

SHORT COMMUNICATION

Disseminated Giant Hyperkeratotic Porokeratosis and Treatment with Acitretin: A Case Report

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Porokeratosis (PK) is a disorder of clonal hyperproliferation of keratinocytes. It has several clinical manifestations characterised by variably sized papules or plaques with a fine peripheral keratotic rim and central atrophy accompanied by a histological finding of cornoid lamella (1).

Cutaneous lesions vary in their appearance and distribution. Disseminated superficial actinic porokeratosis (DSAP) is characterised by numerous papules and plaques distributed over sun-exposed sites. Coexistence of disseminated superficial and disseminated warty PK is very rare (2).

A recent review on DSAP found no published controlled trials evaluating its treatment (3). The knowledge base for treatment is derived exclusively from case reports. To our knowledge, none of the published cases of disseminated superficial and warty PK describe the results of systemic treatment with acitretin.

CASE REPORT

An 86-year-old man presented to the University of Tartu Dermatology Clinic in late 2012 with a 10-year history of a persistent progressive eruption on his gluteal region, thighs and legs. He was treated for presumed verrucous warts with salicylic acid ointment and cryotherapy without success.

He reported that the eruption first appeared on the dorsal aspect of his foot in the form of 1–2 cm plaques. The patient denied any pruritus. However, extensive hyperkeratotic plaques over the ankles caused him considerable discomfort. There were no similar familial cases.

On clinical examination, the patient appeared well. A small (not tender, freely moveable) lymph node was palpable in the left axillar region. Cutaneous examination revealed: (i) several plaques (4–6 cm in diameter) with gross hypertrophic verrucous surfaces confined to the periphery (Fig. 1A) on both legs; (ii) multiple brownish to pink annular macules with raised peripheral ridges (2 mm to 7 cm), with an atrophic and desquamative centre and well-demarcated keratotic and filiform border on the extensor surfaces of the thighs, and on the extensor surface of the patient's right arm; (iii) brownish verrucous annular plaques in the gluteal region (bilaterally).

A biopsy was taken from 2 types of lesions: one from the margin of a delicate annular plaque on the left shin, and one from the well-defined verrucous margin of a lesion on the patient's left foot. Histological examination confirmed the diagnosis of PK in both specimens (Fig. S1¹). PK has also been reported in association with dermal amyloid deposition as a result of apoptosis of keratinocytes (5), and this was also documented in this case.

A comprehensive metabolic screening was normal. Abdominal ultrasonography showed a left polycystic kidney but no enlarged lymph nodes in the abdominal or pelvic cavities. Chest radiograph was normal. Human papilloma virus test (PCR based) from the lesions on the foot was negative.

A complete blood count revealed marked changes in the white blood cell count (WBC; 21.53 (normal range



Fig. 1. Clinical images of some of the skin lesion at the time of diagnosis (A) and after 8 weeks treatment with acitretin (B).

¹<http://www.medicaljournals.se/acta/content/?doi=10.2340/00015555-1908>

3.5–8.8 E9/l) with neutropenia (13% (normal range 40–80%) and lymphocytosis (85% (normal range 20–40%) together with an increase in the absolute lymphocyte count (18.41 E9/l)). The patient was referred to a haematologist and was diagnosed with chronic myeloid leukaemia (treatment was not initiated).

For the treatment of PK, acitretin (30 mg daily) was prescribed. After 8 weeks (May 2013) marked improvement was observed for the grossly verrucous lesions (Fig. 1B) and treatment continued. The patient's symptoms resolved (he had no discomfort). At 16 weeks follow (August 2013) up further improvement was observed. Since then the contact with patient is lost.

DISCUSSION

Reaching a final diagnosis in a case of multiple types of PK is somewhat arbitrary. Our patient had clinical signs of (i) PK of Mibelli (verrucous patches in the gluteal region); (ii) disseminated superficial actinic PK (on the shins and forearms); (iii) and giant hyperkeratotic plaques on the ankles and feet. Whether the latter (giant hyperkeratotic plaques) constitute an extension of the disseminated superficial actinic PK (2, 4, 5) or disseminated PK of Mibelli (6, 7) is hard to determine.

The different types of PK are differentiated solely on clinical criteria. DSAP is the most common form of PK. In addition, disseminated forms have also been described for PK palmaris et plantaris (1) and PK Mibelli (6, 7).

Apart from these clinical variants, a number of morphological forms – giant PK, hypertrophic verrucous, and reticulate PK have been reported (1). Hyperkeratotic or giant forms of PK have been described for PK of Mibelli (6–9), disseminated superficial actinic PK (2, 4, 5), and linear PK (10).

The use of oral retinoids (etretinate, acitretin) in PK has yielded conflicting results. Whereas good results have been obtained in some patients (11, 12), worsening of lesions during the treatment (13) and unusual side effects (developing digitate keratoses following treatment with etretinate) (14) have also been reported. Intuitively, systemic treatment has benefits over local treatment for disseminated and/or markedly hyperkeratotic lesions.

Previous reports of the coexistence of different forms of PK (disseminated PK, DSAP, PK of Mibelli) and haematological malignancies have described PK emerging after treatment for haematological malignancy. To our knowledge there is a scarcity of case reports on PK emerging before the diagnosis and treatment of haematological malignancy. However, it has been speculated that immunosuppression associated with malignancy and chemotherapy may exacerbate or initiate the development of PK in patients predisposed to alterations in cutaneous growth dynamics (15).

In conclusion, coexistence of 2 or more variants of PK in a single patient can occur and whether the different morphological forms of PK (giant, hypertrophic verrucous) consist different forms of PK or a variation on the clinical spectrum of disease warrants further research. Evidence for the treatment of PK is equivocal. Based on our experience in this case, systemic treatment with acitretin (especially for the disseminated warty forms of PK) should be considered as an option.

The authors declare no conflicts of interest.

REFERENCES

1. Sertznig P, von Felbert V, Megahed M. Porokeratosis: present concepts. *J Eur Acad Dermatol Venereol* 2012; 26: 404–412.
2. Kanak K, Jaiswal AK, Reddy P. Disseminated superficial and warty type of porokeratosis: a rare coexistence. *Indian J Dermatol* 2011; 56: 576–577.
3. Skupsky H, Skupsky J, Goldenberg G. Disseminated superficial actinic porokeratosis: a treatment review. *J Dermatolog Treat* 2012; 23: 52–56.
4. Jang KA, Choi JH, Sung KJ, Moon KC, Koh JK. The hyperkeratotic variant of disseminated superficial actinic porokeratosis (DSAP). *Int J Dermatol* 1999; 38: 204–206.
5. Schuldenfrei JA, Fix LW. Disseminated superficial actinic porokeratosis with hyperkeratotic lesions. *Cutis* 1984; 33: 369–370.
6. Marghescu S, Anton-Lambert I, Melz-Rothfuss B. Disseminated bilateral hyperkeratotic variant of porokeratosis Mibelli. *Arch Dermatol Res* 1987; 279: 39–47.
7. Zhou C, Jin Y, Zang D, Zhang J. Disseminated bilateral hyperkeratotic variant of porokeratosis Mibelli with pruritus. *Eur J Dermatol* 2011; 21: 1007–1009.
8. Schaller M, Korting HC, Kollmann M, Kind P. The hyperkeratotic variant of porokeratosis Mibelli is a distinct entity: clinical and ultrastructural evidence. *Dermatology* 1996; 192: 255–258.
9. Uenishi T, Teramura K, Kitamura M, Fujii N, Nakanishi G, Tanaka T, Uehara M. Hyperkeratotic variant of porokeratosis Mibelli with dermal amyloid deposits. *J Dermatol* 2010; 37: 475–479.
10. Lucker GP, Steijlen PM. The coexistence of linear and giant porokeratosis associated with Bowen's disease. *Dermatology* 1994; 189: 78–80.
11. Ludera-Zimoch G, Rubisz-Brzezińska J. [A case of superficial disseminated actinic porokeratosis treated with an aromatic retinoid]. *Przeegl Dermatol* 1989; 76: 54–57 (in Polish).
12. Garg T, Ramchander, Varghese B, Barara M, Nangia A. Generalized linear porokeratosis: a rare entity with excellent response to acitretin. *Dermatol Online J* 2011; 17: 3.
13. Knobler RM, Neumann RA. Exacerbation of porokeratosis during etretinate therapy. *Acta Derm Venereol* 1990; 70: 319–322.
14. Carmichael AJ, Tan CY. Digitate keratosis – A complication of etretinate used in the treatment of disseminated superficial actinic porokeratosis. *Clin Exp Dermatol* 1990; 15: 370–371.
15. Lederman JS, Sober AJ, Lederman GS. Immunosuppression: a cause of porokeratosis? *J Am Acad Dermatol* 1985; 13: 75–79.