Eruptive pruritic papular porokeratosis

Sir,

A 63-year-old Chinese man was referred for a papulosquamous eruption affecting his buttocks, groin, and forearms, which had been recurring over the past 13 years. He gave the history of repeated acute episodes of intense pruritus accompanied by the appearance of new lesions. Previous treatment consisted only of topical corticosteroids prescribed by his family physician. With therapy, his itch resolved and the skin lesions subsided with hyperpigmentation. There were no known triggers for the exacerbations, he was not known to suffer from any chronic illnesses, and he was not on any concurrent medications.
Physical examination of the buttocks revealed multiple erythematous scaly annular plaques with ridged edges forming collarettes of scales, interspersed with older annular hyperpigmented macules [Figure 1]. Similar clinical findings were also noted on examination of his forearms.

A skin biopsy specimen was taken from a lesion on his right gluteal cleft. There was compact hyperkeratosis with oblique parakeratotic tiers (i.e. cornoid lamellae) arising within keratin-filled invaginations [Figure 2a]. Intracorneal neutrophils or fungal elements were not present. There was an irregular epidermal hyperplasia with an absent granular layer and dyskeratotic cells. A superficial perivascular lymphocytic infiltrate was seen in the upper dermis [Figure 2b].

The histology report was consistent with porokeratosis. Together with the clinical picture of a chronic persistent rash marked by recurrent acute pruritic exacerbations, the diagnosis of eruptive pruritic papular porokeratosis (EPPP) was made.

EPPP was first described by Kanzaki et al as a variant of disseminated superficial porokeratosis (DSP). The latter is classically an asymptomatic dermatosis. EPPP differs from DSP in this respect, as it clinically presents with repeated flares of intense pruritus together with the appearance of new lesions, as with our patient. Histological features are identical in both conditions. In a review of 8 previously reported cases, 7 out of 8 had pre-existing lesions lasting at least a year. Regression within 12 months was observed in 6 out of the 8 cases.

EPPP had been reported in a patient with colon cancer. This patient was diagnosed with cancer 6 years prior and underwent surgical resection followed by adjuvant chemotherapy. His cutaneous lesions appeared 3 years later and were initially asymptomatic, with acute pruritic lesions developing subsequently. An abnormal p53 tumor suppressor gene expression had been suggested as a possible common pathway for development of DSP and carcinogenesis, although a causal link was not conclusively established. Our patient did not have a personal history of cancer, and he defaulted follow-up before cancer screening could be instituted.

EPPP is a rare variant of porokeratosis with only around 10 cases previously reported in the English literature. Its peculiar relapsing, pruritic presentation sets this condition apart from other forms of porokeratosis. Although the lesions of EPPP tend to resolve over a period of months, topical and/or systemic treatment may be indicated for severe pruritus. In our patient, only topical steroids and oral antihistamines were prescribed, resulting in partial improvement of symptoms. Etretinate was used in a patient resulting in remission after one year. In general, however, response to therapy is variable.

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Sir,

In 1989 Crocker described Extramammary Paget’s Disease (EMPD) as a skin apocrine gland neoplasm. However, ectopic EMPD (E-EMPD) at skin places lacking apocrine glands has been described. There are 29 cases of E-EMPD reported in English and Japanese literature. We describe a new case.

A 76 year-old woman with hypertension and dyslipidemia came to our center because of an itchy, erythematous-skin colored lesion on her right cheek that has been slowly enlarging since the last year. The patient noticed that the lesion appeared after a wax-depilation burn. Physical examination showed an erythematous star-shaped macule of size 25 × 20 mm on her cheek [Figure 1] and an atypical nevus on her leg. Actinic keratosis, Bowen’s disease, and in situ melanoma were suspected so excisional biopsies were performed. Histological study concluded in situ melanoma on the leg; and histological examination of cheek’s macule showed intraepithelial accumulation of large cells, with eosinophilic cytoplasm, vesicular nucleus and prominent nucleolus [Figure 2a]. In the dermis there was a superficial infiltrate of lymphocytes and plasmocytes. On immunohistochemistry epidermal cells were cytokeratin 7 (Ck7) positive [Figure 2b] and carcinoembryonic antigen (CEA) and cytokeratin 20 (Ck20) negative. The adnexal structures were preserved. Histopathological findings were consistent with E-EMPD therefore a wide margin excision was performed.

Gynecological examination, mammography, upper and lower endoscopy and laboratory tests excluded an internal neoplasm. After four months of follow up, no recurrences have been detected.

EMPD is considered as an intraepithelial adenocarcinoma. It affects elderly people, with a mean age of 67.9 years. It affects more frequently women than men. Its most common locations are first the vulva and secondly the perineum. Its etiology is unknown. However, some authors believe E-EMPD
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