

Terra Firma-Forme Dermatositis Misdiagnosed as Nevroid Acanthosis Nigricans

Gulsen Tukenmez Demirci, MD,* Ayse Tulin Mansur, MD,* and Ebru Demiralay, MD†

Abstract: Terra firma-forme dermatosis (TFFD) is a clinical condition characterized by brown-gray, velvety, pigmented patches or plaques, resembling dirt on the skin. Nevroid acanthosis nigricans (NAN) is a rare and recently described form of acanthosis nigricans occurring during childhood or early adulthood. Herein we describe a patient with TFFD, initially misdiagnosed as NAN. The patient had developed hyperkeratotic and hyperpigmented plaques on and around the umbilicus during pregnancy. Though regular in bathing practices, she could not clear away the lesions and concerned marks for inesthetic appearance. Histopathological findings were compatible with NAN, and she was prescribed 10% urea lotion. On a dramatic healing after 3 weeks, a diagnosis of TFFD is considered and confirmed by the lesions getting wiped away by vigorous rubbing with alcohol pads. We discuss the key points of differentiating TFFD from NAN, and underline the importance of alcohol test for accurate diagnosis.

Key Words: terra firma-forme dermatosis, dermatosis neglecta, nevroid acanthosis nigricans, benign acanthosis nigricans

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Terra firma-forme dermatosis (TFFD) is a clinical condition characterized by brown-gray, velvety, pigmented patches or plaques, resembling dirt on the skin.¹ Nevroid acanthosis nigricans (NAN) is a rare and recently described form of acanthosis nigricans (AN) occurring during childhood or early adulthood.²

Herein we describe a patient with TFFD, and discuss the key points of differentiation from NAN.

CASE

A 35-year-old woman presented with a 7.5-months history of thick and dark colored plaque on and around the umbilicus. She had given birth to twin babies 2.5 months ago by cesarean section. She had first noticed a brownish blotch over the umbilical area and midline of abdomen at the fifth month of pregnancy, which had gradually increased in size and thickened. The lesions were asymptomatic, but the patient was very distressed because of cosmetic disfigurement.

From the Departments of *Dermatology, and †Pathology, Başkent University İstanbul Hospital, İstanbul, Turkey.

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Reprints: Gulsen T. Demirci, MD, Başkent Üniversitesi İstanbul Hastanesi, Mahir İz Cad. No: 43, 34662 Altunizade İstanbul, Turkey (e-mail: gulsentukenmez@yahoo.com).

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She had no history of diabetes, other endocrine disorders, malignancies, or congenital syndromes, and took no medications. There was no family history of a similar skin condition.

Physical examination revealed a brownish to black, hyperkeratotic, verrucous, and velvety plaque on the midline of the abdomen, including the umbilicus. Similar, but less prominent lesions were seen along the linea nigra of pregnancy. The sites of previous piercing were filled with a dark, keratotic material. There were punctate hyperpigmented and slightly hyperkeratotic lesions, beginning from the umbilical plaque, and symmetrically involving the sides of the abdomen (Fig. 1).

The remainder of the skin examination, including those of the flexural areas, was within normal limits. Hair, nails, mucous membranes, and other systems were unremarkable.

Laboratory investigations including complete blood cell count, fasting blood glucose level, hepatorenal function tests, thyroid function tests, insulin and glucose tolerance showed no abnormalities.

A punch biopsy specimen of the umbilical plaque revealed prominent hyperorthokeratosis, irregular mild acanthosis, papillomatosis, and slight melanin pigment along the basal layer.



FIGURE 1. Clinical picture of the lesions before treatment.

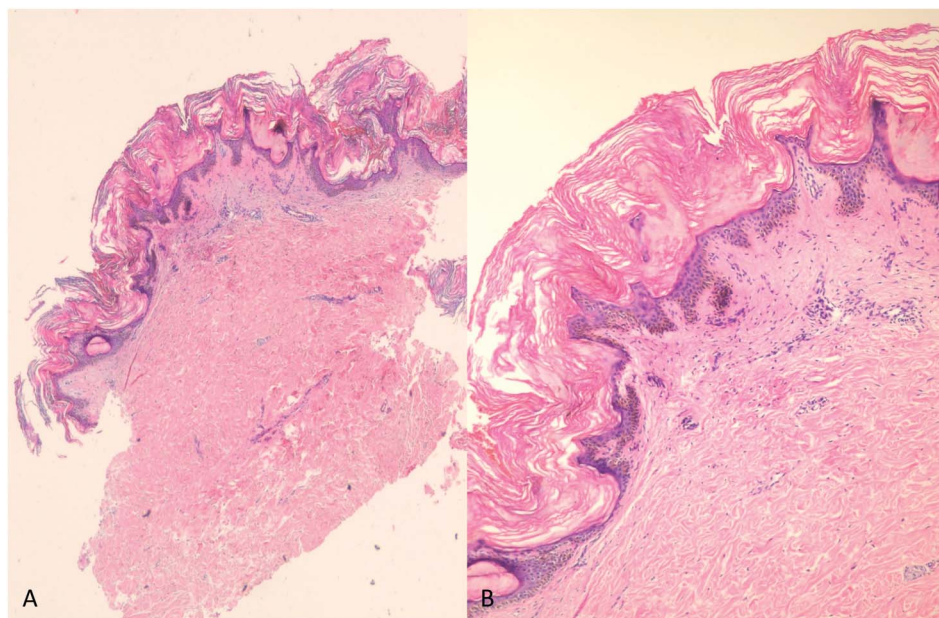


FIGURE 2. A, Microscopic features of skin lesions. B, Closer views shows prominent lamellar hyperkeratosis, finger-like projections of dermal papillae, and basal hyperpigmentation.

There was mild lymphocytic inflammatory infiltrate in the upper dermis (Figs. 2A, B).

These clinicopathologic features were interpreted as being most consistent with NAN. Urea 30% cream twice daily was prescribed. Three weeks later, the patient came for a control visit, with a very satisfying result. There was a significant healing of the umbilical plaque, with almost nearly complete resolution of hyperkeratosis, and a slight residual hyperpigmentation. Punctate lesions peripheral to the main plaque had totally disappeared (Fig. 3A). The rapid and striking healing with only urea cream led us to consider the diagnosis of TFFD. After a vigorous rubbing with ethyl alcohol 70%, the lesions located on the midline cleared, leaving remnants of keratosis on the gauze pad (Fig. 3B).

DISCUSSION

Terra firma-forme dermatosis is characterized by accumulation of hyperpigmented, hyperkeratotic, and verrucous crusts. Rarely palpable verrucous or papillomatous plaques, reticulated patches, and focal slight scaling have been observed. The lesions are resistant to conventional washing with soap and water, but can simply be eradicated by forceful rubbing with alcohol. The etiology of terra firma-forme dermatosis is unclear though it has a predilection for heavier patients and concave skin areas. It does not have any genetic relation and familial tendency. It has been suggested that delayed keratinocyte maturation plays a major role.³ Most



FIGURE 3. A, Clinical features after treatment. B, Friction with 70% alcohol, which resulted in wiping off the keratosis.

TABLE 1. Reported NAN Cases Involving Umbilicus

Case	Author	Age/Sex	Localization	Alcohol Test	Treatment/Result
1	Kim et al ⁶	18/F	Midline of abdomen, umbilicus	ND	NM
2	Lee et al ⁷	18/F	Umbilicus	ND	Tretinoin. Dramatic clearing in 2 wk.
3	Kim SK and Kim YC ⁸	16/F	Umbilicus	ND	Minocycline, adapalene. Weak compliance, no change in 7 wk.

ND, not done; NM, not mentioned; PY, publications.

patients have regular bathing habits, though neglected hygiene due to underlying disabilities may be responsible in some patients. The latter situation is denominated as dermatosis neglecta, and is considered a different spectrum of TFFD. The usual sites of involvement are face, neck, trunk, or ankles. Some unusual sites, such as scalp, axilla, back, umbilical area, pubis, arms, and legs, have also been reported.^{1,2,4} The diagnosis of terra firma-forme dermatosis is classically based upon clinical presentation and subsequent lesion resolution after the application of 70% isopropyl alcohol. Aggressive rubbing with 70% isopropyl alcohol pads is sometimes required to achieve the appropriate shearing forces to remove all pigmentation.⁵

Nevoid acanthosis nigricans has a morphologic pattern similar to other forms of AN, but in contrast, it has no predilection for flexural areas. It presents with circumscribed plaques, mostly originating from the midline with essentially unilateral distribution. Various locations of NAN have been reported, including the face, scalp, chest, abdomen, periumbilical area, back, and thigh. Occasionally it presents along the lines of Blaschko. It is not associated with syndromes, endocrinopathies, drugs, or malignancies, and there is no familial involvement. The lesions manifest at birth, childhood, puberty, or early adulthood.²

Till date, there are only 3 reports of NAN located over umbilicus (Table 1). The clinical features of these cases were strikingly similar to our case, and interestingly, in none of the cases, was alcohol rubbing test performed.^{6–8}

Histopathologic examination of NAN lesions shows hyperkeratosis, papillomatosis, and slight acanthosis in the epidermis, and a mild perivascular lymphocytic infiltration in the superficial dermis.² Biopsy of TFFD is rarely required, therefore, there are only a few reports presenting microscopic findings. The histopathological description encompasses prominent lamellar hyperkeratosis with focal areas of compact whorled orthokeratosis, keratin globules in the stratum corneum, papillomatosis, mild acanthosis, focal perivascular lymphocytic infiltration in the papillary dermis, and focally increased melanin pigment in the basal layer of the epidermis.^{1,3} Microscopic pictures of the 2 entities is highly similar, with no pathognomonic finding which helps for accurate diagnosis.

Patients in the postoperative period may be hesitant to apply pressure over the area due to concerns of pain or discomfort. In our patient, though having adequate hygiene, this approach might have prevented her to apply proper cleansing over the abdomen during her twin pregnancy, and then after cesarean section. It has been proposed that remnants of soaps, cleansers, and emollients may also participate in the prevention of normal keratinocyte shedding and enhance accumulation of scales, dirt, and sebum.^{1,3} Body oils aimed to prevent striae in pregnancy might also have contributed to the development of TFFD lesions of the presented case.

Differential diagnoses of NAN include epidermal nevus, confluent and reticulated papillomatosis, reticulate pigmented flexural anomaly (Dowling–Degos disease), Becker nevus, and hyperkeratotic type of seborrheic keratosis.² We suggest that TFFD has to be included in the differential diagnoses of NAN, especially for lesions located over the umbilical area. In order to avoid unnecessary tests and treatment, a trial of vigorous swabbing of the lesions with 70% isopropyl alcohol pads should be performed, when the diagnosis of TFFD is considered.

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