Abstract

Plaque-type Porokeratosis is a chronic, progressive disorder characterized by the formation of slightly atrophic patches surrounded by an elevated, warty border. It has several clinical forms including a porokeratosis of Mibelli, giant porokeratosis, linear porokeratosis, disseminated superficial actinic porokeratosis, palmoplantar porokeratosis and punctate porokeratosis. We report here a case of giant plaque-type porokeratosis in a 57-year-old patient with disease during of 27 years.

Case Report

A 57-year-old male farmer presented with hyperkeratotic irregular plaques with central atrophy, warts and papules on his lower belly, hip, inguinal, scrotal and foreskin. It has been more than twenty seven years since the onset of his diseases after right hip skin abrasion. The skin injury areas were spreading gradually and peripherally and seemed atrophies in the center, eventually there was formed a circinate and well-defined plaque surrounded by a keratotic wall, and several papule-like warts formed in the center of the skin lesion. There was no family history of similar lesions. In dermatologic examination, he had a 28×16 cm² lesion with central atrophy surrounded by a 0.5-1 cm raised hyperkeratotic wall on his lower belly and same 46×34 cm² lesion in perineal and 28×16 cm² on his lower belly, which is larger than previously reported but the surrounding wall is not up to 1 cm [2-6].

Routine laboratory test results were within normal limits, both tuberculosis and syphilis serology test resultswere negative. Chest X-ray results did not show any signs of abnormal.

The histopathologic examination of the biopsy samples from the two different morphological lesions confirmed the diagnosis of porokeratosis (Figure 2 and 3).

Discussion

Porokeratosis is a heterogeneous group of disorders that are inherited in an autosomal dominant fashion [1]. It has several clinical forms including a porokeratosis of Mibelli, giant porokeratosis, linear porokeratosis, disseminated superficial actinic porokeratosis, palmoplantar porokeratosis and punctate porokeratosis. The giant plaque-type was rarely observed in clinical practice. The pathogenesis is not well understood until recently. Immunosuppression, immunosuppressive diseases, such as AIDS; ultraviolet exposure; renal transplant patients and radiation therapy might exacerbate porokeratosis and promote the development of skin cancers [2,3]. An alternative hypothesis is that an inflammatory mononuclear infiltrate composed of helper T cells, suppressor T cells and Langerhans cells beneath the cornoid lamella may provide a mitotic stimulus for overlying keratinocytes [3].

Giant porokeratosis is a very rare form of porokeratosis. Several reports suggest that it is a morphological variant of porokeratosis of Mibelli, but others consider it as a different clinical form. It has been reported that the lesions were about 10-20 cm in diameter and the surrounding walls rose up to 1 cm [2-6]. In our case, patient has typical Mibelli lesions, with central atrophy surrounded by keratotic walls on his hips, the lesion area are up to 46×34 cm² in perineal and 28×16 cm² on his lower belly, which is larger than previously reported but the surrounding wall is not up to 1 cm [2-6].
6]. According to his medical history, the patient had no other triggering factors except for skin abrasion, and the lesions were unsetting and spreading from the injury area. There are more variant from other reports in literature, our patient had a long course up to 27 years and had no evidence to develop skin cancer, the reason might be lesions of the location is not exposed parts.

Histopathologic examination of the biopsy samples from different morphological lesions confirmed the diagnosis of porokeratosis and no evidence of skin cancer.

Due to lesion size and localization, and functional and aesthetic requirements, the optimal treatment procedure should include excision and grafting, cryotherapy, electrodesiccation, dermabrasion and CO2 laser [5,6]. But our patient refused to be treated with any of these methods. And now the patient was lost to follow-up after 1 month.

References