Eruptive pruritic papular porokeratosis or inflammatory form of disseminated superficial porokeratosis: a new case and review of the literature

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Abstract

Porokeratosis is a heterogeneous group of dermatoses with alterations of keratinization. Histologically, they are characterized by the presence of cornoid lamellae. Eruptive pruritic papular porokeratosis (EPPP) or the inflammatory form of disseminated superficial porokeratosis (or eruptive disseminated porokeratosis) is an infrequent variant, characterized by pruritic erythematous papules or annular lesions. We present a 72-year-old woman with EPPP, exhibited by pruritic lesions on the extremities and back, and review the literature concerning this condition. We found 32 cases of EPPP or inflammatory disseminated superficial porokeratosis (including the current case) reported in the literature, with a median age of 66 years (range, 13-84); 59.3% were men. Eruptive pruritic papular porokeratosis was associated with various neoplasms in 31.2% of cases. Six patients had an associated viral infection. Response to treatment was poor in most cases. Eruptive pruritic papular porokeratosis resolved spontaneously in 75% of cases. Median time to resolution was 6 months (range, 1-24). Eruptive pruritic papular porokeratosis (or inflammatory disseminated superficial porokeratosis/eruptive disseminated porokeratosis) is an infrequent variant of porokeratosis characterized by intense pruritus and spontaneous resolution in most individuals. Eruptive pruritic papular porokeratosis can be associated with neoplasms and screening for malignancies is recommended if clinically indicated.

Keywords: porokeratosis, eruptive pruritic papular porokeratosis, disseminated superficial porokeratosis, eruptive disseminated porokeratosis, inflammatory, cornoid lamella, malignancies

Introduction

Porokeratosis is a heterogeneous group of dermatoses with alterations of keratinization. Histologically, they are characterized by the presence of cornoid lamella. Eruptive pruritic papular porokeratosis (EPPP) or the inflammatory disseminated superficial porokeratosis, is an infrequent variant, characterized by pruritic erythematous papules or annular lesions [1–3]. We present a case of EPPP and review the literature.

Case Synopsis

A 72-year-old woman had been diagnosed four years prior to being seen with disseminated superficial actinic porokeratosis on the legs. She was suffering from an eruption of pruritic skin lesions on the extremities and trunk with four months of evolution. Physical examination showed multiple macules and annular erythematous papules on the thighs, legs (**Figure 1**), arms, and back. Dermoscopy revealed an erythematous ring with hyperkeratotic areas. Histopathology showed the presence of cornoid lamella, vacuolation of the basal membrane, and a perivascular lymphoid inflammatory infiltrate (**Figure 2**). A diagnosis of EPPP was made. The patient received topical and oral corticosteroids (prednisone 10mg/day) and antihistamines without clinical improvement. Narrow band UV-B phototherapy was started with mild improvement and reduction of pruritus. Logistical complications forced the suspension of phototherapy. Acitretin (25mg/day) was ineffective. Diclofenac sodium 3%, 5-fluorouracil 5%, and imiquimod 5% led to adverse

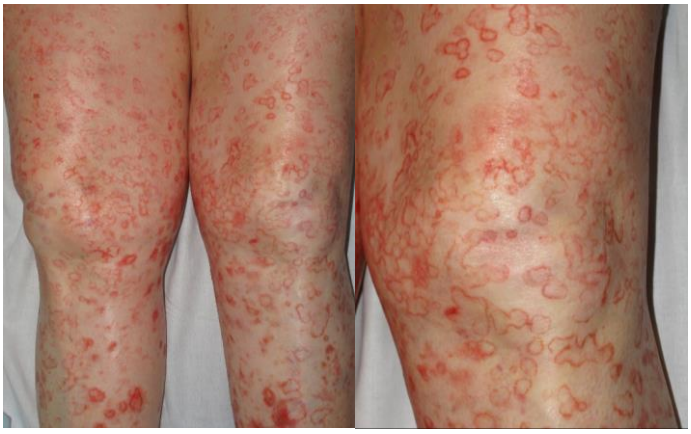

Figure 1. *Eruptive pruritic papular porokeratosis. Multiple macules and annular erythematous papules on the thighs, legs, and on the left knee.*

skin effects without clinical improvement. After three years, the cutaneous lesions of the arms and trunk spontaneously remitted and those of the lower extremities no longer presented an inflammatory aspect. The patient has not developed neoplasms or systemic diseases after four years of follow-up.

Case Discussion

Eruptive pruritic papular porokeratosis was first described by Kansaki et al. in 1992 [1]. They described three patients with DSP who presented with pruritic erythematous papules. Histology showed the presence of cornoid lamella and eosinophils in the dermis. An inflammatory infiltrate rich in CD8+ T lymphocytes has also been described, unlike disseminated superficial porokeratosis in which CD4+ lymphocytes are common [2].

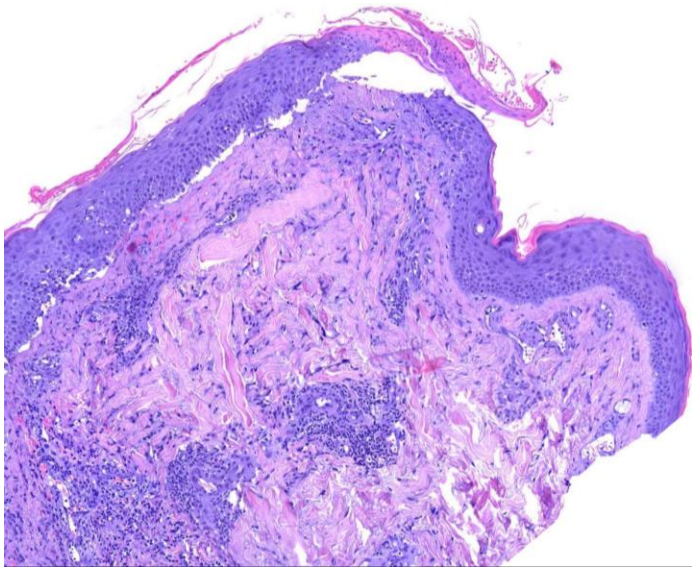

Figure 2. *Histopathology. Atrophic epidermis with the presence of cornoid lamella, vacuolation of the basal membrane, and a perivascular lymphoid inflammatory infiltrate. H&E, 100×.*

Eruptive pruritic papular porokeratosis is exceedingly rare. We found only 32 cases of EPPP or the inflammatory form of disseminated superficial porokeratosis (including the current case) reported in the literature (**Table 1**), with a median age of 66 years (range, 13-84); 59.3% were men. Nearly 52% (14/27) had previously presented with diverse subtypes of porokeratosis, the most frequent being disseminated superficial porokeratosis (8/14). All the individuals experienced intense pruritus. On histology, the presence of a cornoid lamella and a dermal inflammatory infiltrate were the most frequent findings (9 cases of lymphohistiocytic infiltrate and 8 with eosinophils). Eruptive pruritic papular porokeratosis was associated with various neoplasms in 31.2% (10/32) of cases (three hepatocellular carcinomas, one acute myeloid leukemia, one acute lymphoblastic leukemia, one cholangiocarcinoma, one pancreatic carcinoma, one colorectal carcinoma, and one mammary carcinoma). There was one case of myelodysplastic syndrome (1/32). Six patients had an associated viral infection: hepatitis C virus (4/6), hepatitis B (1/6) and recurrent herpes simplex virus (1/6). Response to treatment was poor in most cases. Topical corticosteroids and antihistamines were the most frequently used. Eruptive pruritic papular porokeratosis resolved spontaneously in 75% of cases. Median time to resolution was 6 months (range, 1-24).

Eruptive pruritic papular porokeratosis could be an immunological response directed against clones of abnormal keratinocytes. The etiology of this immune response remains elusive, although in some cases it could be secondary to neoplasms or viruses [4, 5]. The association with neoplasms found in our review (especially hepatopancreatic and hematological) is noteworthy; screening for these should be considered if clinically indicated. Additional reports would be helpful to establish whether the

observation of cancer in patients with EPPP is bona fide or coincidental.

Conclusion

Eruptive pruritic papular porokeratosis (or inflammatory form of disseminated superficial porokeratosis) is an infrequent variant of porokeratosis characterized by intense pruritus and spontaneous resolution in most individuals. Eruptive pruritic papular porokeratosis can be associated with neoplasms and viral infections.

Potential conflicts of interest

The authors declare no conflicts of interests.

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Table 1. *Cases with eruptive pruritic papular porokeratosis (or the inflammatory form of disseminated superficial porokeratosis) described in the literature*

	Sex/age (years)	Comorbidities	Previous dermatoses	Histopathology	Spontaneous resolution	Treatment	Response to treatment
Kanzaki, 1992 [1]	F/71 M/60 M/75	No Stroke Treatment with oral CS	DSP DSP DSP	Cornoid lamella. Lymphocytes and eosinophils in dermis Cornoid lamella. Lymphocytes and eosinophils in dermis Cornoid lamella. No eosinophils	Yes (5 months) Yes Yes (6 months)	Topical CS, AHs ND Topical and intralesional CS, AHs	No response ND No response
Tanaka, 1995 [2]	M/78	No	DSP	Cornoid lamella. Colloid bodies, mononuclear cells in dermis	Sí (15 meses))	Topical 5-FU	No response
Stork, 1997 [6]	M/69	Eosinophilia and increased IgE	No	Cornoid lamella. Intraepidermal vesicles with eosinophils. Subepidermal detachment. Mixed perivascular infiltrate	No (16 months of follow-up)	Oral CS, UVA y heliotherapy. Etretinate	No response Good response
Ricci, 1999 [7]	M/77	No	DSAP	Cornoid lamella. Lymphocytes and eosinophils in dermis	Yes (2 years)	Etretinate, CS, AHs, AZT. CS & topical 5-FU. Grenz rays	moderate response with Grenz rays
Levin, 1999 [8]	M/70	Myelodysplastic syndrome, hemolytic anemia (oral CS)	ND	Cornoid lamella	ND	No treatment	
Knoell, 1999 [9]	M/56	Renal transplant, HT	No	Cornoid lamella, papillomatosis, acanthosis	yes (within weeeks)	Cryotherapy	Good response
Kono, 2000 [5]	M/67 M/62 F/58	HCV, HCC (2 months after EPPP) HCV, HCC (6 months after EPPP) HCV, HCC (after EPPP)	No No No	Cornoid lamella Diagnosis of DSP Diagnosis of DSP	ND ND ND	Percutaneous ethanol injection (HCC) Percutaneous ethanol injection (HCC) No treatment	Good response Good response
Kang, 2001 [10]	M/50	No	DSP	Cornoid lamella, hypogranulosis, Lymphohistiocytic infiltrate in dermis	ND (3 months of follow-up)	Topical 5-FU	No response
Pearson, 2003 [11]	F/30	Pregnancy	Linear porokeratosis	Cornoid lamella	ND	ND	ND
Jang, 2004 [12]	F/57 F/53	Recurrent HSV Dysphagia	ND ND	Cornoid lamella, necrotic keratinocytes, dermal edema Compatible with porokeratosis	ND ND	ND ND	ND ND

Makino, 2005 [3]	M/74	Diabetes, gout, prostatic hyperplasia	DSAP	Cornoid lamella. Lymphocytes and eosinophils in dermis	Yes (13 months)	Topical CS & oral AHs	No response
Kanekura, 2006 [13]	M/82	No	DSP	Cornoid lamella. Interphase dermatitis. Lymphohistiocytic infiltrate in dermis	Yes (8 months)	Topical CS	No response
Lee, 2006 [14]	F/73	Cholangiocarcinoma	No	Cornoid lamella, dyskeratosis	No	No treatment	
Klein, 2009 [15]	M/71	Hepatitis B	Porokeratosis of Mibelli	Cornoid lamella. Eosinophils in dermis	ND	Topical CS, tacrolimus & retinoids. Oral CS & AHs	Intermittent response to oral CS
Choi, 2009 [16]	M/84	Colorectal carcinoma (6 years of evolution). Chemotherapy	DSP	Cornoid lamella. Lymphocytes and eosinophils in dermis	Yes (4 months)	Topical CS & oral AHs	Good response after 1 month
Goulding, 2009 [17]	M/63	Hip replacement, Pulmonary embolism (after surgery)	No	Cornoid lamella. Lymphohistiocytic infiltrate in dermis	Yes (5 weeks)	Topical CS & urea	Symptomatic relief
Schena, 2010 [17]	F/77	Hepatitis C, pancreatic carcinoma	No	Cornoid lamella, dyskeratosis. Lymphohistiocytic infiltrate in dermis	No (died in the following 2 weeks)	ND	ND
Duhm, 2011 [18]	F/68	No	DSAP	Cornoid lamella. Lymphocytic infiltrate in dermis	Yes (1 year)	CS, acitretin	Symptomatic relief
Pini, 2011 [19]	M/13	Acute lymphoblastic leukemia, allogenic transplant (one year previously)	ND	Cornoid lamella, dyskeratosis. Lymphohistiocytic infiltrate in dermis	Yes (after suspending immunosuppressants)	Topical urea	No response
Tee, 2012 [20]	M/63	No	DSP	Cornoid lamella, dyskeratosis. Lymphohistiocytic infiltrate in dermis	No	Topical CS	Good response
Shoimer, 2014 [21]	F/54	ND	ND	Compatible with porokeratosis	ND	Topical Calcipotriol, cryotherapy, oral CS	No response
Bednarek, 2015 [22]	F/39	Asthma, history of histoplasmosis, oral CS therapy	No	Cornoid lamella. Lymphocytic infiltrate in dermis	Yes (2 months)	emollients	ND

Marks, 2016 [23]	M/56	Acute myeloid leukemia (Imatinib therapy)	Angiokeratomas. GVHD	Cornoid lamella	ND	ND	ND
Yalcin, 2016 [24]	M/65	Diabetes mellitus type 2	No	Cornoid lamella. Eosinophils in dermis	ND	ND	ND
Mangas, 2017 [25]	F/62	Breast cancer (trastuzumab & exemestane therapy)	ND	Cornoid lamella, hypogranulosis	Yes	Topical CS & salicylic acid	Symptomatic relief
Rigo, 2018 [26]	F/76	COPD, pneumonia, CS therapy	No	Cornoid lamella. Subepidermal blister	ND	ND	ND
Current report	F/72	Hypothyroidism, dyslipidemia	PASD	Cornoid lamella. Interface dermatitis. Lymphocytes in dermis	Yes (2 years)	Oral CS, acitretin & AHs. Topical 5-FU, diclofenac & imiquimod. Narrow band UVB	Mild improvement with narrow band UV-B

Abbreviations: DSP, disseminated superficial porokeratosis; DSAP, disseminated superficial actinic porokeratosis*; EPPP,* eruptive pruritic papular porokeratosis*; CS, corticosteroids; AHs, antihistamines; HSV, herpes simplex virus; HCC, hepatocellular carcinoma; COPD, chronic obstructive pulmonary disease; ND, not described; GVHD, graft versus host disease; HT, hypertension; 5-FU, 5-fluoracil; AZT, azathioprine.*