Coexistence of Linear Porokeratosis and Disseminated Superficial Actinic Porokeratosis: A Type 2 Segmental Manifestation

Oscar Suárez-Amor, Mercedes Pereiro-Ferreirós, Manuel Ginarte, Carmen Peteiro and Jaime Toribio
Department of Dermatology, Faculty of Medicine, San Francisco sn. ES-15782, Santiago de Compostela, Spain. E-mail: mejaime@usc.es
Accepted September 28, 2006

Sir,

Six main clinical forms of porokeratosis are recognized: classical porokeratosis or porokeratosis of Mibelli, punctate palmoplantar porokeratosis, linear porokeratosis (LP), disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis (DSAP), and disseminated palmoplantar porokeratosis (1). The coexistence of more than one form in a single individual is infrequent. We describe here a 48-year-old woman who had a small LP lesion since infancy and developed DSAP in her fifth decade of life.

CASE REPORT

A 48-year-old Caucasian woman without relevant antecedents presented with pale rough-surfaced lesions on her forearms and legs, first noticed 4 years previously. Over the preceding 2 years they had become pruritic, particularly during the summer, when they increased in number. She had no family history of similar lesions. Physical examination revealed small plaques 5–10 mm in diameter on the extensor surfaces of the forearms and legs, clearly delimited, rounded or oval, with lighter-coloured and slightly atrophic centres, and a hyperpigmented keratotic outer ring. These lesions were not confluent and did not show any clear spatial organization (Fig. 1A). In the right axillary region and extending towards the inner surface of the arm, the patient showed several lesions very similar to those on the forearms and legs, but more conspicuous and confluent, arranged in a linear pattern (Fig. 1B). The patient reported that she had had these lesions from childhood. Neither palmoplantar regions nor mucosal tissues were affected. Histopathological study of the linear lesion and one of the leg plaques confirmed the diagnosis of porokeratosis. The epidermis showed a compact column of parakeratotic cells with loss of granular layer, superficial mononuclear infiltration with areas of vacuolization in the basal layer, and presence of isolated melanophages in the papillary dermis. General examination did not detect any other manifestations, and the patient's overall health was good. Standard biochemical analyses, immunological studies and haemograms likewise did not reveal pathological findings. Treatment was started with topical 0.1% tretinoin, once daily for 3 months. This treatment had a modest effect, and was changed to cryosurgery, with acceptable results to date. We continue to monitor the patient at periodic intervals.

DISCUSSION

In the patient described here, the lesions had developed 4 years previously, were slightly pruritic, worsened on exposure to sunlight, and had gradually increased in number; by contrast, the linear lesion had arisen in infancy, and was asymptomatic and unchanging. On physical examination the lesions on her legs and forearms were subtle and non-evident, while the linear lesion was much more evident.

The coexistence of different clinical forms of porokeratosis in a single individual is uncommon (2, 3). There have been a few previously reported cases of association between LP and DSAP, mostly in children and young

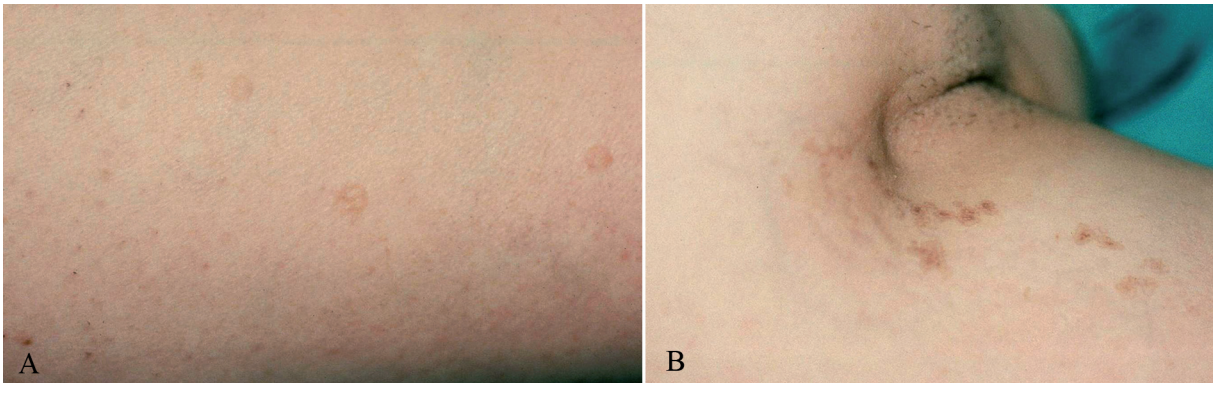

Fig. 1. (A) Plaques 5–10 mm in diameter localized on the extensor surface of the right leg, with atrophic centre and hyperpigmented keratotic peripheral ring. (B) Linear lesion, a few cm long, with polycyclic outline, in the right axillary region and on the inner surface of the arm.

doi: 10.2340/00015555-0215

adults (4–9). According to Happle (10), such cases may be a type 2 segmental manifestation of an autosomal dominant condition, in this case DSAP. This happens when there is loss of the wild-type allele in one primitive skin unit in a patient heterozygous for the underlying condition (loss of heterozygosity), giving rise to a pronounced segmental manifestation superimposed on the disseminated lesions of DSAP.

A type 2 segmental involvement in the present case is supported by the fact that the LP lesions appeared earlier in life than the DSAP lesions and were more severe, as it has been described in previously reported cases of coexistence of LP and DSAP (4–9). Furthermore, previous studies have found that both patients with LP-DSAP and patients with isolated LP often have a family history of DSAP lesions, strongly suggesting that LP is aetiologically related to DSAP (8). Porokeratosis is a genodermatosis with malignant potential, with the risk of malignancy being higher for large lesions, long-term lesions, and lesions in older or immunocompromised patients (11). Allelic loss may represent an initial stage in the process of carcinogenesis, and may explain why the linear form has the highest risk of malignancy (12).

REFERENCES

1. Schamroth JM, Zlotogorski A, Gilead L. Porokeratosis of Mibelli. Acta Derm Venereol 1997; 77: 207.
2. Pearson IC, Cliff S. Case 6: plaques extending in a linear pattern from left ankle to hip forming over a 2-year period. Diagnosis: linear porokeratosis with disseminated superficial porokeratosis erupting in pregnancy. Clin Exp Dermatol 2003; 28: 345.
3. Suh DH, Lee HS, Kim SD, Cho KH, Kim KH, Park KC. Coexistence of disseminated superficial porokeratosis in childhood with congenital linear porokeratosis. Pediatr Dermatol 2000; 17: 466.
4. Welton WA. Linear porokeratosis in a family with DSAP. Arch Dermatol 1972; 106: 263.
5. Dover JS, Miller JA, Levene GM. Linear porokeratosis of Mibelli and DSAP. Clin Exp Dermatol 1986; 11: 79.
6. Dover JS, Phillips TJ, Burns DA, Krafchik BR. Disseminated superficial actinic porokeratosis. Coexistence with other porokeratotic variants. Arch Dermatol 1986; 122: 887.
7. Morton CA, Shuttleworth D, Douglas WS. Porokeratosis and Crohn's disease. J Am Acad Dermatol 1995; 32: 894.
8. Freyschmidt-Paul P, Hoffmann R, König A, Happle R. Linear porokeratosis superimposed on disseminated superficial actinic porokeratosis: report of two cases exemplifying the concept of type 2 segmental manifestation of autosomal dominant skin disorders. J Am Acad Dermatol 1999; 41: 644.
9. Boente M, López-Baró AM, Frontini M, Asial RA. Linear porokeratosis associated with disseminated superficial actinic porokeratosis: a new example of type II segmental involvement. Pediatr Dermatol 2003; 20: 514.
10. Happle R. A rule concerning the segmental manifestation of autosomal dominant skin disorders. Review of clinical examples providing evidence for dichotomous types of severity. Arch Dermatol 1997; 133: 1505.
11. Lucker GPH, Steijlen PM. The coexistence of linear and giant porokeratosis associated with Bowen's disease. Dermatology 1994; 189: 78.
12. Happle R. Cancer proneness of linear porokeratosis may be explained by allelic loss. Dermatology 1997; 195: 20.