Case Report

Bilateral nevus of Ota

Vandana Mehta, C. Balachandran

Department of Skin & STD, Kasturba Medical College, Manipal, India.

Abstract

Nevus of Ota is a dermal melanocytosis seen in the distribution of ophthalmic, maxillary and mandibular divisions of the trigeminal nerve. Most of the cases reported are in females with a typical unilateral distribution. We describe a case of bilateral nevus of Ota in a male.

Key words

Nevus of Ota, bilateral

Introduction

Nevus of Ota or nevus fuscocaeruleus ophthalmomaxillaris was first described by the Japanese dermatologist Ota in 1939 as a dermal melanocytic hamartoma that presents as bluish hyperpigmentation along the ophthalmic, maxillary and mandibular branches of the trigeminal nerve. It is usually unilateral and bilateral involvement is described in less than 5% of cases. We report an unusual case of bilateral nevus of Ota in a young male.

Case report

A 42-year-old male presented with asymptomatic bluish grey pigmentation on both the cheeks, extending up to the forehead and temples with involvement of both the eyelids and right nasal ala since birth (Figures 1 and 2). He also had pigmentation of the sclera on the right side, but the left eye, oral or palatine mucosa were not involved (Figure 3). There was no family history of the same and general physical examination failed to reveal any associated abnormalities. Biopsy showed pigmentation of the basal layer with elongated spindle shaped melanocytes in the dermis (Figure 4).

Discussion

Ota’s nevus, originally described as nevus fuscocaeruleus ophthalmomaxillaris by Ota and Tanino in 1939 is a dermal melanocytosis.1 It is usually congenital, however familial cases have been reported.2 Two peak ages of onset, in early infancy and early adolescence suggest a probable hormonal influence. It is said to be most prevalent in Japan where the incidence among the dermatology outpatients lies between 0.2% to 1%. In Indians it is comparatively rare with the male to female sex ratio being 1:4.8, women being more frequently affected than men. It is usually unilateral and is located in the areas innervated by the first and second branches of the trigeminal nerve. The pigmentation of Ota’s nevus is composed of flat blue black or slate grey macules intermingled with
Figure 1 Bluish to slate grey pigmentation on the right malar region, forehead, upper eyelid and nasal ala.

Figure 2 Bluish pigmentation on the left side of the face.

Figure 3 Bluish grey pigmentation on the Right sclera.

Figure 4 10X photomicrograph showing dendritic melanocytes in the upper dermis.

small brown specks. The intensity of pigmentation may be influenced by fatigue, menstruation, insomnia and weather. Based on the distribution and extent of pigmentation, Ota’s nevus has been classified into four types by Tanino: mild (type 1), moderate (type 2), intensive (type 3) and bilateral (type 4). Bilateral involvement is rare and is described in less than 5% of patients. Our case corresponds to type 4 according to Tanino’s classification with bilateral extensive involvement of the face.

Ocular melanosis in 22-77% cases is almost always ipsilateral and deep in the conjunctiva. Pigmentation may also affect the sclera, cornea, iris, choroid and less commonly the optic nerve, retrobulbar fat, orbit, periosteum and extraocular muscles. The pigmentation of mucous membranes of the head and neck is variable; tympanic membrane being most frequently affected although nasal, buccal, pharyngeal and rarely palatine mucosa may be involved. No mucosal pigmentation was seen in our patient. Histology typically shows darkly pigmented, spindle shaped dendritic melanocytes in the upper and mid dermis. Though the patients are mainly concerned about the cosmetic appearance, glaucoma and ocular melanoma are the more serious complications that may occur in less than
10% cases, which makes regular ophthalmological follow up mandatory for patients with ocular melanosis.67 Sturge-Weber syndrome, Klippel-Trenaunay syndrome, neurofibromatosis, ipsilateral deafness, congenital cataract and spinocerebellar degeneration have all been associated with nevus of Ota.6 Acquired bilateral nevus of Ota-like macules (ABNOM) or Hori’s nevus clinically and histologically resembles nevus of Ota, but can be differentiated by its late age of onset in the fourth and fifth decade and the lack of conjunctival, mucosal and tympanic membrane involvement.9 Selective photothermolysis with Q switched ruby laser is considered to be a safe and effective treatment for this pigmentation.10

References


Erratum

In the October-December 2007 issue of the *Journal of Pakistan Association of Dermatologists*:

1. In the Review article entitled ‘Miltefosine: a breakthrough in treatment of leishmaniasis’ the name of second author was missed. The correct authors list is ‘Arfan ul Bari, Simeen Ber Rahman’.

2. In the case report ‘Epidermodysplasia verruciformis: a rare genodermatosis with risk of malignant transformation’ the names of second and third authors were wrongly mentioned. The correct authors list is ‘Arfan ul Bari, Simeen Ber Rahman’.

We are sorry for the typographical errors.