CASE REPORT

Terra firma-forme dermatosis: A rare case report and review of literature

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ABSTRACT

Terra firma-forme dermatosis (TFFD) was first described in 1987 as Duncan's dirty dermatitis, in honor of the physician who initially described this condition in Houston. The lesions appear as brown-gray pigmented plaques and patches in the face, neck, and trunk and can feel rough when palpated. The exact etiology of TFFD is still not clear. The pathomechanism is thought to be attributed to abnormal keratinization. TFFD represents retention, rather than a proliferative hyperkeratosis. Disappearance of the lesions by rubbing with 70% isopropyl alcohol is diagnostic for disease. The condition is only little known and is rarely described in the medical literature or in textbooks. Usually, the lesions of TFFD do not cause any medical threat to the patient, but they are cosmetically unsightly and cause psychological distress to the patient. Here, a new case of 22 year old man is reported to the department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University, Bangladesh with presentation of multiple asymptomatic dirty-brown muddy gray colored skin eruption on his forehead, nose and both cheeks for 2 years, despite having good hygiene. Awareness of TFFD facilitates prompt diagnosis and thereby prevents unnecessary biopsy and extensive endocrine evaluation.

KEY WORDS:Terra firma-forme dermatosis, Duncan's dirty disease, Isopropyl alcohol

INTRODUCTION

Terra firma-forme dermatosis (TFFD) is a recently described benign, cutaneous pigmentation disorder that presents with asymptomatic brown-grey, velvety, pigmented patches or plaques that resembles dirty skin.[1,2] Terra firma-forme means dirty, dry surface in Latin, but in TFFD the patients practice good hygiene. This condition has most often been seen in children, but also in adults, with equal incidence in men and women.[3] It is especially observed in the face, neck, and trunk or ankles in young patients, although unusual sites such as scalp, lips, chest, axilla, back, umbilical area, pubis, arms and legs have been reported.[2,4-8] The plaques can be localized or generalized. Currently, the etiology remains unknown, but it is hypothesized that TFFD is a disorder of abnormal and delayed keratinization with incomplete keratinocyte maturation and retention of keratinocytes and melanin within the epidermis.[4,5,7,8] TFFD does not cause any symptoms. The disease is commonly considered as benign, but cosmetic distress in affected patients can occur.[1,2] Brown, dirt-like discoloration that cannot be removed by bathing with water or rubbed off with routine detergent soap.[3] Some

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reports have focused on sunlight exposure as a triggering factor.[1,5,7] An association with atopic dermatitis and xeroderma has been reported.[4] Even though TFFD was reported in association with pregnancy,[9] it can be assumed that there is no etiological association, but simply a higher rate of detection during this phase of life. An association between acanthosis nigricans and insulin resistance, obesity has been reported.[10] The possibility for a syndromic disease has been postulated, which will remain disputed though until further evidence arises.[6,8] Terra firma-forme dermatosis and dermatosis neglecta are widely considered to be synonymous, but some authors have proposed a separation of the terms. Although, isolated cases of terra firma-forme dermatosis have been reported in infants, it occurs mainly in older children and adolescents with characteristic hygiene habits and with a distinctive distribution of the lesions.[6] Whereas, dermatosis neglecta affects patients of any age whose hygiene in specific areas is insufficient.[11] Skin lesions of dermatosis neglecta can be routinely removed while bathing.[12,13] Histopathologically, prominent lamellar hyperkeratosis with focal areas of compact whorled orthokeratosis,[1,2] papillomatosis, mild acanthosis and deposition of keratotic material within the valleys between the papilla.[8] There is no parakeratosis.[4,8] Other findings with special stains include increased melanin content in the compact hyperkeratotic and basal areas of the epidermis (Fontana-Masson).[1,2] Dermatoscopic examination of the lesions show polygonal brown scales, arranged in a mosaic pattern.[14] The diagnosis of TFFD is confirmed by forceful rubbing with a gauze pad immersed into 70% isopropyl alcohol or ethyl alcohol.[1,2,5] This diagnostic test prevents unnecessary laboratory work-up or biopsy.[4] In addition, it offers a magical and prompt therapeutic cure for the disorder.[1,2] A pink normal skin underneath is exposed after the wiping procedure.[8]

CASE REPORT

A 22-year-old man presented at the department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University, Bangladesh with multiple asymptomatic dirty brown-muddy gray colored plaques on his forehead, nose and both cheeks for 2 years. Despite having good hygiene, including showers and washing with various medicated soaps 2 to 3 times a day no improvement had been observed. The patient was non-atopic, healthy, and had not been receiving any topical and systemic medication for this condition. Physical examination revealed brownish, hyperpigmented plaques and patches that were slightly verrucous in some areas. The lesions were distributed symmetrically on the forehead, anterior aspect of neck, cheek and nose.(Fig. a, b, c) There were also islands of normal skin within the patches. Dermoscopy revealed no melanocytic pattern of hyperpigmentation and no vascular abnormalities, but there were polygonal areas of brownish pigmentation that followed a linear pattern in some areas and spared the natural folds of the skin.

Histopathological examination revealed prominent lamellar hyperkeratosis with orthokeratotic

Fig. 1 (a,b,c) showing a 22-year-old man with multiple dirty brown plaques on face

whorls; keratin globules were prominent in the stratum corneum, increased melanin in the basal layer, papillomatosis and acanthosis was also seen.(Fig. d)

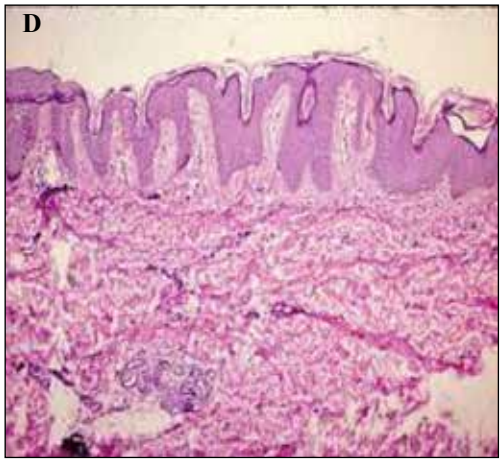

Fig. 1 (d) histopathological findings of skin lesion.

Skin scrapings for KOH examination revealed no fungal components. With a suspicion of TFFD, swabbing with 70% isopropyl alcohol pads was performed that completely removed the lesions confirming the diagnosis.

DISCUSSION

Antonela Stiube reported a 38-year-old woman to their clinic with brown skin eruptions on her areolae, which first appeared in puberty. The patient reported that black crusts erupt on the nipples and even without therapy over time fall off. As a consequence, mildly painful wounds would arise. Now that she was nursing her baby, the crusts were detaching more rapidly. She feared that the baby's health could be affected, which lead her to seek consultation. At presentation, symmetrical, dark-brown, hyperkeratotic plaques on the areolae were noted. The patient reported no pruritus and no pain associated with it. Due to the presentation, they considered TFFD, dermatosis neglecta, an atypical presentation of acanthosis nigricans, or dermatosis papulosa nigra. Rubbing of the skin eruptions with gauze immersed in 70% isopropyl alcohol cleared the skin immediately and almost completely, which confirmed the diagnosis of TFFD. Further care with Vaseline was recommended, and the patient was advised to repeat the treatment with isopropyl alcohol in case of recurrence of the skin changes.[15]

Aslan et al evaluated a retrospective analysis of patients with a diagnosis of TFFD between June 2013 and February 2016. There were 79 patients with TFFD; 70 were children (aged 1 to 17 years), and 9 were adult (aged 18 to 42 years). Fifty-one (65.6%) of the patients were female. Accompanying dermatological diseases Atopic dermatitis (7.6%), Acne (3.8%), Rosacea (1.3%), Seborrheic dermatitis(1.3%) and Vitiligo (1.3%). Regarding systemic diseases allergic rhinitis/allergic asthma (11.4%), urinary tract infection (6.3%), malignancy (osteosarcoma and chronic myeloid leukemia) (2.5%) and hypothyroidism (2.5%). In 34.2% of cases, lesions were found in more than one area of the body. They appeared on the trunk (27.8), extremities (26.6), folded zones (8.9), and head and neck (2.5) in decreasing order. The lesions were symmetrically distributed in 61 (77.2%) patients. The lesions disappeared after being wiped with 70% isopropyol alcohol-soaked gauze.[16]

Martín-Gorgojo A, et al. reported a 10-year-old girl with no relevant past medical history. Her mother brought her to their unit for assessment of a persistent asymptomatic skin rash that had appeared several months earlier. Physical examination revealed brownish, reticulated, macular areas that were slightly papillomatous to the touch in some zones. The lesions were distributed symmetrically and dermatoscopy revealed no mela-

nocytic pattern and no vascular abnormalities, but there were polygonal areas of brownish pigmentation that followed a linear pattern in some places and spared the natural folds of the skin. Swabbing with a cotton ball soaked in 70% ethyl alcohol was performed as a diagnostic test that doubles as treatment. This procedure cleared the lesions, revealing skin of a normal appearance in the treated area. They established a diagnosis of dermatosis neglecta and instructed the patient to apply an exfoliant cream containing keratolytic agents (silica granules, salicylic acid, triclosan, aluminum oxide, and zinc oxide) and then wash the affected areas in order to accelerate the healing and complete resolution of the lesions.[17]

Erkek, et al. describe two patients with terra firma-forme dermatosis in the setting of xerosis cutis and atopic dermatitis. From a clinical point of view, they lay emphasis on its unique expression and diagnosis/treatment. From a histological perspective, they highlighted its resemblance to dermatosis neglecta and speculate on the role of 'neglect' in a patient with seemingly adequate hygiene. Erkek, et al. presented one case of a 30-year-old woman with a 6-months history of persistent scaly pigmented patches on the armpits and torso. Subjective complaints were intense pruritus and cosmetic embarrassment. Her medical history was notable for xerosis cutis since childhood. She had been instructed by several doctors to avoid frequent bathing and to keep away from abrasive sponges and scrubs. She was bathing with tepid water without cleansers twice a week and promptly applying 10% urea containing emollients on moist skin. Dermatological examination revealed brown-grey patches on both sides of the abdomen and axillae. There were islands of normal skin within the patches. Initial tentative clinical diagnoses were terra firma-forme dermatosis, pomade crust, confluent and reticulated papillomatosis and acanthosis nigricans. Histopathological examination displayed prominent lamellar hyperkeratosis with whorls, keratotic plugging of follicular orifices, keratin globules in the stratum corneum, papillomatosis and sharp invaginations between the papilla, increased melanin pigment in the basal layer and minimal lymphocytic liquefaction. Focal findings were noted in dermis; i.e., edema in the papillary dermis, pigment-laden macrophages, perivascular lymphocytic infiltration and erythrocyte extravasation. A Fontana-Masson stain showed focally increased melanin pigment in the basal layer of the epidermis. Complete cure was attained by powerful swabbing of the skin with isopropyl alcohol-soaked gauze pads. Erkek, et al. presented case 2 as a 20-year-old man with a 2-year history of itchy, thick and dark skin lesions. Previously he had received several prescribed and OTC topical medications without beneficial effects. His medical history was notable for atopic dermatitis. His washing habits consisted of daily shower and instant application of 10% urea containing emollients. On dermatological examination there were reticular, brown-grey, pigmented plaques with a chamois leather appearance on the neck and upper back. Features of atopic dermatitis was also observed, i.e., Dennie-Morgan periorbital skin folds, allergic shiner, xerosis cutis and keratosis pilaris. Biopsy was not attempted, since a 'wipe test' with 70% ethyl alcohol accomplished clearance of lesions. A diagnosis of TFFD was established and patient was sent home with adequate information on how to get rid of this 'dirty' appearance. The patient was satisfied with the outcome of this in-

tervention (urea containing emollients) and there was no recurrence at 6 months of follow-up.[18]

Chetna Singla reported a 15-year-old girl to the tertiary care center with crusting of the both upper and lower lips since the last 3–4 days associated with burning sensation. On careful physical examination, multiple lesions that were bilaterally symmetrical were present in the form of dirty brown-colored adherent crusts over forehead, bilateral cheeks covering the malar area, and the temples. On inquiring, she gave a history of such asymptomatic dirty brown plaques over the face gradually progressive in nature and present for the last 5 months. Lesions started over the forehead and further lesions started appearing over both cheeks. When asked about cleanliness, she reported trying to clean the lesions with water only, but with no success. Throughout this period of 5 months, she never used soap. Although, she reported the presence of acne before the onset of the present lesions. Under considerable distress, as the lesions were present over the face, she had consulted many physicians but with no diagnosis and had given up hope of getting treated for the same. After thorough examination, differential diagnosis of the plane warts, seborrheic keratosis and TFFD were kept. Biopsy was not done as it was refused by the patient. Their diagnosis of TFFD in this patient was confirmed when the lesions completely cleared off following rubbing with a spirit swab, with completely normal underlying skin. The relieved and considerably happier patient was sent home with instructions to use a spirit swab in case of any recurrence of TFFD. No recurrence of TFFD was seen.[13]

CONCLUSION

Terra firma-forme dermatosis (TFFD) can be easily diagnosed with careful evaluation and can be treated, but if the physician does not consider it early in the diagnosis, unnecessary diagnostic and treatment procedures may result.

Conflict of interest: Author declares no conflict of interest.

Ethical consideration: Informed consent was taken from the patient for publication of this case report and accompanying images.

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