and acquired bilateral nevus of Ota like macules (ABNOM or Hori nevus). Treatment options were limited prior to the advent of laser therapy but nowadays the Q-switched lasers such as Ruby 694nm, Alexanderite 755nm and Nd:YAG 1064nm, have become the mainstay of therapy. Although malignant changes are rare to occur but detailed systemic neurologic, ophthalmological and oral examinations should be done in all cases of nevus of Ota.

REFERENCES