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Received 28 September 2021; accepted 8 November 2021; Available online 14 December 2022

https://doi.org/10.1016/j.abd.2021.11.010

Keratotic papules on the thigh: underrecognized skin manifestations of dermatomyositis or Wong-type dermatomyositis?☆

Dear Editor,

A 53-year-old female, who was admitted to the Rheumatology Department of our hospital, complaining of muscle weakness and joint pain, was referred to us regarding the skin symptoms which appeared 4 months previously. Physical examination revealed forehead and nasolabial erythema, periungual erythema, scaly erythema on the left knee, keratotic erythematous patch on the dorsum of metacarpophalangeal joint (Gottron's sign), and keratotic lesion on the radial aspect of the second fingers (mechanic's hand). In addition, small reddish, keratotic papules and papular erythemas were disseminated on the lateral aspects of the right thigh (Fig. 1). Laboratory examination showed elevation of aspartate aminotransferase (180 U/L), alanine aminotransferase (100 U/L), lactate dehydrogenase (487 U/L), creatine kinase (3506 IU/L), aldolase (35.8 U/L), and myoglobin (1230 ng/mL). The antinuclear antibody was positive (1:1280, speckled). Serum antibodies against TIF-1γ (48.0 index; normal <32) and Mi-2 (>150 index; normal <53) were elevated, whereas both anti-Jo-1 and anti-MDA-5 antibodies were normal. Serum KL-6 level was normal, and no interstitial lung disease (ILD) was detected. Skin biopsy revealed a keratotic plug, epidermal atrophy, liquefaction of the basal layers, individual cell keratinization, and infiltration of inflammatory cells in the papillary dermis (Fig. 2). No internal malignancy was detected by a detailed examination. An electromyogram of the biceps muscle revealed a myogenic

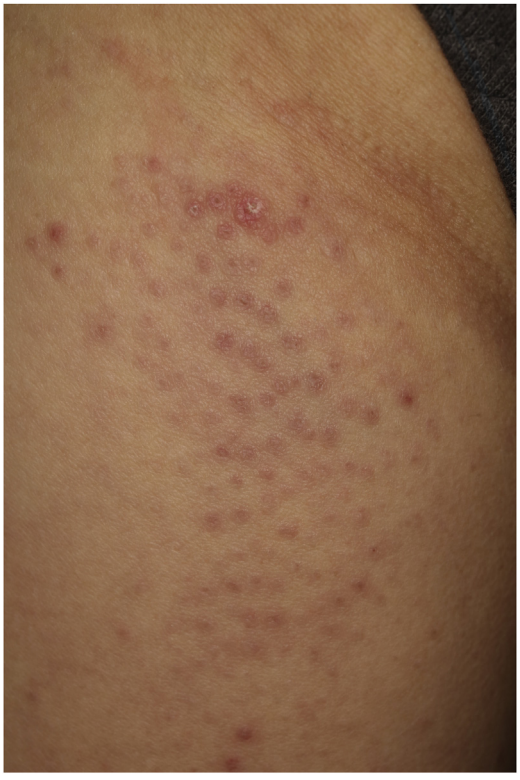

Figure 1 Clinical features of follicular papules on the thigh.

☆ Study conducted at the Fukushima Medical University, Fukushima, Japan.

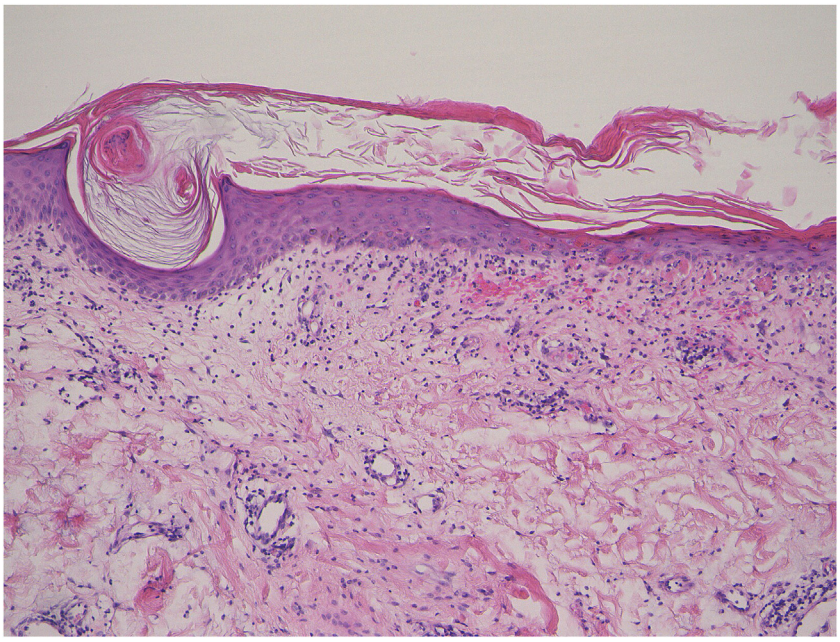

Figure 2 Histological features showing keratotic plug, epidermal atrophy, liquefaction of the basal layers, individual cell keratinization, and mononuclear cell infiltration in the upper dermis: (Hematoxylin & eosin ×100).

pattern. MRI showed high signal intensity in the upper arm and thigh muscles. She was successfully treated with methylprednisolone pulse therapy followed by oral prednisolone.

Wong-type dermatomyositis (DM) is characterized by keratotic erythematous follicular papules, which histopathologically show follicular hyperkeratosis with keratotic plugs filling dilated follicular infundibula.[1] Whether this type is a distinct subtype of DM or not is still controversial. Our case presented various skin symptoms related to DM, and thus we think that keratotic papules on the thigh in the present case may better be considered as a rare manifestation of DM, rather than a Wong-type DM.

The association between Wong-type DM and either malignancy or ILD remains unclear. As far as we searched, 35 cases of Wong-type DM have been reported in the literature, which included cases with pityriasis rubra pilaris-like appearance, and cases with follicular keratotic papules on the buttock or extremities along with other various skin manifestations compatible with DM. Wong et al.[1] reported that 52% of 23 patients (11 Wong-type and 12 typical DM) had complications of malignancy, but the frequency of either malignancy or ILD in Wong-type DM is uncertain. After excluding this report, we examined the 24 cases with Wong-type DM. Internal malignancy was observed in 3 cases among 17 cases with a description of malignancy (7 were unknown).[2–4] And ILD was observed in only 1 case among 8 cases (16 were unknown). Our case did not have either ILD or internal malignancy; however, the patient is under careful follow-up because she had a positive anti-TIF-1γ antibody. The anti-TIF-1γ antibody is closely related to cancer-associated DM, and adult patients with this antibody are reported to have malignancy at a rate of 65%.[5] Further studies are necessary to examine the relationship between myositis-specific antibodies and keratotic papular lesions.

Financial support

None declared.

Authors' contributions

Miyuki Yamamoto: Data collection, analysis, and interpretation; preparation and writing of the manuscript.

Toshiyuki Yamamoto: Approval of the final version of the manuscript.

Conflicts of interest

None declared.

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Received 23 March 2021; accepted 30 March 2021;
Available online 19 December 2022

https://doi.org/10.1016/j.abd.2021.03.018

Primary ductal carcinoma of ectopic breast☆

Dear Editor,

Ectopic breast carcinoma accounts for approximately 0.3% to 0.6% of all breast cancers, with 95% arising from aberrant breast tissue.[1,2] Clinical diagnosis may be delayed due to the atypical location, similarity to other diseases, and flawed laboratory tests.[3] This case report describes a patient with primary ductal carcinoma of the ectopic breast in the axilla, diagnosed and treated as hidradenitis suppurativa (HS).

A 62-year-old female patient presented with an erythematous, well-defined nodule measuring 1.5×2.0 cm, with a retracted center, painful and hard on palpation in the left axilla (Fig. 1).

The clinical diagnosis suggestive of HS was confirmed by ultrasound, which showed an area of subcutaneous hypoechoic thickening and a nodular area. Topical and oral antibiotics were prescribed for 14 days, in addition to intralesional corticosteroid injections, without improvement.

Two 0.5 cm elliptical incisional biopsies were performed and anatomopathological analysis showed preserved epidermis and the presence of rows and clusters of atypical epithelial cells in the dermis (Fig. 2), while the immunohistochemistry showed positivity for pankeratin, suggestive of cutaneous metastasis.

The neoplastic screening did not disclose a primary site. Therefore, total excision of the lesion was performed, which showed dermal infiltration of carcinomatous cells over ectopic mammary gland tissue (Fig. 3). Immunohistochemistry was positive for estrogen and negative for progesterone and human epidermal growth factor receptor-type 2 (HER-2). It was concluded that it was a primary ectopic breast carcinoma with characteristics of invasive ductal carcinoma.

The patient was referred to the mastology and oncology service, where surgical margins were enlarged and an ipsilateral axillary lymphadenectomy was performed, due to the presence of lymph node metastasis. She underwent complementary treatment with radiotherapy and anastrozole. There has been no recurrence one year on outpatient follow-up and control mammography.

Ectopic breast tissue is subject to the same pathophysiological processes as the topical breast, but malignant changes are more frequent than benign ones.[1] It can consist of glandular tissue, nipple and areola. It is subdivided into supernumerary breast or aberrant breast tissue. The latter is characterized by the presence of an isolated mammary gland, close to the topical breast and without communication with overlying skin.[1]

Ectopic breast cancer predominates in the female sex. The axillary region is most frequently affected and infiltrating ductal carcinoma accounts for 79% of cases.[1,4] The most common clinical manifestation is the presence of a unilateral, subcutaneous, irregular, erythematous, indurated nodule showing progressive growth, with or without a nipple and areola.[5] Ultrasonography is the initial preferential examination, which may show an irregular, hypoechoic, heterogeneous, poorly-defined nodule; the accessory mammary gland may be detected.[5] The diagnosis is usually a late one, with an average delay of 40 months, which can lead to worse prognosis. The confirmation is achieved with histopathological analysis.[3,5] The management of ectopic breast carcinoma follows conventional breast cancer treatment and staging. There is no consensus on the prophylactic excision of the ectopic breast tissue.[3,5]

Although it is a rare condition, it is essential to recognize it and consider tumors along the milk line. It is important to emphasize the importance of the histopathological analysis, even in the presence of a probable benign lesion.

Financial support

None declared.

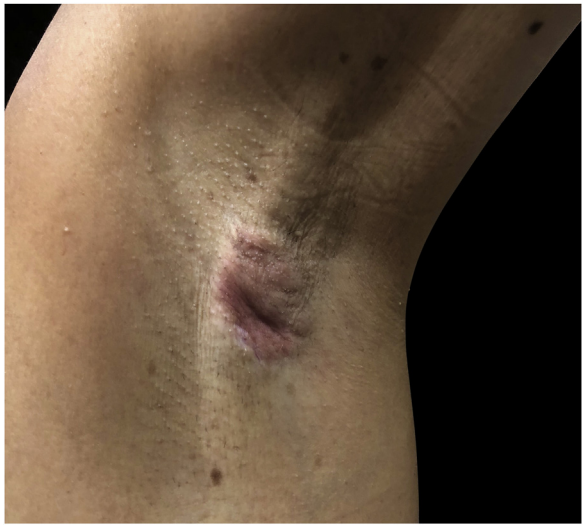

Figure 1 Erythematous, mobile nodule with retraction area.

☆ Study conducted at the Hospital Federal de Bonsucesso, Rio de Janeiro, RJ, Brazil.