A Case of Squamous Cell Carcinoma Arising in Giant Porokeratosis

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We report a case of squamous cell carcinoma arising in giant porokeratosis in an 88-year-old woman. She had a 20×12 cm sized erythematous scaly patch with central ulceration on the right parietal area. On histopathologic examination, a skin biopsy specimen from the peripheral ridge of erythematous lesion revealed porokeratosis, and a specimen from the central ulceration showed well differentiated squamous cell carcinoma.
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Key Words : Giant porokeratosis, Squamous cell carcinoma.

Giant porokeratosis is a rare variant of porokeratosis and morphologically characterized by atrophic plaques, surrounded by a raised wall, mimicking porokeratosis of Mibelli[1]. It can be differentiated from the latter by the diameter of the lesion and their configuration which is usually polycyclic rather than annular. It has a well defined potential for malignancy. Malignancies have been reported in all types of porokeratosis[2]. To date there have been reports of squamous cell carcinomas, basal cell carcinomas, Bowen's diseases and metastatic squamous cell carcinomas arising in porokeratosis[3,4]

REPORT OF A CASE

An 88-year-old woman presented with an erythematous scaly patch on the right parietal area. The lesion had been growing slowly during the previous 3 years and was occasionally pruritic. Several months prior to visiting our clinic, ulceration developed in the center of the erythematous lesion. She was initially treated with various kinds of steroid creams without improvement. There was no family history and prevous history of similar lesions.

Physical examination revealed a 20× 12 cm sized, well-defined, circinate erythematous patch with an elevated delicate keratotic wall on the right parietal area. It involved the forehead, right auricle, right parietal and posterior occipital area. In the center of the erythematous patch, an ulcerated lesion with crusts, measuring 2×3 cm was noted (Fig. 1). Regional lymph nodes were not palpable and the remainder of physical examination was not contributory.

On completion of laboratory tests includiding a complete blood cell count, urinalysis, liver function test, BUN/creatinine and chest PA, the results were within normal limits or negative. A swab from a patch grew on culture candida albicans.

Biopsy specimens were obtained from the peripheral ridge of the patch and the central ulceration. The epidermis of the peripheral ridge showed hyperkeratosis and parakeratosis. There was a parakeratotic column, so-called cornoid lamella. Beneath the parakeratotic columns, there was no granular layer and some of the keratinocytes showed an eosinophilic cytoplasm. Chronic inflammatory cell infiltrate was seen in the upper dermis and perivascular areas (Fig. 2). A biopsy specimen taken from the ulcerated lesion in the center of the giant lesion showed irregular masses of

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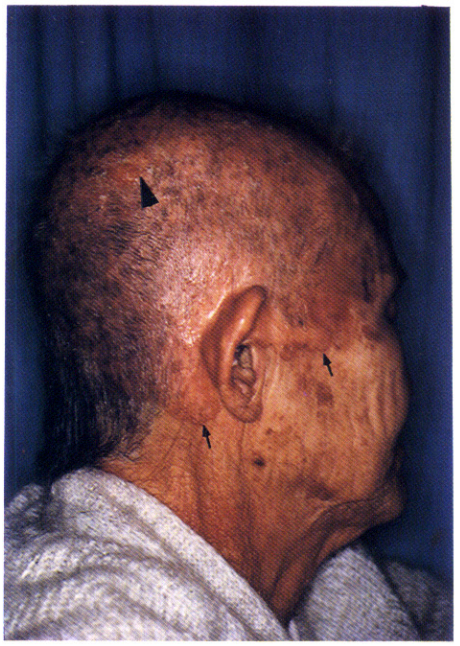

Fig. 1. A 20×12 cm sized irregularly shaped, erythematous scaly patch (arrow) with 2×3 cm sized central ulceration on right parietal area (arrow head).

epidermal cells with pleomorphic and hyperchromatic nuclei with some horn pearls and mitotic figures suggesting well differentiated squamous cell carcinoma (Fig. 3).

DISCUSSION

Porokeratosis is a rare skin disorder featuring one, several, or even hundreds of skin lesions, varying from a few milimeters to a few centimeters in diameter and is inherited as an autosomal dominant trait, occasionally occuring sporadically[5]. Several clinically distinctive forms have been described[1]: (1) porokeratosis of Mibelli (2) giant porokeratosis (3) linear porokeratosis (4) disseminated superficial porokeratosis (5) disseminated superficial actinic porokeratosis and (6) palmoplantar porokeratosis. Giant porokeratosis is morphologically characterized by atrophic plaques, surrounded by a raised wall, mimicking porokeratosis of Mibelli[6]. It can be differentiated from the latter by the diameter of the lesion which may be up to 10 to 20 cm. Histopathological features of giant porokeratosis are similar to those found in porokeratosis of Mibelli. In our case, the lesion was a well defined erythematous patch with elevated keratotic

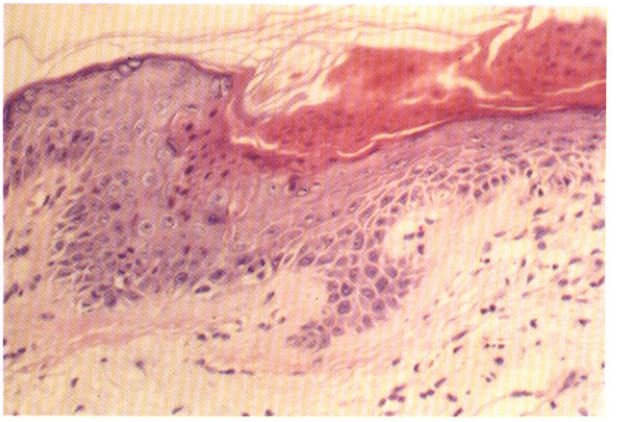

Fig. 2. A biopsy specimen taken from the pepheral ridge of the patch shows hyperkeratosis, parakeratosis and parakeratotic column with inflammatory cells infiltration in the upper dermis. (H&E, ×100).

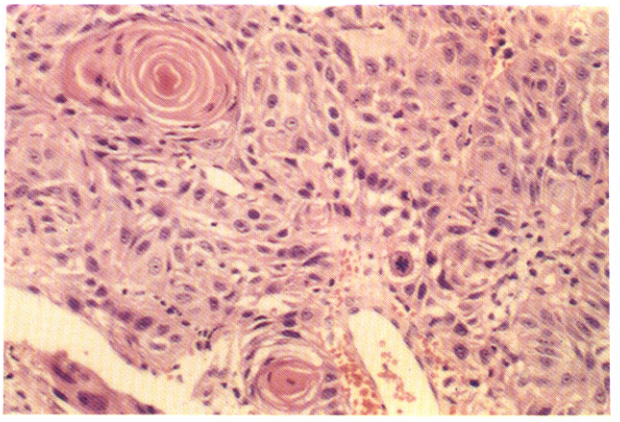

Fig. 3. A biopsy specimen taken from the central ulceration shows hyperplastic epidermal cell masses showing hyperchromasia of the nuclei. Several horn pearls and atypical mitotic figures are also seen. (H&E, ×200).

wall measuring 20×12 cm in size and demonstrated typical histopathologic changes of porokeratosis.

Porokeratosis is a well-defined potential for malignancy but the relationship between porokeratosis and skin cancer is not clearly defined[7]. Malignant degeneration occuring in porokeratosis was first noted in 1942 by Vigne[8]. The incidence of malignant changes was 7 % in 250 cases of porokeratosis reviewed in 1974[2]. Malignancies have been reported in all types of porokeratosis. To date there have been reports of squamous cell carcinomas, basal cell carcinomas, Bowen' s diseases and metastatic squamous cell carcinoma in porokeratosis[3,4]. In Korea, only three cases of malignancies associated with porokeratosis were reported. They are basal cell

epithelioma associated with porokeratosis plantaris, palmaris et disseminata, squamous cell carcinoma arising in porokeratosis of Mibelli, and squamous cell carcinoma associated with porokeratosis plantaris, palmaris et disseminata[9-11]. In our case, squamous cell carcinoma was developed in the center of the giant porokeratotic lesion. The giant form of porokeratosis is considered to be more prone to the development of malignancies and there was a report of giant porokeratosis associated with Bowen's disease[6].

The mutated clone theory has been used to explain the increased incidence of carcinomas associated with porokeratosis[12]. Reed and Leone[13] suggested porokeratosis to be a clonal disease with an inheritable tendency to develop abnormal clones. Furthermore, chromosomal instability in cultured skin fibroblasts from patients with porokeratosis was reported, suggesting a relation to the induction of malignancy[14]. External stimuli such as immunosuppressive therapy, irradiation with X-ray or ultraviolet light and repeated trauma may contribute to the expression of the disease and possibly to the malignant transformation of the abnormal epidermal clones[5]. Altered gene transcription at specific levels of epidermal differentiation results in a cellular milieu potentiating proto-oncogene amplifications and/or mutations, and the subsequent development of carcinomas[12].

We subscribe potency for malignant transformation of giant porokeratosis, confirming the need for a careful follow-up of these patients.

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