

Hypertrichotic Plaque-type Blue Naevus – A Novel Type of Dermal Melanocytosis: Report of an Unusual Case

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Sir,

In 1977, the term “dermal melanocytosis” was coined by Delacrétaz for the various forms of dermal melanocytic accumulations (1). Since then, the Mongolian spot, the naevus of Ota and several variants of the blue naevus, such as the plaque-type, have been grouped in this category (2–5). The patient presented here, while displaying a striking clinical similarity to Delacrétaz’s case, demonstrates a number of unique features suggesting a new entity.

CASE REPORT

A 19-year-old Caucasian male patient presented with lesions that had been regarded as angiodysplasia. They had first been noted at the age of 2 years and had progressed during childhood. Treatment had been repeatedly attempted with sclerotherapy. During puberty, conspicuous terminal hairs of fair colour, in accordance with the patient’s skin type II, appeared within the affected regions.

A poorly defined bluish-grey discoloration was noted over the back of the right hand and on the sides of the proximal phalanges of the third and fourth fingers and in the fold between these fingers extending to the back of the hand. Within the discoloured area, several sharply defined, flat, dark-blue or, more rarely, brown papules were seen (Fig. 1). These were of firm consistency and not attached to the deeper tissue. Conspicuous terminal rather fair hairs were evident within the areas of discoloration on the back of the fingers. Conversely, on the back of the fingers of the healthy hand, the patient had no hairs. On the palmar side of the hand, individual black macules were visible, and an oval nodule of about 1 cm in diameter, painful on palpation, was located subcutaneously on the distal palm; on the proximal volar side of the forearm and in the anticubital fossa two ill-defined, light blue areas measuring 3 and 6 cm in diameter were noted. Poorly defined, deep-blue, dermal and subcutaneous nodules measuring roughly 0.5 cm in diameter were regularly distributed within these macular areas (Fig. 2). The skin lesions did not follow Blaschko’s lines or any other definite pattern.

A skin biopsy of a blue papule from the back of the hand showed a cellular blue naevus extending throughout the mid-dermis into the subcutis. No atypical mitoses or proliferating cells were seen.



Fig. 1. Bluish-grey discoloration with individual dark blue papules. Conspicuous terminal hairs within the areas of discoloration on the backs of the fingers which are rather fair, in accordance with the patient’s complexion.



Fig. 2. Ill-defined light blue plaques with deep blue nodules and terminal hairs of the cubital and the volar side of the forearm.

Since the patient had in the past been erroneously diagnosed as having Klippel-Trenaunay’s syndrome, phlebography and arteriography had been performed and shown no anomalies. Thermography showed that the surface temperature over the pigmented spots was up to 4° lower than the skin of the left hand.

DISCUSSION

Our patient represents a new type of dermal melanocytosis unique on the basis of several clinical features, including time of onset, appearance, location of the lesions and associated hypertrichosis. We therefore propose the term hypertrichotic plaque-type blue naevus for this condition.

Plaque-type blue naevi are known to arise either before birth (6–8) or between 10 and 30 years (1, 9–11). Our patient is unusual because of the early onset of the lesions. The progression during puberty has been observed before, although no triggers, such as hormonal factors, have been described (2, 9, 12).

The majority of patients show involvement of the scalp (6, 9, 10, 13). There is only one report of localization on the arm (11) and one on the hand (1, 2), making the combined localization on arm and hand, as displayed by our patient, highly unusual.

So far, most cases reported with non-scalp location have been devoid of terminal or vellus hair (6, 10). The differential diagnosis of our patient also includes trichoplasic nevoid variants such as the “pilar neurocristic hamartoma” (13, 14). However, these tumours contain neuromesenchymal and fibrogenic components not present in our patient’s lesions.

Plaque-type blue naevi are usually benign and asymptomatic lesions, but a definite prognosis cannot be made for individual patients (2). Malignancy has been reported to arise exclusively in scalp lesions (9, 13). On the other hand, it is well known that enlarging, benign blue naevi can displace or damage structures such as nerves, making neurologic follow-up evaluation advisable.

Since phlebography as well as arteriography were normal, the reduced surface temperature over the pigmented spots is an effect more of dermal melanin than of reduced blood supply. There are recent reports in the literature on successful therapy of dermal melanocytoses with the Q-switched-Neodyn-YAG laser, although

poorer results are reported in the treatment of papular areas of the extremities (5). Our patient is currently undergoing YAG laser treatments, with moderate improvement.

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