More than a century ago, two Italian dermatologists independently described a new keratotic disorder [1,2] which was called porokeratosis by Mibelli [1]. After these first descriptions, at least four variants of porokeratosis have been recognized. Clinically all these dermatological disorders are characterized by diffuse annular lesions surrounded by a raised, sharply marginated, keratotic border. The typical histological feature is represented by the cornoid lamella, which is a column of parakeratotic cells. The most common form is the disseminated superficial porokeratosis, characterized by diffuse small lesions, generally distributed over the legs, more rarely also on the buttocks and the arms. Idiopathic disseminated porokeratosis affects both males and females, usually around the 5th decade. The disease is transmitted as an autosomal dominant trait with variable penetrance. It has been hypothesized that porokeratosis may be triggered by a proliferation of abnormal clones of epidermal cells in response to several stimuli, the most important of which are sunlight and artificial UV [3].

A number of case reports and small series of disseminated porokeratosis have been reported in immunosuppressed patients [4–6], leading to the opinion that immunosuppression can cause porokeratosis. In a recent large series [7] 24 cases of disseminated porokeratosis in patients given immunosuppressive agents were reported; of these 13 patients had received a renal transplant. There were no apparent differences between patients given azathioprine or cyclosporin. The mean interval between the start of immunosuppression and the onset of skin disease was around 30 months. No clear-cut correlation with sunlight exposure was seen. Age, sex, area of skin involvement and dermatological appearances were similar in immunosuppressed patients and in a control group of patients with idiopathic porokeratosis, but in more than half of immunosuppressed patients there were concomitant warts and there was an actinic keratosis in about one-fourth of cases.

The mechanisms by which immunosuppressive therapy may induce disseminated porokeratosis are still unclear. Immunosuppression might trigger the expression of a mutant clone of epidermal cells, either directly or by disrupting the growth dynamics of the epidermis, or by impairing the immune surveillance. A genetic predisposition is likely. While sun exposure plays an important pathogenetic role in idiopathic cases, it does not seem to be essential for the development of porokeratosis in immunosuppressed patients.

Theoretically, the chronic growth activation of keratinocytes may expose patients with porokeratosis to an increased carcinogenetic risk. However no evidence of malignancy was seen either in a histological review of 61 patients with disseminated porokeratosis [8] or in our series of 24 patients who received immunosuppressive treatment [7]. Nonetheless, in view of the increased risk for skin malignancy caused by immunosuppression, a careful clinical surveillance of immunosuppressed patients with disseminated porokeratosis is advisable.

References

Fig. 1. Clinical features of disseminated superficial porokeratosis. Small annular lesions with keratotic borders.

Fig. 2. Typical lesions of porokeratosis associated with actinic keratosis.

Fig. 3. The typical cornoid lamella column of porokeratotic cells (more intense pink).