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2.学会発表

H.知的財産権の出願・登録状況

1.特許取得

なし。

2.実用新案登録

なし。

3.その他

なし。

III. 研究成果の刊行に関する一覧表

雑誌

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Sugiura K, Nakasuka A, Kono H, Kono M, Akiyama M.	Impetigo herpetiformis with IL36RN mutations in a Chinese patient: A founder haplotype of c.115+6T>C in East Asia.	J Dermatol Sci	79(3)	319-20.	2015
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Himoto T, Tanaka N, Saito A, Muro Y, Sugiura K, Tani J, Miyoshi H, Morishita A, Yoneyama H, Haba R, Masaki T.	Diversity of humoral responses to thecentromere proteins among HCV-relatedchronic liver disease, PBC and AIH patients.	Clin Res Hepatol Gastroenterol	39(2)	222-9	2015
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IV. 研究成果の刊行物・別刷

JAPANESE DERMATOLOGICAL ASSOCIATION
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LETTER TO THE EDITOR

Unilateral generalized linear porokeratosis with nail dystrophy

Dear Editor,

Porokeratosis is a disorder of abnormal keratinization characterized clinically by well-defined annular lesions with hyperkeratotic ridges. Here, we report a case of unilateral generalized linear porokeratosis with dystrophy of the toenail.

A 36-year-old man showed unilateral, brownish-red plaques and round macules on the right side of his trunk and the right extremities, with slight itching (Fig. 1a). The eruptions had been present since childhood. At elementary school age, toenail dystrophy occurred after the appearance of cutaneous hyperkeratotic lesions. The lesions consisted of multiple pigmented hyperkeratotic and verrucous papules and plaques with a sharply demarcated border in a linear distribution. The lesions were distributed along the lines of Blaschko (Fig. 1a). Some of the macules had coalesced, and in some parts of the lesions, annular macules with hyperkeratotic rims were also seen. The linear lesions on the right leg were connected to the lesions in the toes. The nail of the right first toe was dystrophic and showed irregular grooving and pterygium (Fig. 1b,c). No bony abnormality of the affected toe was detected by X-ray examination (Fig. 1d). The patient had no cutaneous lesions on the hands. No nail dystrophy was seen on any finger. There were no mucosal or visceral abnormalities. He had neither immunosuppression nor any systemic disease associated with porokeratosis. He had no family history of porokeratosis.

Skin biopsies were taken from affected sites of the abdomen and the leg. Cornoid lamellae were found in the interfollicular epidermis (Fig. 1e).

From these clinical and histopathological findings, this case was diagnosed as linear porokeratosis.

Two forms of linear porokeratosis exist. In the rare generalized form, the lesions are multiple, and they affect several extremities and involve the trunk. Unilateral generalized linear porokeratosis is extremely rare.[1]

Nail involvement in linear parakeratosis is rare. To date, there have been only a few case reports of onychodystrophy in linear porokeratosis.[2,3] In one case, there was bony narrowing of the digits.[3] Six main variants of porokeratosis have been described.[4] In those variants, nail involvement was reported not only in linear porokeratosis, but also in porokeratosis of Mibelli[4] and porokeratosis plantaris palmaris et disseminata.[5] Though the exact pathogenesis of dystrophic nail is not yet understood, it is possible that nail changes occur through the involvement of the nail matrix and nail bed by atypical hyperproliferative keratinocytes, ultimately resulting in destruction of the whole nail.[2] Cases with nail involvement in linear porokeratosis are not as rare as those in the other porokeratosis variants. We speculate that this is because many cases of linear porokeratosis involve the distal portions of the extremities.

Local porokeratosis can be treated by topical therapy, such as with 5-fluorouracil, vitamin D_3 analogs, imiquimod or tretinoin creams.[4] For generalized porokeratosis, topical therapies may show variable effectiveness, but they are not practical. Therefore, systemic retinoids were anecdotally used for diffuse porokeratosis.[1] Our patient declined etretinate, the only internal retinoid available in Japan.

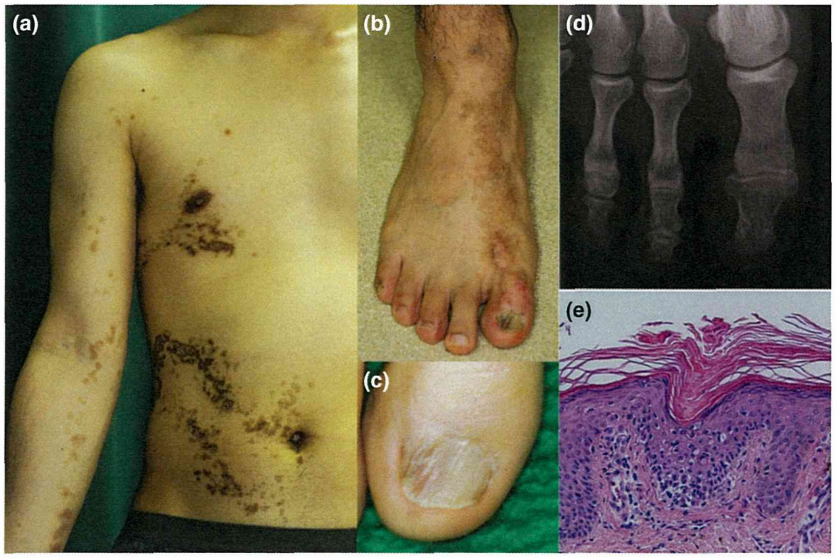

Figure 1. Unilateral generalized linear porokeratosis with nail dystrophy. The clinical and histopathological features of the current case. (a) A clinical photograph indicating the distribution pattern of the lesions. (b) A clinical photograph of the continuous linear lesion from the right dorsum of the foot to the toenail. (c) Nail dystrophy and pterygium on the right hallux next to the lesion. (d) An X-ray photograph of the affected toe shows no bony abnormalities. (e) A microphotograph of hyperkeratosis and a cornoid lamella in the interfollicular epidermis (hematoxylin–eosin stain, original magnification ×200).

Correspondence: Michihiro Kono, M.D., Ph.D., Department of Dermatology, Nagoya University Graduate School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya 466-8550, Japan. Email: miro@med.nagoya-u.ac.jp

To our knowledge, this case is the first report of unilateral generalized linear porokeratosis with nail dystrophy.

CONFLICT OF INTEREST: None declared.

Michihiro KONO,[1] Nozomi YOKOYAMA,[1] Yasushi OGAWA,[1] Hiromichi TAKAMA,[2] Kazumitsu SUGIURA,[1] Masashi AKIYAMA[1]

[1]*Department of Dermatology, Nagoya University Graduate School of Medicine, Nagoya, and* [2]*Takama Dermatology Clinic, Kasugai, Japan*

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