

Localised nasal actinic porokeratosis: An independent subtype of porokeratosis?

Nidhi Sharma, Yukti Aggarwal, Suresh Kumar Malhotra, Kuldeep Singh Chahal

Departments of Dermatology and Pathology, Government Medical College, Amritsar, Punjab, India

Corresponding author: Dr. Nidhi Sharma, E-mail: drnidhisharma@hotmail.com

ABSTRACT

Porokeratosis (PK) are a group of keratinizing disorders clinically characterized by atrophic plaques with peripheral hyperkeratotic ridge which gradually spread in a centrifugal manner. The sites of predilection are trunk or extremities and facial lesions are rarely reported. We report a sporadic adult-onset case of porokeratosis of a 20 year old female with a solitary, asymptomatic, persistent classical plaque confined to the ala of nose. On reviewing the literature, nasal and perinasal area has been found to be exclusively involved in many cases of facial porokeratosis. Such cases have unique yet common epidemiological and clinical profile thereby deserving a separate category while being classified.

Key words: Porokeratosis; Nasal alae; Localized; Solar; Non-familial

INTRODUCTION

Porokeratosis (PK) is an epidermal keratinizing disorder, characterized clinically by distinct, indolently evolving, annular plaques with atrophic center and irregular, peripheral, elevated hyperkeratotic ridge as well as histologically by the cornoid lamella [1].

The etiopathogenesis of PK is complex and has been broadly attributed to the peripheral expansion of abnormal and mutant clone of parakeratotic keratinocytes [2]. The attributing risk factors for clonal proliferation may be intrinsic i.e. genetically determined or extrinsic factors like immunosuppression, immunodeficiency (AIDS), ultraviolet radiation, infections, mechanical trauma, drugs (thiazide diuretics), hematological malignancies, autoimmune diseases, and occupational exposure to benzene etc [3].

Clinically, six distinct types of PK are well recognized i.e. PK of Mibelli (PM); disseminated superficial actinic PK (DSAP); disseminated superficial PK (DSP); PK palmaris et plantaris disseminate (PPPD); punctate PK and linear PK [4]. The usual pattern of inheritance is autosomal dominant but at times it may be sporadic. A few of these subtypes exhibit propensity for

malignant transformation as conversion into squamous cell carcinoma, Bowen disease and basal cell carcinoma has been described in 6.8 -11.0% cases of PK [5].

Common sites of predilection are trunk and extremities although sporadic cases of involvement of palms and soles, face, oral mucosa, lips have been reported [5]. The present study highlights the high incidence of nasal PK among the reported cases in the world literature which is clinically and epidemiologically distinct from other types of PK.

CASE REPORT

A twenty year old female patient presented with persistent, asymptomatic plaque over nose since two years. It started as a small papule which slowly progressed in size in a centrifugal manner. Photosensitivity was present. Past medical and family history was non-significant. On clinical examination, there was a single plaque of size 1*1.5 cm confined to the right alae of nose (Fig. 1). There was atrophy in the center of the lesion and margins were hyperkeratotic and peripherally raised to a height of 3 mm with perimarginal fissure. Otherwise general physical and systemic examination

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was unremarkable. Routine laboratory investigations were within normal limits. Viral markers and ELISA for HIV-1, 2 were non-reactive. The differential diagnosis of discoid lupus erythematosus, PK, basal cell carcinoma, lupus vulgaris and sarcoidosis were considered. An incisional skin biopsy revealed parakeratotic invaginations in the epidermis (Fig. 2) consistent with the diagnosis of PK.

DISCUSSION

Although, PK was first described by Mibelli in 1893, a case of facial PK was first reported in 1979 by Mehregan. He reported that 197 cases out of 165,000 consecutive skin biopsy specimens verified histologically for PK, of which only seven patients had primary facial lesions (3.5%) [6]. On reviewing the literature, 49 cases of Facial/Nasal PK, with variable categorization have been identified. Their demographic and clinical features are shown in Table 1.

As evident from the table, there was exclusive involvement of nose and the perinasal area in 26 cases



Figure 1: A single plaque of size 1*1.5 cm confined to the right alae of nose with central atrophy and peripherally raised hyperkeratotic margins.

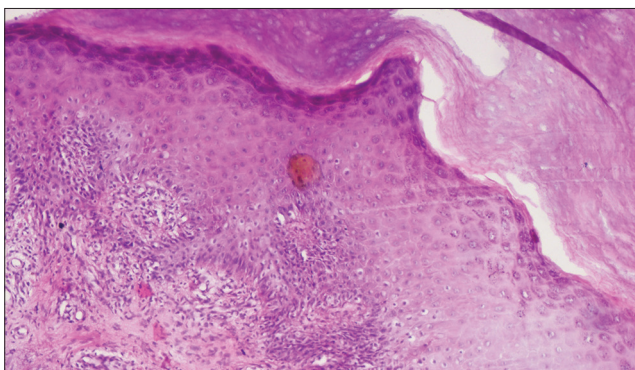


Figure 2: Parakeratotic invaginations in the epidermis. (H&E 1000x).

Table 1: Published case reports of porokeratosis on face and nose

S. No	Author	No of patients	Median age(yrs)	Sex	Duration of disease (yrs)	Distribution on face	Nasal involvement	No. of lesions (single/Multiple)	Size of lesions (cm)	Family history	Family history	Family history	H/o photo-sensitivity	Histological findings
1	Nabai et al (1979)[4]	1	18	M	2	Upper lip, chin, right cheek, right hand	0N	1M	----	NR	ND	ND	NR	PK, CL, Hg, DII
2	Mehregan et al (1980) [4]	7	37.3	2F/5M	3.4	Lips, lower eyelid, cheek, neck, ears	1N*	4S+3M	----	NR	ND	ND	NR	PK, CL,
3	Enk et al (1991) [4]	1	51	M	0.3	Right side of face	0N	1M	0.2-1.0	NR	ND	ND	NR	PK, CL, Hg, DII
4	Rahbari et al (1995) [6]	2	26	2M	?	Right side of face and nose	1N**	2M	----	NR	2D	2D	NR	PK, CL, Hg, DII
5	Navarro et al (2000)	1	18	M	3	Left side of face	0N	1M	Pinhead to 1.0	No	ND	ND	Absent	PK, DII
6	Chowdhury et al (2000) [2]	1	15	M	5	Bridge of nose	1N	1S	0.5	NR	ND	ND	Present	PK, CL, Hg, DII
7	Sharquie et al (2003) [1]	15	26.9	12F/3M	4	Nose & adjacent area	15N (7alae)	4S+11M	0.5-1	No	ND	ND	Present	PK, CL, Hg, DII

(Contd...)

Table 1: (Continued)

S. no	Author	No of patients	Median age(yrs)	Sex	Duration of Disease (yrs)	Distribution on face	Nasal involvement	No. of lesions (Single/Multiple)	Size of lesions (cm)	Family history	Destructive/ Nondestructive	H/o photo-sensitivity	Histological findings
8	Baccouche et al (2004) [7]	1	25	F	2	Left side of nose	1N (alae)	1M	NR	No	ND	NR	PK, CL, Hg, DII
9	Miranda et al (2004)	1	29	M	12	Upper lip, nose	1N	1S	3	No	D	NR	PK, CL, Hg, DII
10	Zhang et al (2005)	1	27	M	27	Right cheek	0N	1M	6*3.5	No	ND	NR	PK, Hg, DII
11	Feranbas et al (2006)	1	42	F	20	Left side of nose ext. to cheek & eye	1N	1S	7*4	No	D	Present	PK, CL, Hg, DII
12	Carranza et al (2008) [8]	1	34	M	10	Nose	1N	1M	-----	No	ND	Present	PK, CL, Hg, DII
13	Chaudhary et al (2009)	1	21	M	2	Nose and upper part of the lip	1N (alae)	1S	10	No	ND	NR	CL(f), Hg, DII
14	Gutierrez et al (2010) [5]	6	25.8	5F/1M	1.5	Left & Right side of face & nose	2N** (2alae)	3S+3M	-----	No	ND	NR	PK, CL, Hg, DII
15	Ghorpade et al (2010) [9]	1	22	M	0.6	Left & Right side of nose	1N (alae)	1M	0.2-0.3	No	ND	NR	PK, CL, Hg, DII
16	Wang et al (2010)	1	25	F	1	Supratip of nose	1N	1S	----	No	ND	Present	CL(f), Hg, DII
17	Rocha-Sousa et al (2011)	1	19	M	4	Distal part of Nose	1N (alae)	1M	0.2-1.0	No	ND	Absent	CL(f), Hg, DII
18	Robati et al (2011)	1	30	M	4	Right side of upper lip to lower lip & dorsum of both hands	0N	1M	4*5	No	ND	NR	PK, CL, Hg,
19	Lapresta et al (2011)	1	37	F	10	Upper part of the lip & Right nasal alei	1N (alae)	1M	1.3	No	ND	NR	PK, CL, Hg, DII
20	Ragunath et al (2012)	1	22	M	0.6	Left side of nose	1N (alae)	1S	-----	No	ND	Absent	PK, CL, DII
21	Ataseven et al (2012) [10]	1	23	M	3.5	Nasal alae	1N (alae)	1M	0.8-2	No	ND	Present	PK, CL, Hg, DII
22	Raid et al (2013) [4]	1	19	F	7	Bilateral malar area & nasal alei	1N (alae)	1M	0.3-2	No	ND	Absent	PK, CL, Hg,
23	Nur et al (2014)	1	34	F	3	Right side of nose	1N (alae)	1S	1	No	ND	Absent	CL(f), Hg, DII
	*	1	14	M	3	Cheek & nose	1N	1M		No	ND	No	PK, CL
	**	1	32	M	?	Right Cheek & nose	1N	1M		No	ND	No	PK, CL, Hg, DII
	***	1	38	F	1	Right side of the nose	1N (alae)	1S		No	ND	NR	PK, CL, Hg, DII
	****	1	22	M	1.5	Right and left side of the nose	1N (alae)	1M		No	ND	NR	PK, CL, Hg, DII

NR-Not Reported, ND-Non Destructive, PK-Parakeratosis, CL-Cornoid Lamella, CL(f)-Cornoid Lamella(follicular), Hg- Hypogranulosis, DII-Dermal Inflammatory Infiltrate

out of 49 and yet another 7 cases of nasal PK had additional involvement of cheek, lips and eye. Among these 26 cases, there were 16 females and 10 males. Their ages ranged from 15-38 years with mean age being 25.49 ± 6.73 SD.

The duration of the disease ranged from 6 months to 10 years with a mean \pm SD of 3.02 ± 2.66 years. The size ranged from 0.2-2 cm in the reported cases. The lesions were solitary in 9 cases and multiple in 17 cases.

19 out of 26 cases of nasal PK had a definite history of photosensitivity which may have acted as an aggravating factor. But in 4 cases, photosensitivity was not reported while in 3 cases it was found to be altogether absent.

The family history was noticeably negative in all these patients. None of these patients reported any kind of destructive change supervening over primary lesions of PK. The histopathological hallmark of PK, the cornoid lamella was observed in all the cases. The granular layer was absent under the cornoid lamella and perivascular lymphoid infiltrate in the dermis was noted in most of the cases.

Clinically, our case did not fit completely into any of the six clinical variants of PK. Interestingly, this case had gross similarity to the single largest published series (12 female and 3 male patients) [1] and the subsequent individual case reports of nasal PK [5,7-9].

In most of these publications, similar lesions were not reported in preceding or succeeding generations, so genetics probably played no role in the etiopathogenesis of this entity. In almost all the case reports, history of photosensitivity was present thereby implicating sunlight or artificial ultraviolet light either in induction or exacerbation of cutaneous lesions over nose. UV-exposure causes p53 mutations by inducing thymidine dimers thereby causing local immunosuppression. Although face, neck, and limbs all are exposed to sunlight, in these cases a few small sized lesions of PK are limited to the nose and adjacent perinasal area only. Thus, this subtype differs from all other types of PK except DSAP in relation to sensitivity to sunlight.

In DSAP, 15% of patients with facial lesions have lesions on other sun-exposed areas of the body [1].

DSAP is inherited as an autosomal trait with the gene locus identified at chromosome 12q and 15q [4]. The lesions begin to develop in the teenagers of affected families, with penetrance nearly complete by the third and fourth decades of life.

This variant neither exhibits any inheritable pattern nor any pre-malignant potential. Thus the actinic damage seems to cause limited injury, thereby resulting in localized form of PK.

This case is reported due to exclusive location of PK on nasal alae which owing to specific morphology, distribution and clinical course may be classified into separate category of "localized nasal actinic Porokeratosis". As more and more similar cases will be reported, the existence of this entity will be further clarified.

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