Eur. J. Pediat. Dermatol.
20, 123-7, 2010

Bilateral nevus of Ota associated with Turner syndrome.

Alsaif F., Alshahwan H.
Department of Dermatology, King Khalid University Hospital,
King Saud University, Riyadh, Saudi Arabia

Summary Here is reported a case of bilateral nevus of Ota in a 10-year-old girl with Turner syndrome and growth hormone deficiency. The cutaneous lesions got visible at the age of 6 months. In the following five years the skin lesions got more evident and widespread. They also darkened during the administration of the growth hormone. The ophthalmological examination showed, besides the conjunctival and scleral pigmentation, esotropia and nystagmus. The histological examination showed the presence of dermal melanocytes, confirming the clinical diagnosis of nevus of Ota. The cutaneous lesions were treated with Q-switched alexandrite laser. According to the Authors, this is the first case of nevus of Ota associated with Turner syndrome reported in the literature.

Key words Nevus of Ota, Turner syndrome.

In 1939, Ota described the first case of a benign dermal melanocytosis known as nevus of Ota. This condition involves the skin and mucosa innervated by the ophthalmic, maxillary and, rarely, the mandibular division of the trigeminal nerve. It is frequent in people of Japanese descent but is also seen in individuals of Chinese, Indian, African and European descent. Nevus of Ota is more frequent in females than males.

Unilateral presentation is typically seen, but bilateral involvement is described in 5% to 10% of patients. We report a 10-year-old Saudi girl with bilateral nevus of Ota associated with Turner syndrome. This is the first report of such an association.

Case report

A 10-year-old Saudi girl with a known case of Turner syndrome [chromosomal analysis, 46, X, del (X) (p22)] with growth hormone deficiency presented to the dermatology clinic with an asymptomatic blue-gray lesion on the upper part of the face that had been present since the age of 6 months.

The lesions had gradually progressed with age and extended to the forehead, temple and bilateral periorbital areas. Then they had been static for 5 years. There was a history of lesion darkening coincident with growth hormone treatment.

Her parents were non-consanguineous, and no one else in the family had similar pigmentation.

Physical examination revealed a short stature and the subtle facial features of Turner syndrome. Cutaneous examination revealed a bilateral, diffuse, homogeneous, slate-colored, blue-gray patch over the forehead, periorbital area, temples, nasal bridge and sclerae (Fig. 1, 2, 3). Nasal and oral mucosae were not affected. Other Turner syndrome features, including wide spaced nipples and wide carrying angle, were also present. Ophthalmologic examination revealed scleral and conjunctival pigmentation, nystagmus and

esotropia. Renal and ovarian ultrasounds were normal. Cardiologic and audiometric examination were normal.

The histopathologic study showed normal epidermis and the presence of numerous elongated, spindle-shaped melanocytes with their long axis parallel to the skin surface. They were scattered between the collagen bundles in the mid-dermis (Fig. 4 and box).

Based on clinical findings and the complementary examinations performed, the diagnosis of Turner syndrome associated with nevus of Ota was made in this patient. The patient is now undergoing treatment with the Q-switched alexandrite laser with 755 nm wavelength, 3 mm spot size, and 7 J/cm^2.

Discussion

Nevus of Ota was first described by a Japanese dermatologist, Ota, who reported an unusual

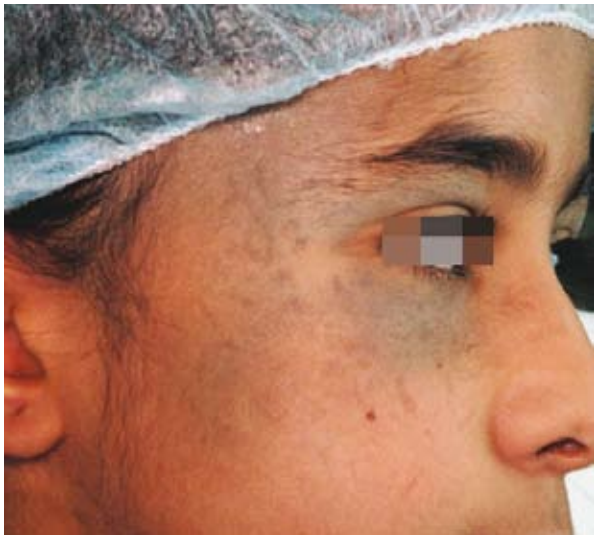

Fig. 1

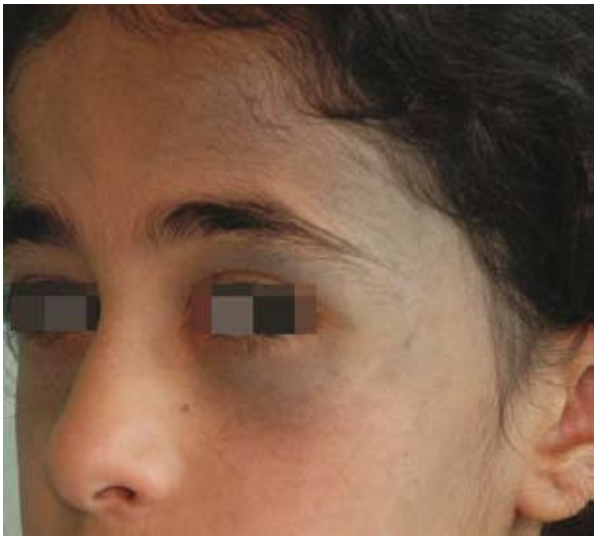

Fig. 2

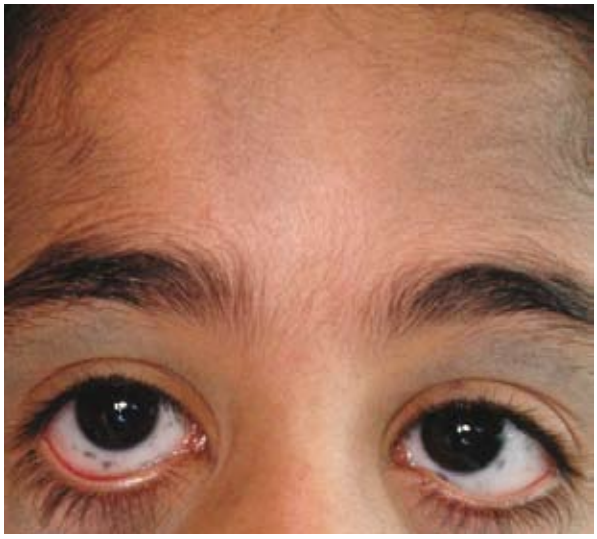

Fig. 3

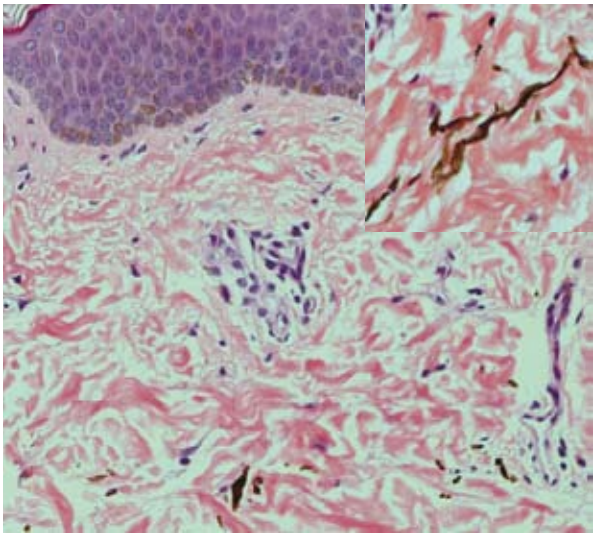

Fig. 4

Fig. 1, 2, 3, 4: Bilateral nevus of Ota in a 10-year-old girl with Turner syndrome (Fig. 1, 2, 3). The histological examination showed dendritic melanocytes in the dermis (Fig. 4 and box).

syndrome of Nevus fusco-ceruleus ophthalmomaxillaris in 1939 (24, 25). It is very common in Asians, especially in the Japanese population, but it is also seen in people of Chinese, Indian, African and European descent. Several epidemiological studies have shown higher prevalence in Japan with rates between 0.4 and 1.1% (10, 29, 36). A Canadian study in Black populations revealed an incidence of up to 0.014% (7).

About 80% of the patients were females (16). Forty-eight percent of patients developed nevus of Ota at or after birth compared to 11% between 1 and 10 years of age and 36% at puberty (10).

The pathogenesis of nevus of Ota is likely multifactorial, including genetic factors, as the lesion has a much higher incidence among Asians than in other populations (19). Other stimulus factors are female sex hormones, ultraviolet light exposure and trauma (16).

Histopathology of affected skin shows the presence of typical dendritic melanocytes containing melanin in the dermis (13).

The clinical manifestations are usually unilateral, and only 5% of cases are bilateral (4). Its presentation is characterized by the presence of blue-gray macular pigmentation with irregular borders and commonly involves skin and mucosa innervated by the ophthalmic, maxillary and rarely mandibular division of the trigeminal nerve (4).

Tanino classified nevus of Ota into 4 types (Table 1), depending on the extent and distribution of pigmentation (29). According to this classification, this case would correspond to type IV.

Apart from skin and scleral involvement, pigmentation has been documented in the episclera, conjunctiva, cornea, iris, retina and uveal tract. Similar discoloration can be observed in oral and nasal mucosae and in the tympanic membrane. Leptomeninges can also be affected (26). Open-angle glaucoma (18) and melanoma (5, 8, 14) are rarely associated with nevus of Ota with only about 10 cases of cutaneous melanoma in nevus of Ota documented to date. Melanoma arising in the brain and optical structures are more frequently associated with this type of lesion (26, 30).

Turner syndrome is a sex chromosome disorder with an incidence between 1:2000 to 1:5000 live female births (6, 11). The defining clinical triad of impaired sexual development and webbed neck was first described in the English-language literature by Turner in 1938 (31), and later ovarian failure and streaked gonads were noted in 1944 by Wilkins e Fleischmann (34). The main features of this syndrome are short stature, impaired sexual development, infertility, webbed neck and lymphedema.

Turner syndrome has been associated with many coincident cutaneous dermatoses. Even though most patients with Turner syndrome have an increased number of acquired melanocytic nevi (1, 37), it is still unclear if this predisposes these individuals to melanoma, as it does in the rest of the population, or if Turner syndrome has

TABLE 1: TYPES OF NEVUS OF OTA

Type I: IA. Mild orbital type: distribution over the upper and lower eyelids, periocular and temple regions; IB. Mild zygomatic type: pigmentation is found in the infrapalpebral fold, nasolabial fold and the zygomatic region; IC. Mild forehead type: involvement of the forehead alone; ID. Mild nasal: 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, eyebrows and nose.

Type IV: Bilateral type: both sides are involved.

some protective effect against melanoma (6, 28). Because many case reports of melanoma have been reported with nevus of Ota, the risk of melanoma in our case needs to be considered. Other cutaneous associations with Turner syndrome are halo nevus (2, 22), vitiligo (9, 20), neurofibromatosis (9), lymphedema (20), alopecia areata (27) and psoriasis (20, 27).

Various therapies have been used to treat nevus of Ota successfully. Cosmetic cover-up products can be used for camouflage. Cryosurgery and microsurgical treatments can leave disfiguring scars and are not recommended. Combined dermabrasion and carbon dioxide lasers have yielded good results (33). Treatment with photothermolysis with Q-switched lasers is safe and effective for lightening nevi of Ota (3, 12, 15, 23, 35). Side effects such as mild purpura, erythema and edema can follow the treatment and resolve after a few days, but pigmentation changes, such as hyperpigmentation or hypopigmentation, are more frequent. Topical tretinoin, hydroquinone, and corticosteroid creams can also be used in the treatment of post-inflammatory hyperpigmentation (2, 21).

The response to laser treatment has been reported to be dependent on lesion color. Darker lesions, which have a greater number of melanocytes, require more treatments for eradication (3). Overall, laser therapy is very effective in the treatment of nevus of Ota, and recurrence is rare (2, 23).

Address to:
Dr. Fahad Al Saif
Chairman and Consultant
Department of Dermatology
P.O. Box - 2925
Riyadh - 11461
e-mail: fsaif1000@hotmail.com

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