A case of porokeratosis plantaris palmaris et disseminata and literature review

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Abstract

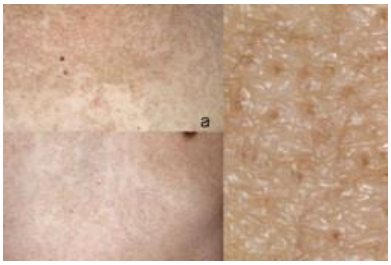
Porokeratosis plantaris palmaris et disseminata (PPPD) is a quite rare variant of porokeratosis. We report a 56-year-old male patient. He first noted brownish, asymptomatic, annular macules scattered on the trunk and extremities at about age 31 and these skin lesions, continued to increase in number. Ten years prior to presentation, similar lesions appeared on the palms. About one to two years ago, the patient noted painful wart-like, keratotic punctuate papules 2-3 mm in diameter on the soles, which disturbed walking. Histological examination showed the characteristic feature of the cornoid lamella. The family tree of this patient showed an autosomal dominant mode of transmission. We review sixteen typical cases of PPPD previously reported in the English literature.

Introduction

Porokeratosis plantaris palmaris et disseminata (PPPD) is a rare variant of porokeratosis, originally described by Guss et al [1] in 1971. They demonstrated that PPPD has its onset in the teens and early twenties, is autosomal dominant, affects males twice as often as females, and in some cases is exacerbated during the summer by sun exposure. PPPD initially occurs in the palmoplantar areas with subsequent involvement of the other areas of the body, including sites unexposed to ultraviolet radiation. Morphologically, its clinical features resemble disseminated superficial (actinic) porokeratosis (DSP/DSAP). We report herein a Japanese case of PPPD and review twelve typical cases of PPPD previously reported in the English literature.

Case report

A 56-year-old man first noted brown, asymptomatic, annular macules on the trunk and extremities at about age 31 and these skin lesions continued to increase in number. Ten years prior to presentation, similar lesions appeared on the palms. About one to two years ago, the patient noted painful, small wart-like lesions on the soles, which disturbed his walking. There was no sign of worsening during the summer. The patient had surgery for colon cancer 6 months prior to his consultation.



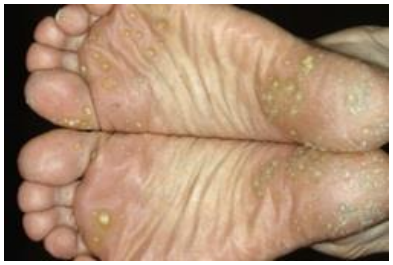


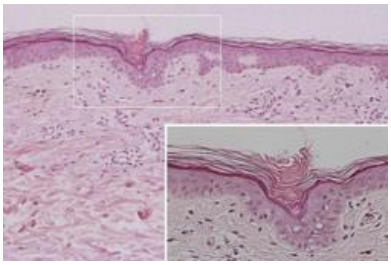
Figure 1

Figure 2

Figure 1. 1a and 1b: Hundreds of brownish annular papules and plaques of various sizes up to 1 cm in diameter with slightly raised keratotic margins were disseminated on the trunk and extremities. Some of lesions tended to coalesce in irregular, concentric patterns. 1c: There are a few pinhead sized brown papules in the center of annular plaques.

Figure 2. Wart-like keratotic punctuate papules are seen on the soles, especially on pressure points such as the heels and metatarsal heads.

On physical examination, hundreds of brownish annular plaques of varying size, up to 1cm in diameter with slightly raised keratotic margins, were disseminated on the trunk and extremities (Figure 1). Some of the lesions tended to coalesce in irregularly concentric patterns. There were a few brown, pinhead-sized papules in the center of the annular plaques. Similar annular lesions were seen on the palms, although these were smaller than the lesions on trunk and extremities.



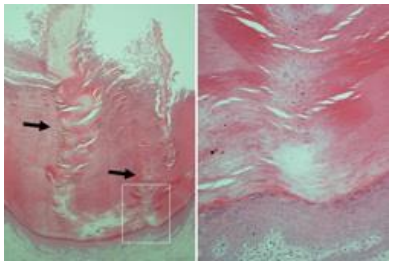


Figure 3

Figure 4

Figure 3. Histopathology of the lesion on the back. Tightly packed columns of keratotic cells in the cornified layer (cornoid lamella), lost granular layer, mild dyskeratosis, and vacuolar cells in the epidermis below the cornoid lamella are observed. Focal lymphocytic infiltrate is present in the superficial dermis.

Figure 4. Histopathology of the planter lesion indicating wedge-shaped hyperkeratosis. Two cornoid lamellae were found in the orthokeratotic material (indicated by arrows), beneath which the granular layer is thin or absent. A small quantity of vacuolar cells can be seen in the epidermis.

Furthermore, wart-like, keratotic, punctuate papules, 2-3 mm in diameter, occurred on the soles, especially the pressure points such as the heel and metatarsal head (Figure 2). There were no lesions on the face or oral mucosa. Results of laboratory examinations were within the normal range. Histopathological examination taken from the edge of an annular macule on his back (Figure 3) showed tightly packed columns of keratotic cells in the cornified layer (cornoid lamella), loss of the granular layer, mild dyskeratosis, and vacuolar cells in the epidermis below the cornoid lamella. Another specimen from the sole (Figure 4) also showed two cornoid lamellae in the orthokeratotic material (indicated by arrow), beneath which the granular layer was thin or absent. A small quantity of vacuolar cells were seen in the epidermis.

The patient's father, uncle, and paternal grandfather had similar lesions with a similar distribution.

Discussion

PPPD is an uncommon clinical type of porokeratosis. To our knowledge, only 16 typical cases were previously described in the English literature (**Table 1**) [1-16]. In general, PPPD initially appears on the palms and soles and subsequently involves other skin areas. Although the morphology and distribution of the lesions were typical of PPPD in our patient, the initial lesions appeared on the trunk and extremities and 15 years later on the palms and soles. This sequence was previously observed in only four families [**2**,**3**, **4**, **5**] besides our case. Some cases of PPPD are difficult to differentiate from DSP/DSAP when the symptoms manifest late as plantar and palmar lesions. PPPD lesions on the trunk and extremities closely resemble those of DSP/DSAP. This suggests that prior cases of PPPD may have been misdiagnosed as DSP/DSAP because of the late appearance of the lesions on the palms and soles. Although Guss et al [**1**] emphasized the early appearance of plantar and palmar lesions, we do not consider the initial site as being as important. Misdiagnosis because of the late appearance of the plantar and palmar lesions can occur. The age of onset seems to be in the teens to thirties, the higher end of the age range being slightly greater than Guss has described. Porokeratosis is a well-known disorder that complicates other phenotypes, so it is difficult to distinguish PPPD from punctate porokeratosis with DSP/DSAP clinically and histologically. But it would be a low probability that this could occur through three consecutive generations.

It has been suggested that the inheritance pattern of PPPD is consistent with an autosomal dominant mode of transmission. Ten of seventeen reported families (59%) showed autosomal dominant transmission. The family tree in our case shows three consecutive generations of male-to-male transmission. In X-linked transmission normal females are necessary between affected males. Interestingly it is significantly different from DSP/DSAP in which the sporadic pattern is predominant. Autosomal dominant transmission in the Japanese literature shows DSP, 7 of 20 (35%) and DSAP, 11 of 55 (20%) [**17**]. Although some reports classify PPPD as another clinical variant of porokeratosis [**18**], we believe that PPPD is an independent clinical entity. PPPD not only has characteristic clinical features, but it has palmoplantar lesions and

an earlier age of onset than in DSAP, which typically begins in the third or fourth decade [6]. More importantly, the characteristic inheritance pattern confirms the diagnosis. Examination of 17 reported cases of PPPD showed that the overwhelming majority of patients (12 persons) were male, with only 4 affected females. Male-to-male transmission was reported in 3 families besides our case (Table 1). This fact is also corroborated by the observation of Guss et al [1]. This gender pattern is similar to the other types of porokeratosis. The abnormal gene is inherited by 50 percent of the offspring; both males and females equally inherit the defective gene, but not all will develop the disease. Despite equal genetic transmission of the disease, the clinical phenotype is more prevalent in males than females. The basis for this intriguing sex-related distinction is unknown.

Porokeratosis is reported to be a premalignant condition of the skin. The premalignant potential is well illustrated in all varieties of porokeratosis by the numerous reports of squamous cell carcinoma and Bowen disease developing in preexisting porokeratotic lesions [9]. Five cases of squamous cell carcinoma were reported as co-occurring with PPPD (Table 1), as is often the case with other types of porokeratosis. Continued careful observation of this disease is therefore required.

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