Porokeratotic eccrine ostial and dermal duct nevus with extensive linear distribution: a case report

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Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is known as an uncommon disorder of keratinization in which acrosyringium is involved; however, its causes have yet remained unknown. It is presented clinically by discrete keratotic papules on distal extremities arranged in a linear pattern along Blaschko lines. Histopathologically, it is characterized by cornoid lamella, which is almost always located overlying an acrosyringium. We present a case with an unusual presentation of extensive PEODDN lesions along the lower extremity. Our case was a young woman presented with multiple keratotic elevated papules on her left sole since her birth. In continuity with the lesions on her sole, a linear plaque was extended up to her buttock. Histopathology showed hyperkeratosis with cornoid lamella overlying the acrosyringium. The rarity of PEODDN and interesting presentation of the lesions on the sole of our patient with extension in a linear Blaschko pattern up to her buttock resembling verrucous linear epidermal nevus encouraged us to present this case.

Keywords: porokeratotic, nevus, Blaschko

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INTRODUCTION

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is an extremely rare disorder of keratinization involving acrosyringium and hair follicles. It is presented clinically by discrete keratotic papules on palms, soles or distal extremities arranged in a linear pattern along Blaschko lines. The lesions resemble comedones but are found at locations free of pilosebaceous follicles. PEODDN is present at birth in most cases, but they may appear during adulthood [1,2]. Histopathologically, it is characterized by cornoid lamella, a column of parakeratotic cells over an epidermal invagination that is almost always located overlying an eccrine duct with a dilated acrosyringium [3]. Considering that PEODDN is a very rare skin condition with less than 50 cases reported previously in the literature, we present a case with this disorder with an unusual presentation of extensive PEODDN lesions along the lower extremity [4].

CASE PRESENTATION

A 25-year-old young woman presented with multiple keratotic papules on her left sole. In

continuity with the lesions on her sole, a linear plaque following Blaschko lines was extended up to her buttock. The lesions were present since her birth and were asymptomatic. The patient had consulted to a dermatologist at the age of 9, and the mid-portion of the linear plaque on her calf and lower thigh had been resected. Nevertheless, the remaining lesions were stable until our first visit. In her clinical examination, multiple keratotic papules were present on her left sole, and a linear verrucous plaque following Blaschko's line was distributed over her left leg and her left buttock resembling linear verrucous epidermal nevus. The linear plaque was interrupted on her lower thigh and popliteal fossa due to the past resection (Figures 1a-c). Our patient was otherwise healthy with no significant past medical history; the physical examination and routine lab data were also normal.

Two incisional biopsies were taken from her skin lesions. Linear verrucous epidermal nevus, linear porokeratosis, linear lichen planus, linear psoriasis and linear Darier's disease were considered as primary clinical differential diagnoses. Histopathology showed hyperkeratosis and acanthosis with cornoid lamella overlying the acrosyringium with dyskeratotic cells in upper epidermis under the porokeratotic column (Figures 2a, b). Regarding both clinical and histopathological findings, PEODDN was determined as the proper diagnosis. The patient was referred to the laser clinic for ablative erbium laser therapy.

DISCUSSION

PEODDN is an extremely rare dermatological disorder. Its pathogenesis is unknown, but some studies regarded genetic mosaicism and abnormal keratinization as the possible mechanisms of the disease. Recently, it has been proposed that PEODDN is probably a mosaic form of Keratitis ichthyosis deafness (KID) syndrome. The mutant gene in KID syndrome, GJB2, encodes connexin 26, which is a gap junction protein [5]. The condition is usually present at birth, but cases of PEODDN presenting during adulthood have been reported [6]. It is commonly presented clinically as keratotic papules on palms and soles, but when other sites are involved, keratotic plaques are reminiscent of linear verrucous epidermal nevus, as they were seen in our patient. Furthermore, cutaneous lesions of PEODDN extended to the proximal leg in our patient, which is a rare presentation in this disease [7].

Although PEODDN is considered a benign congenital hamartoma, an association between PEODDN and a variety of disorders, such as Bowen's disease, squamous cell carcinoma, sensory polyneuropathy, deafness and hyperthyroidism, have been reported. Histopathologically, PEODDN is identified by cornoid lamella appeared over acrosyringia [8]. The treatment options are ablative methods, particularly laser therapy, and for localized lesions, excision by surgery is

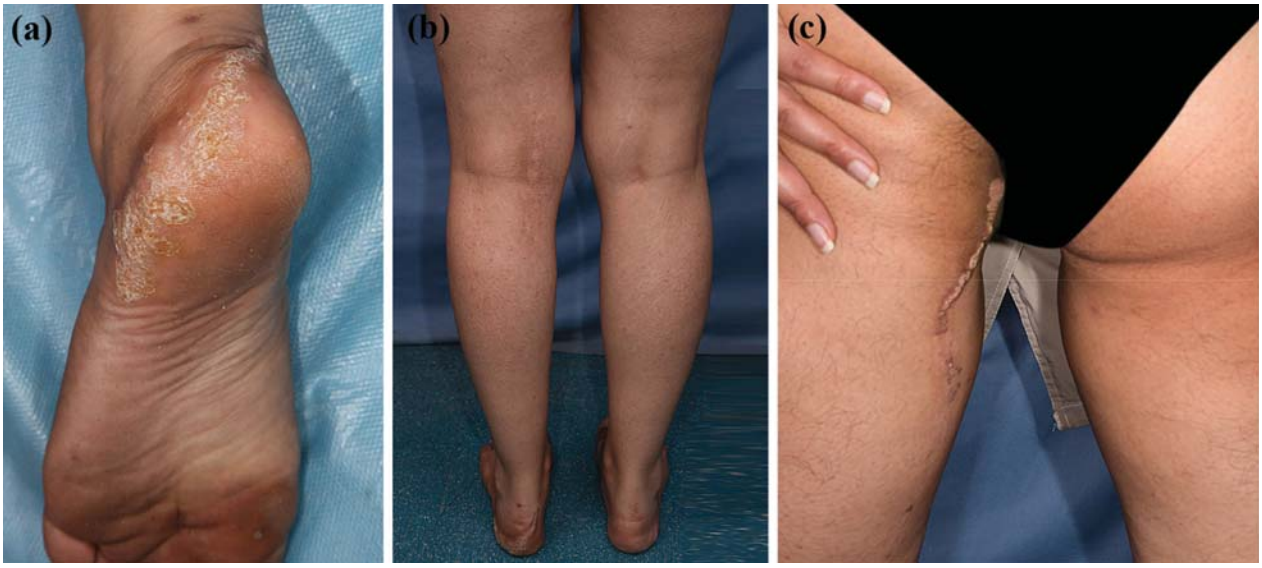

Figure 1. (a) Multiple keratotic papules on the sole; (b) Linear verrucous plaque following Blaschko's line with extensive distribution from the sole to the leg, (c) thigh and the left buttock.

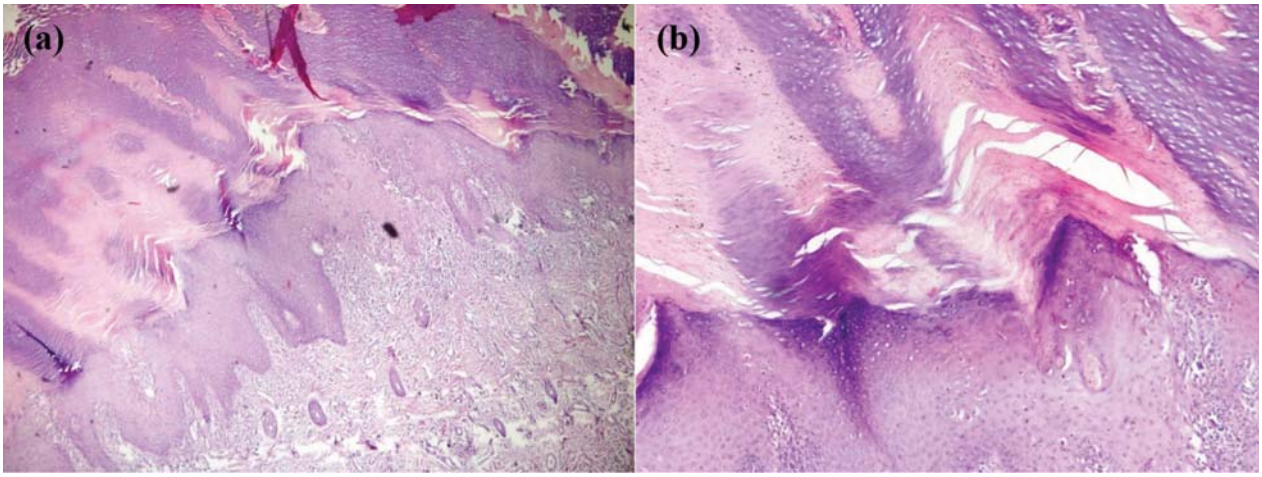

Figure2. (a) Hyperkeratosis and acanthosis with cornoid lamella overlying the acrosyringium (focal porokeratosis above eccrine duct) (H & E ×40) (b) Dyskeratotic cells in the upper squamous layer under the porokeratotic column (H & E ×100).

recommended. Laser therapy with ultrapulse CO2 laser and erbium/CO2 laser has been reported as a successful treatment [9].

CONCLUSION

In the present report, the rarity of PEODDN and interesting presentation of the lesions on the sole of our patient with extension in a linear Blaschko pattern to the leg, thigh and buttock resembling verrucous linear epidermal nevus, promoted us to present this case.

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Conflict of interest None declared.

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