

Pruritic Acquired Nevus of Ota

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Abstract

Nevus of Ota is a unilateral, asymptomatic cutaneous and mucosal hyperpigmentation of the face that is congenital or may appear during childhood. We present a case of symptomatic acquired nevus of Ota in an adult, associated with intense pruritus, not described in the literature so far. A 32-year-old woman presented with brownish mottled macules which appeared on her face progressively over 8 days, following the distribution of the first and second divisions of the left trigeminal nerve and partially covering the iris and sclera of the left eye. She reported an intense pruritus in this area. We performed a biopsy on the left forehead, which confirmed the diagnosis of nevus of Ota. Specific stains and immunohistochemistry revealed increased numbers of mast cells. Ophthalmological tests showed acute acquired melanocytosis of the left iris and sclera. The origin of the nevus is still unclear. Several hypotheses suggest a reactivation of melanocytes during their migration from the neural crest. The pruritus reported in our patient may be explained by the increased

quantity of mast cells observed in the lesion and/or neuronal stimulation of the ophthalmic and maxillary divisions of the fifth cranial nerve.

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Introduction

Nevus of Ota is a unilateral cutaneous and mucosal hyperpigmentation, appearing as bluish or brownish macules that occur on the areas of the face innervated by the maxillary and ophthalmic branches of the trigeminal nerve, and rarely by the mandibular branch [1, 2]. Besides skin, it involves ocular tissue and sclera in most cases; it also increases the risk of glaucoma, and can be associated with melanotic tumors, especially uveal melanoma [3]. Nevus of Ota has been classified into type I, II, III and IV (i.e. mild, moderate, intense and bilateral, respectively) [1]. The Orientals, Blacks, Asians and rarely the Caucasians are affected, with the highest prevalence in women with a sex ratio of 1:4.8 [2, 4]. Nevus of Ota is a congenital unilateral melanocytic hyperpigmentation which may appear in early childhood or in puberty [4]. We report here a case of acquired unilateral nevus of Ota in a 32-year-old Kurdish Iraqi woman.

Case Report

A 32-year-old Kurdish Iraqi woman presented to our clinic with brownish mottled macules that had appeared progressively, starting 8 days earlier, on the left cheek, eyelid and forehead and partially on the iris and sclera of the left eye, following the distribution of the maxillary and ophthalmic divisions of the left trigeminal nerve (fig. 1, 2). She was in good health and denied taking any medication or using any topical skin preparation on the face. She described a burning and very itchy sensation that occurred with the lesion, which was strictly located on the pigmented area. Clinically, there were no erythematous changes, and the examination under Wood's lamp revealed a slightly higher contrast of the pigmented zones. Histological analysis of the skin biopsy performed on the left forehead showed a proliferation of non-atypical pigmented spindle-shaped cells in the superficial and reticular dermis (fig. 3). The immunohistochemical study revealed positivity for S-100 protein, HMB-45 and melan-A in these cells. A special stain of Toluidine blue and immunohistochemistry for CD117 (data not shown) showed an increased number of mast cells in the lesion (>15 mast cells per high-power field; fig. 4).

The proliferation index (Ki-67) in the spindle-shaped pigmented cells remained less than 5%. Immunolabeling and PCR analysis for herpes simplex virus and varicella zoster virus were negative. An MRI showed a discrete nonspecific hypersignal in the cutaneous tissues, with no abnormalities of the orbital region and the brain. An ophthalmological examination concluded an acute acquired melanocytosis of the left iris and sclera associated with heterogeneity of the iris structure, but no visual defect. An ultrasound biomicroscopy scan revealed no disorganization of the ocular structures. Considering the clinical and histopathological findings, we diagnosed the lesion as an acquired nevus of Ota.

Discussion

Dermal melanocytosis, which is described to be congenital in most cases, can appear during the first year of life and less often in adolescence [5]. Cases of late-onset dermal melanocytosis in adults are rare. Also referred to as acquired dermal melanocytosis (ADM), several hypotheses have been discussed to explain its origin. During embryological development, melanocytes migrate from the neural crest to the basal stratum. In dermal melanocytosis, a defect in this process is described. In ADM, the latent dermal melanocytes are thought to be reactivated by unknown triggers such as sex hormones and local trauma, in addition to sun damage and inflammation [6]. These factors activate the pathway to synthesize melanin. The stability of the cells is explained by a protective extracellular sheath surrounding dermal melanocytes in adult life [6].

In 1861, Hulse [7] was the first to describe a unilateral hyperpigmentation of the face and sclera. Then, in 1939, Ota [1] defined it as a clinical entity named nevus of Ota. It is also referred to as oculodermal melanocytosis, nevus fuscoceruleus ophthalmomaxillaris, or congenital melanosis bulbi [8, 9]. In 1984, Hori et al. [10] were pioneers in describing the acquired bilateral nevus of Ota-like macules as ADM. This bilateral dermal melanosis appears in the 3rd or 4th decade of life, with prevalence in females. It involves the first and second branch of the trigeminal nerve, including the forehead, temples, eyelids, cheeks and nose. The lesions are circumscribed, blue-brown or slate-gray macules, and the mucous membrane, eyes and



Fig. 1. Brownish mottled macules on the left side of the face.



Fig. 2. Brownish mottled macules on the iris and sclera.

nasal and oral cavity are not pigmented [10]. In 1987, Sun et al. [11] described the acquired nevus fuscoceruleus zygomaticus, known as acquired unilateral nevus of Ota, while in our case it involves the ophthalmic and the zygomatic divisions. In 1991, Whitmore et al. [12] reported a type IB nevus of Ota in a 30-year-old Caucasian woman. In 1994, Lynn et al. [13] reported a case of acquired nevus of Ota in an 80-year-old man. In 2002, 2 cases of late-onset nevus of Ota were reported by

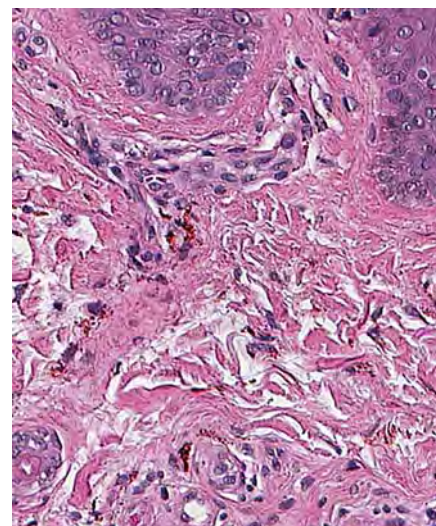


Fig. 3. Preserved epidermis overlying a dermal proliferation of intensely pigmented spindle-shaped cells. HE. $\times 20$.

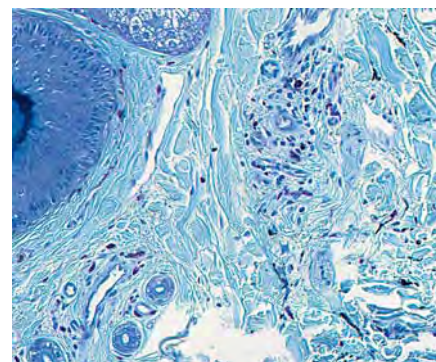


Fig. 4. Special stain of Toluidine blue confirming an increased number of mast cells ($\times 20$).

Chang et al. [14], the first in a 44-year-old man and the second in a 49-year-old woman.

Different treatment modalities are proposed to treat ADM. Laser therapy such as Q-switched Ruby, Q-switched Alexandrite and Q-switched Nd-Yag lasers have been reported to be effective in the treatment of ADM, with some adverse effects observed after treatment [6, 15]. The principle of these lasers is to destroy melanin of the dermal melanocytes and melanophages via se-

lective photothermolysis. Other modalities also exist, even if poorly successful, such as cryotherapy, microsurgery, dermabrasion, chemical peeling and carbon dioxide laser vaporization [6].

Our patient is a 32-year-old Caucasian woman, with an acquired nevus of Ota associated with intense pruritus on the hyperpigmented area. Neurons and skin are both issued from the same embryological tissues. The origin of this pruritus could be explained

by the increased quantity of mast cells. It can also be related to neuronal stimulation of the ophthalmic and maxillary divisions of the fifth cranial nerve in the whole process.

Interestingly, the pruritus spontaneously faded 10 weeks after the onset of the lesion. As a therapeutic approach for the pigmentation, the patient is currently undergoing Q-switched ND-YAG laser sessions (1,064 nm) and progressive fading is observed.

To our knowledge, this is the first case report of acquired nevus of Ota associated with pruritus.

Disclosure Statement

The authors declare no conflicts of interest.

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