

# Eruptive pruritic papular porokeratosis

Wan-Lu Zhang, Dan Huang, Wei Zhang, Ying-Da Wu, Su-Ying Feng, Yi-Qun Jiang, Cheng-Rang Li

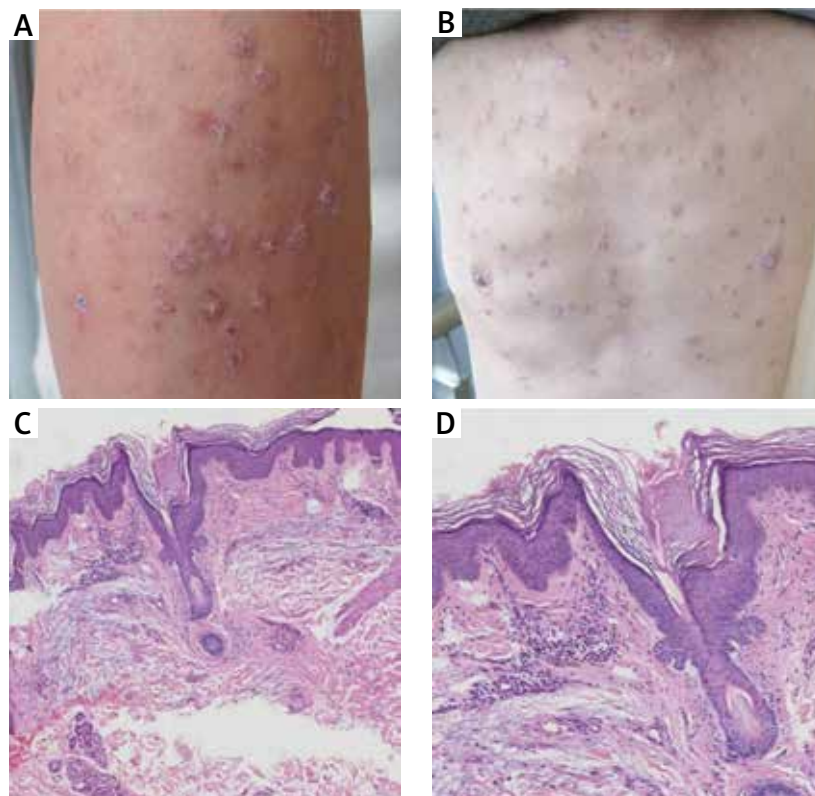
Department of Dermatology, Chinese Academy of Medical Sciences and Peking Union Medical College, Nanjing, China

Adv Dermatol Allergol 2021; XXXVIII (1): 167–169

DOI: <https://doi.org/10.5114/ada.2021.104293>

“Eruptive pruritic papular porokeratosis (EPPP)”, first proposed by Kanzaki in 1992 [1] and designated as “Inflammatory disseminated superficial porokeratosis (Inflammatory DSP)” by Tanaka in 1995 [2], is a special variant of porokeratosis. The typical clinical course consists of asymptomatic DSP for months to years, followed by the acute pruritic exacerbations, and then subsides spontaneously within 12 months [3]. Histopathological examination reveals the characteristic cornoid lamellae. This paper describes a typical case of EPPP in China and summarizes previously reported EPPP cases.

Herein we describe a typical case in China – a 68-year-old man with EPPP for 5 years. After the initial 3 years of asymptomatic DSP, typical eruptions such as annular lesions with a raised and sharply margined keratotic border suddenly presented with recurrent multiple, pruritic, erythematous papules on his extremities and trunk. The lesions subsided in the winter and recurred in the summer. Physical examination revealed numerous nodular erythematous papules like prurigo nodularis lesions (Figure 1 A), and hyperpigmented brown macules on his trunk (Figure 1 B). The patient had a history of mild hypertension, no oth-



**Figure 1.** Clinical and histopathological features of the case

**Address for correspondence:** Cheng-Rang Li, Department of Dermatology, Chinese Academy of Medical Sciences and Peking Union Medical College, 12 Jiangwangmiao St, Nanjing, China, e-mail: [nylcr72@163.com](mailto:nylcr72@163.com)

**Received:** 10.10.2018, **accepted:** 3.03.2020.

This is an Open Access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 International (CC BY-NC-SA 4.0). License (<http://creativecommons.org/licenses/by-nc-sa/4.0/>)

**Table 1.** Summary of previously reported cases of EPPP

Case no.	Age of onset	Gender	Pre-existing asymptomatic porokeratosis	Period of pre-existing lesions	Period of significant regression	Other previous diseases/suspicious triggering factors	Tissue eosinophilia	Author
1	71	F	+	Several years	5 months	–	+	Kanzaki <i>et al.</i> [1]
2	60	M	+	Nearly 10 years	NM	CVA/drugs	+	Kanzaki <i>et al.</i> [1]
3	75	M	+	Over 10 years	6 months	Systemic steroids	–	Kanzaki <i>et al.</i> [1]
4	78	M	+	Several years	3 months	–	–	Tanaka <i>et al.</i> [2]
5	69	M	–	–	1 year	–	+	Stork <i>et al.</i> [4]
6	77	M	+	20 years	4 months	–	+	Ricci <i>et al.</i> [5]
7	50	M	+	10 years	NM	–	–	Kang <i>et al.</i> [6]
8	74	M	+	3 years	2 months	Cerebral and myocardial infarction, diabetes, gout, and prostatic hypertrophy	+	Makino <i>et al.</i> [7]
9	82	M	+	1 years	6 months	–	–	Kanekura <i>et al.</i> [8]
10	71	F	+	35 years	NM	–	+	Klein <i>et al.</i> [9]
11	84	M	+	3 years	1 month	Colon cancer	+	Choi <i>et al.</i> [10]
12	68	M	+	4 months	2 months	–	+	Wakatabi <i>et al.</i> [11]
13	63	M	+	Over 13 years	NM	–	–	Tee <i>et al.</i> [3]
14	77	F	+	Over 20 years	NM	–	+	Yamamoto <i>et al.</i> [12]
15	80	M	+	2 years	NM	Hypertension and hyperuricemia	–	Inazawa <i>et al.</i> [13]
16	63	F	+	3 years	2 months+	Basedow's disease and oral thiamazole	–	Patrizi <i>et al.</i> [14]
17	68	M	+	3 years	Up to now	Hypertension	–	Present case

F – female, M – male, + – positive, – – negative, NM – not mentioned, CVA – cerebrovascular accident.

er relevant diseases and he was not on medication. Biopsy specimen of a reddish papule on his right upper limb did not show the granular layer and patchy parakeratosis of keratinocytes in the characteristic cornoid lamellae (Figure 1 C). Mild focal lymphocytes (without eosinophils) infiltrates around the blood vessels in upper dermis (Figure 1 D).

Based on the above symptoms, the patient was definitely diagnosed as EPPP. These pruritic papules did not respond well to topical steroids or oral antihistamines. Interestingly, as the winter progressed, the papules levelled off and the itching was reduced, leaving behind some brownish non-inflamed patches in the follow-up.

By analysing 17 cases of EPPP including the one reported here, we found that EPPP might be manifested with the following characteristics (Table 1 [1–14]): (1) history of pre-existing porokeratosis varied from months (> 2 months) to several years except 1 patient who had no pre-existing lesions and no tendency to spontaneous resolution in a 16-month follow-up [4]; (2) less than half of patients had other pre-existing diseases or suspicious triggering factors, including cardiovascular disease, medication history, autoimmune disease, cancer or others;

(3) elderly males might be the susceptible population. The mean age of 17 patients was 71 years, including 13 males in contrast to 30–40 years at onset and the 1 : 3 ratio of male : female for the classic porokeratosis [11]; (4) the typical clinical case presents with sudden multiple erythematous papules, accompanied by strong itching, followed by asymptomatic porokeratosis skin lesions; (5) with the cornoid lamellae, histological examination revealed the presence of eosinophils infiltration in 9 patients, more than half of the cases; and (6) the eruptions could subside within 12 months spontaneously in the follow-up [3].

The reasons responsible for EPPP sudden appearance are still not clear. And the interventions for this particular type of porokeratosis are identical to the classic DSP. Some therapeutic options were used for acute phase lesions such as topical steroids, 5-fluorouracil, assisted freezing, and laser etc. [4, 5], but the efficacy is unsatisfied. Oral antihistamines can be effective against pruritus.

In conclusion, we reported a Chinese man with typical EPPP characteristics. In low temperatures, the eruption tends to resolve, which might be related to season, sun-

light, humidity or sweat stimulation. Whereas, the mechanisms and characteristics of EPPP need to be further studied with more similar cases.

### Acknowledgments

We thank Qi Li for helping search the literature. This study was funded by CAMS Innovation Fund for Medical Sciences (grant numbers 2016-I2M-1-002), Six Talent Peaks Project in Jiangsu Province (WSN-126) and Technology Foundation for Selected Overseas Chinese Scholar, Ministry of Personnel of China.

### Conflict of interest

The authors declare no conflict of interest.

### References

1. Kanzaki T, Miwa N, Kobayashi T, Ogawa S. Eruptive pruritic papular porokeratosis. *J Dermatol* 1992; 19: 109-12.
2. Tanaka M, Terui T, Kudo K, Tagami H. Inflammatory disseminated superficial porokeratosis followed by regression. *Br J Dermatol* 1995; 132: 153-5.
3. Tee SI, Chong WS. Eruptive pruritic papular porokeratosis. *Indian J Dermatol Venereol Leprol* 2012; 78: 758-60.
4. Stork J, Kodetova D. Disseminated superficial porokeratosis: an eruptive pruritic papular variant. *Dermatology* 1997; 195: 304-5.
5. Ricci C, Rosset A, Panizzon RG. Bullous and pruritic variant of disseminated superficial actinic porokeratosis: successful treatment with grenz rays. *Dermatology* 1999; 199: 328-31.
6. Kang BD, Kye YC, Kim SN. Disseminated superficial actinic porokeratosis with both typical and prurigo nodularis-like lesions. *J Dermatol* 2001; 28: 81-5.
7. Makino E, Inaoki M, Fujimoto W. Inflammatory stage of disseminated superficial porokeratosis. *J Dermatol* 2005; 32: 890-3.
8. Kanekura T, Yoshii N. Eruptive pruritic papular porokeratosis: a pruritic variant of porokeratosis. *J Dermatol* 2006; 33: 813-6.
9. Klein N, Enk A, Hartschuh W. Inflammatory stage of disseminated superficial porokeratosis in a 71-year old patient. *Hautarzt* 2009; 60: 420-3.
10. Choi KH, Kim TY. A case of inflammatory disseminated superficial porokeratosis in a colon cancer patient. *Ann Dermatol* 2009; 21: 150-3.
11. Wakatabi K, Kakurai M, Yamada T, et al. Inflammatory disseminated superficial porokeratosis with an unusual clinical feature of the pruritic, erythematous papules preceding annular brownish pigmentation. *J Dermatol* 2012; 39: 946-8.
12. Yamamoto T, Furukawa H, Ohtsuka M. Amyloid deposition in disseminated superficial porokeratosis with inflammatory stages. *J Dermatol* 2013; 40: 1059-60.
13. Inazawa M, Satoh T, Yokozeki H. Hyperkeratotic variant of inflammatory disseminated superficial porokeratosis with lichenoid reaction and extensive amyloid deposition. *Int J Dermatol* 2014; 53: e94-5.
14. Patrizi A, Viridi A, Misciali C, Bardazzi F. Eruptive pruritic papular porokeratosis in a Caucasian woman: a transient inflammatory stage of porokeratosis. *G Ital Dermatol Venereol* 2017; 152: 529-31.