Eruptive Blue Nevi of the Scalp in a Patient with Mayer-Rokitansky-Küster-Hauser Syndrome during Isotretinoin Therapy

Nevena Skroza, Ilaria Proietti, Ersilia Tolino, Nicoletta Bernardini, Veronica Balduzzi, Anna Marchesiello, Alessia Anzalone, Daniela Colapietra, Simone Michelini, Alessandra Mambrin, Natale Porta[1], Vincenzo Petrozza[1], Concetta Potenza

From the Department of Medical-Surgical Sciences and Bio-Technologies, Dermatology Unit "Daniele Innocenzi", Sapienza University of Rome, Fiorini Hospital, Polo Pontino, Terracina, [1]Department of Medical-Surgical Sciences and Bio-Technologies, Pathological Unit, Sapienza University of Rome, Polo Pontino I.C.O.T. Hospital, Latina, Italy.
E-mail: proiettilaria@gmail.com

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Sir,

Blue nevi are benign melanocytic lesions located in the deeper reticular dermis, usually asymptomatic and solitary.[1] Multiple blue nevi have rarely been reported and are defined as plaque-type, agminated, or eruptive blue nevi. They are generally clustered on a cutaneous surface measuring <10 cm.[2]

This condition is characterized by a rapid appearance of multiple melanocytic nevi mainly developed in immunosuppressed patients or those affected by blistering disease, varicella zoster virus infection, severe sunburn, or trauma.[2,3]

The abrupt eruption of melanocytic lesions may also occur in physiological conditions such as puberty or pregnancy.[3]

A 23-year-old Caucasian woman with Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome type 1 presented with abrupt onset of small, well-restricted, blue macules with dermatomal distribution on the right temporal region during isotretinoin treatment for severe acne.

No personal or family history of melanoma was reported, although her mother notified to have several blue nevi, one of them localized on the right temporal region.

The clinical history revealed no previous trauma, severe sunburn, or other cutaneous diseases.

Physical examination revealed a 4-mm blue papule with discreet 2-mm blue satellite macules within a normally pigmented 5-cm area of the right temporal scalp [Figure 1a].

Dermoscopic examination revealed the presence of diffuse and homogeneous bluish pigmentation. We ruled out primary melanoma due to the absence of atypical network, vessel, irregular globules and dots, pseudopods regression areas, and we excluded trichoepithelioma due to abscence of large blue-gray ovoid nests, leaf-like areas, and arborizing vessels [Figure 1b]. Considering the sudden onset, the number and appearance of the lesions might pose problem of differential diagnosis of cutaneous metastases from occult malignant melanoma. However, clinical examination revealed neither atypical pigmentation nor partially regressed lesions. No regional lymphadenopathy was found.

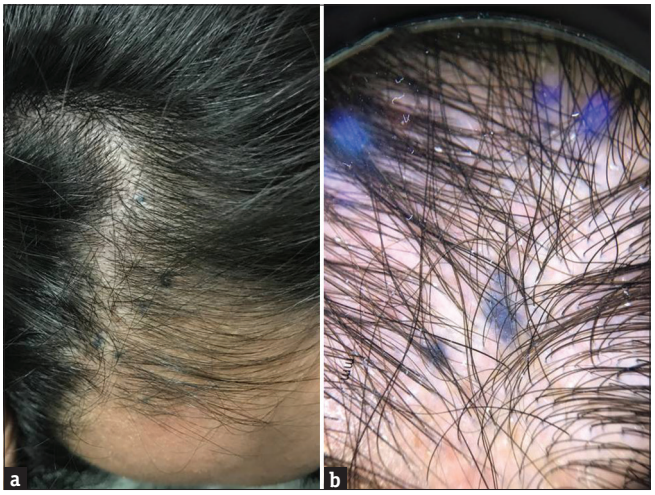

Figure 1: (a and b) Clinical and dermoscopic features of blue nevi with dermatomal distribution on the right temporal region

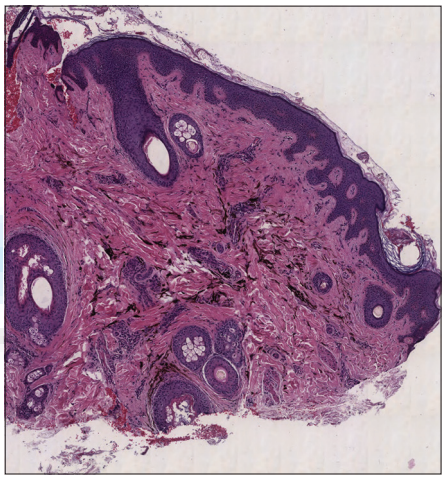

Figure 2: Aggregates of elongated, finely branching, melanocytes and scattered melanophages in the interstices of the dermal collagen bundles of the mid- and upper dermis, separated by normal skin (H and E, ×20)

To exclude the suspicion of cutaneous metastasis from occult malignant melanoma, surgical removal of all lesions [Figure 2] was performed. The histopathological examination revealed aggregates of elongated, finely branching, melanocytes and scattered melanophages

in the interstices of the dermal collagen bundles of the mid- and upper dermis, separated by normal skin. Therefore, on the basis of the clinical and histopathological features, a conclusive diagnosis of multiple disseminated common blue nevi was made.

The blue nevus is defined as a benign neoplasm of acquired or congenital aggregate of aberrant dermal melanocytes actively producing melanin, presents in 0.5%–4% of healthy caucasian adults. It is typically located in the head and dorsa of the hands and feet.

According to common nosological classifications, blue nevi belong to the disease group of dermal melanocytoses such as nevus of Ito, nevus of Ota, Mongolian spot, and other forms of dermal pigmentation, characterized by the presence of dermal melanocytes. Recently, eruptive and slowly progressing blue nevi in combination with cutis marmorata teleangiectatica congenita were described.[4]

Furthermore, multiple blue nevi can present as a part of the Carney complex/lentigines, atrial myxomas, blue nevi syndrome, along with lentigines, atrial myxomas, and mucocutaneous mastocytosis.[4]

Our patient was affected by a congenital defect found at birth in normal 46, XX karyotype female named MRKH syndrome. It presents aplasia or severe hypoplasia of vagina, uterus, and fallopian tubes, renal agenesis, and cervicothoracic somite anomalies. External genitals as well as hormonal profiles are normal.[5]

This patient developed eruptive blue nevi during isotretinoin therapy and this might be hypothesized as triggering factor since the patient had no clinical features related to other mentioned syndromes or diseases nor any other possible triggering factor.

Besides, given the rarity of both the conditions (blue nevi and MRKH), we report this unusual association.

Furthermore, we highlight the importance of the differential diagnosis with cutaneous metastasis of occult melanoma, based on the correlation of clinical, dermoscopic, and microscopic data.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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