

Original Research Article

A retrospective clinicopathological study of cases of perforating dermatoses in a tertiary care centre

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ABSTRACT

Background: Perforating dermatoses is a group of diseases characterized by extrusion of dermal materials such as inflammatory cells or extracellular substances through the epidermis. They can be classified as primary and secondary perforating dermatoses. This study was done to assess the prevalence and associated systemic diseases of perforating dermatoses in our setting.

Methods: This is a retrospective study done by reviewing the hospital records of patients who attended the skin OPD, Government Rajaji Hospital, Madurai Medical College, Madurai during the period June 2008 to June 2010. All patients diagnosed clinically as perforating dermatoses and supported by histopathological examination were included.

Results: Kyrle's disease (54%) was the commonest type, followed by reactive perforating collagenosis (43%) and perforating calcific elastosis (3%). About 53% of the patients presented as hyperkeratotic papules and 43% as hyperkeratotic, plugged, umbilicated papules and plaques. Erythematous papules arranged in annular and serpiginous pattern in the periphery of the central atrophic plaques were noted in 1 patient (3%). Lower extremities (73%) were the most commonly involved site. (67%) patients had at least one systemic disease and commonest association was diabetes mellitus (53%), chronic renal failure (36%), hypothyroidism (3%).

Conclusions: Perforating dermatoses is a uncommon dermatological condition characterized by Trans Epidermal Elimination of dermal substances. Since it has a strong association with systemic diseases screening for systemic diseases is required for all cases.

Keywords: Transepidermal elimination, Primary, Secondary, Extremities, Systemic associations

INTRODUCTION

Perforating dermatoses is a heterogenous group of disorders characterized by trans epithelial elimination (TEE) in which material from the dermis is extruded through the epidermis to the exterior with little or no disruption of the surrounding structures. The extruded material may include inflammatory cells, red cells, microorganisms and extracellular substances, such as

mucin or altered connective tissue components.¹ Primary perforating dermatoses - Kyrle's disease (hyperkeratosis follicularis et parafollicularis in cutem penetrans), reactive perforating collagenosis (RPC), elastosis perforans serpiginosa (EPS), perforating folliculitis (PF). Associated with systemic diseases -renal disease and /or diabetes mellitus, malignancy, hepatic, endocrinological disorders, AIDS, tuberculosis, pulmonary aspergillosis, neurodermatitis, atopic dermatitis and scabies.²⁻⁹ The

clinical and histopathological findings may resemble any of these four primary perforating dermatoses (Kyrle's, RPC, EPS, PF).¹⁰ As a secondary component in primary dermatoses such as granuloma annulare, pseudoxanthoma elasticum, chondrodermatitis nodularis helices, lichen nitidus, lichen planus, keratoacanthomas and necrobiosis lipoidica.

Aims and objectives

- To delineate the clinical and histopathological features of various types of perforating dermatoses.
- To find out the potential relationship existing between the perforating dermatoses and systemic disease.

METHODS

Study design: This was a retrospective study.

Study place and period

The study was carried out by reviewing the records from June 2008 to June 2010 of patients with perforating dermatoses in the department of Dermatology, Government Rajaji Hospital, Madurai Medical College, Madurai.

Study group

Patients who were clinically diagnosed as perforating dermatoses and supported by histopathological confirmation were included in this study.

Procedure

This study was done by reviewing the records of patients who were clinically diagnosed as perforating dermatoses. Brief clinical history regarding the duration of disease, symptoms at time of presentation and history regarding

associated systemic diseases were reviewed. Histopathological correlation of the clinically diagnosed cases was reviewed. Comorbidities and associated systemic diseases of those cases was also obtained from the records.

RESULTS

Thirty patients were diagnosed as perforating dermatoses with clinical and histopathological features during the study period from June 2008 to June 2010.

Type of perforating dermatoses

Among the 30 patients of perforating dermatoses, Kyrle's disease was the commonest type as shown in Table 1.

Table 1: Types of perforating dermatoses.

Types of perforating dermatoses	No. of patients	%
Kyrle's disease	16	54
Reactive perforating collagenosis	13	43
Perforating calcific elastosis	1	3
Total	30	100

Sex distribution

20 patients were males and 10 patients were females as shown in Table 2.

Table 2: Sex distribution.

Types of perforating dermatoses	Male	Female
Kyrle's disease	11	5
Reactive perforating collagenosis	9	4
Perforating calcific elastosis	-	1
Total	20	10

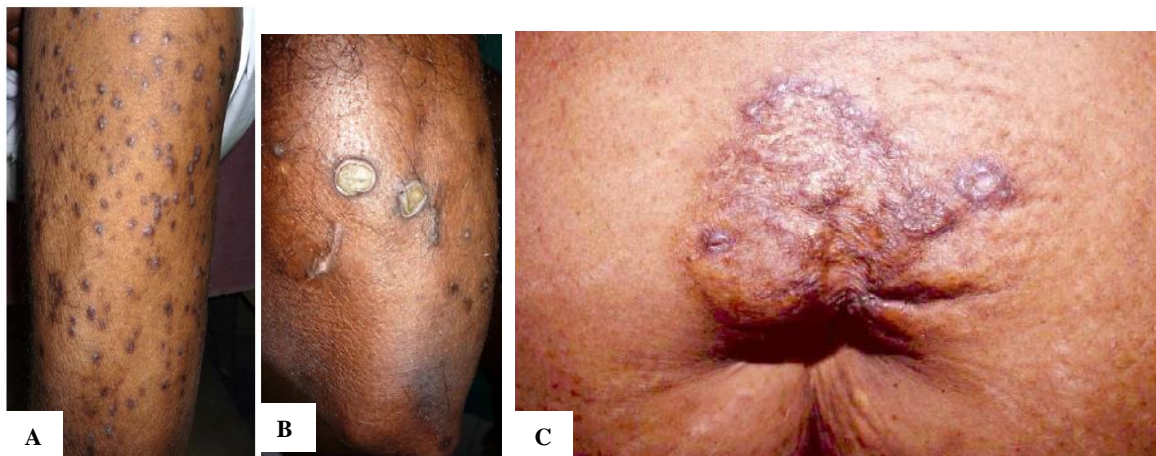


Figure 1: Clinical picture of perforating dermatoses. (A) Kyrle's disease: hyperkeratotic papules over thigh and back; (B) reactive perforating collagenosis: umbilicated papules and plaques over forearm and legs; (C) perforating calcific elastosis: plaque over periumbilical region.

Morphology of lesions

In 16 (53%) of the patients, the lesions were hyperkeratotic papules as in Figure 1 (A) and in 13 (43%) hyperkeratotic, plugged, umbilicated papules and plaques as in Figure 1 (B) were apparent. Erythematous papules arranged in annular and serpiginous pattern in the periphery of the central atrophic plaques as in Figure 1 (C) were noted in 1 patient (3%).

Most commonly involved site

Lower extremities (73%) were the most commonly involved site as shown in Table 3.

Table 3: Sites of involvement.

Region	No. of patients	Percentage (%)
Head	5	23
Trunk	17	57
Upper extremities	19	63
Lower extremities	22	73

Symptoms

The presenting symptoms of the patients were itching in 63% and pain in 7%. 30% of patients were asymptomatic as shown in Table 4.

Table 4: Symptoms.

Symptoms	No. of patients	Percentage (%)
Itching	19	63
Pain	2	7
Asymptomatic	9	30

Table 5: Biopsy findings

	KD	RPC	PCE
Biopsy findings			
Parakeratosis	12	8	-
Hyperkeratosis	16	10	1
Keratotic plugging	15	9	1
Acanthosis	16	11	1
Dyskeratotic cells	5	-	-
Transepidermal channel			
Keratin	16	9	1
Cellular debris	13	7	1
Altered collagen fibers	-	13	-
Altered elastic fibers	-	-	1
Papillary dermis			
Lymphocytes	10	4	1
Neutrophils	12	6	1
Altered collagen fibers	-	13	-
Altered elastic fibers	-	-	-
Reticular dermis			
Altered elastic fibers	-	-	1

KD: Kyrle’s disease; RPC: Reactive perforating collagenosis; PCE: Perforating calcific elastosis.

Histopathology

Histopathological evaluations revealed three types of lesions:

- Histopathological features of 16 (54%) cases showed epidermal invagination filled with keratotic plug admixed with cellular debris and neutrophils as shown in Figure 2 (A and B). In biopsy specimens of these 16 patients, Masson trichrome and elastic van Gieson stains were negative in the epidermis and in the crater as shown in Figure 2. Hence, the overall histological appearance in these 16 cases was consistent with Kyrle’s disease as shown in Table 5.

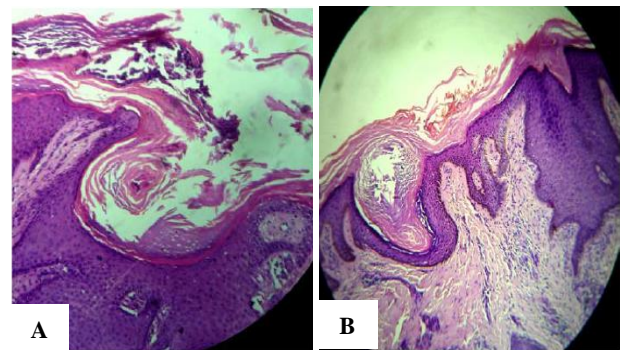


Figure 2: Histopathology of Kyrle’s disease. (A) Structure of epidermis showing a cup shaped invagination; (B) epidermal invagination filled with keratotic plug and cellular debris.

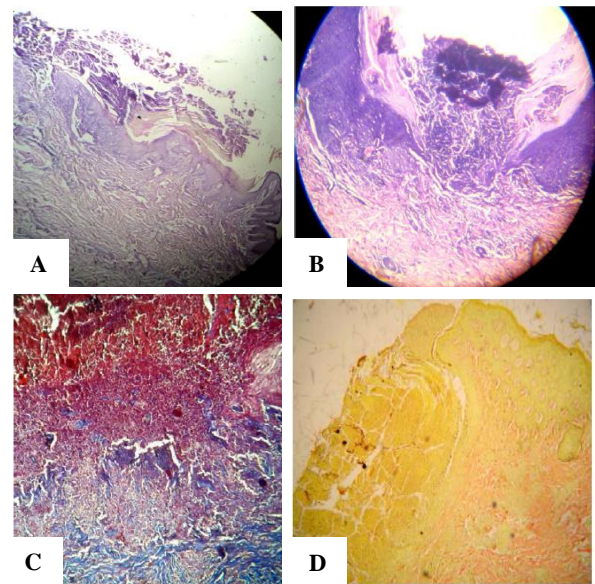


Figure 3: Histopathology of reactive perforating collagenosis. (A) Cup shaped invagination of epidermis filled with plug containing keratin and cellular debris; (B) transepidermal elimination of collagen fibres; (C) Masson trichrome stain for collagen showing transepidermal elimination of collagen stained blue; (D) Verhoeff van Gieson stain showing elimination of collagen stained red.

- Histopathological features of 13 (43%) cases showed cup shaped invagination of the epidermis filled with a plug consisting of keratin, cellular debris and neutrophils as in Figure 3 (A). There were vertically oriented collagen bundles at the base of the lesions in five of these thirteen cases as in Figure 3 (B). Masson trichrome staining showed transepidermal elimination of collagen in all these thirteen cases as in Figure 3 (C). Elastic van Gieson stains were negative in the epidermis and in the crater as in Figure 3 (D). The overall histological appearance in these 13 cases was consistent with reactive perforating collagenosis as shown in Table 5.
- In the last case, numerous altered basophilic elastic fibres encrusted with calcium salts seen in the reticular dermis as shown in Figure 4. Von Kossa stain was used to stain the calcium. Channels extruding inflammatory and elastotic debris from the reticular dermis to the overlying epidermis was also seen. Verhoeff van Gieson stain was used to stain the elastic fibres. The overall histological appearance in this case was consistent with perforating calcific elastosis as shown in Table 5.

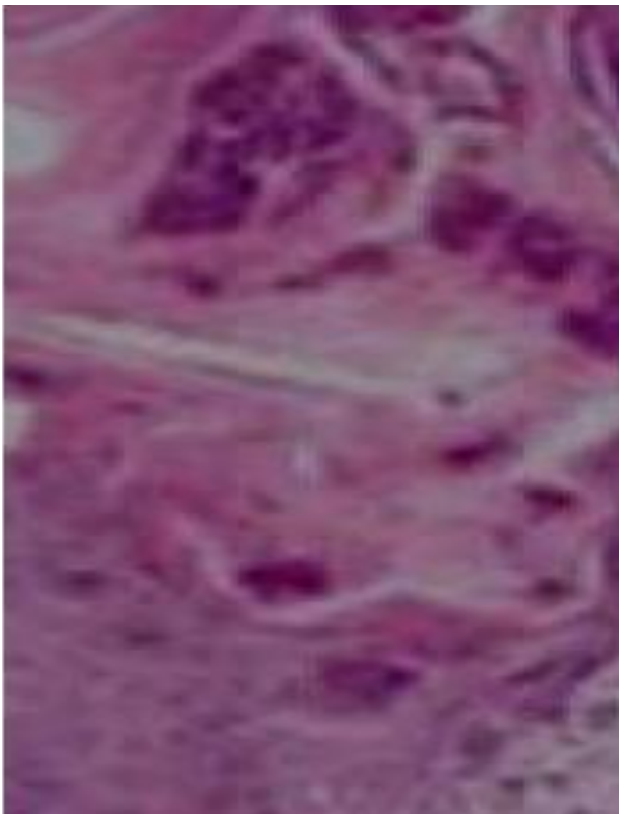


Figure 4: Perforating calcific elastosis: irregular calcified elastic fibres in reticular dermis.

Associated diseases

Twenty (67%) patients had at least one systemic disease. The commonest association was diabetes mellitus

followed by chronic renal failure, hypothyroidism and hypertension as shown in Table 6.

Table 6: Associated diseases.

Types of perforating dermatoses	DM	CRF	Hypothyroid	HT
Kyrle’s disease	12	11	1	-
Reactive perforating collagenosis	4	-	-	-
Perforating calcific elastosis	-	-	-	1
Total	16	11	1	1

DISCUSSION

30 cases of perforating dermatoses were diagnosed in about 1,15,368 patients who attended the skin OPD during the period of study.

Types of perforating dermatoses

In our study, Kyrle’s disease was the commonest type (54%) followed by reactive perforating collagenosis (43%) and perforating calcific elastosis (3%).

Sex distribution

Preponderance of males was noted in our study which is in concordance with the study by Saray et al.¹¹ On analyzing the incidence in the literature, it is varied in different studies.^{12,13} In Kyrle’s disease, a female preponderance has been noted in some series whereas Joseph et al has recorded an equal sex distribution.²⁵

Morphology

The lesions of perforating dermatoses had varied morphologies resembling Kyrle’s disease (hyperkeratotic papules) in 54%, reactive perforating collagenosis (umbilicated papules with central keratotic plug) in 43% and perforating calcific elastosis (erythematous papules arranged in the periphery of a central atrophic plaque in annular and serpiginous pattern) in 3%.

Distribution of lesions

Lower extremities are the most commonly involved site in acquired perforating dermatoses as stated by Saray et al who reported a frequency of 73 percent.¹¹ This was the case in our study, with extensor surfaces of the lower extremities involvement in 17 patients (81%), upper extremities in 12 patients (57%), trunk in 11 patients (52%) and head in 3 patients (14%) of acquired perforating dermatoses. Multisite involvement was noted in 15 patients (71%). However unusual presentation on the face and scalp were also observed. In perforating

calcific elastosis, Hicks et al and Kazakis et al suggested that the traumatic effect of repeated pregnancies on elastic tissue explain the localization of skin lesion to the periumbilical area as observed in our patient.^{15,18} Lower extremities are the most commonly involved site as stated by Saray et al who reported a frequency of 73%. This was the case in our study also.¹¹

Symptoms

Itching was the main complaint in 63% of our patients, which was most. Patient with early onset (14 years of age) of skin lesions presented to us at the age of 60 years with large intensely pruritic lesions which healed with scarring, indicating that the lesions become more profuse and enlarged with age unless treatment is given. Similar observations were also made by Kachhawa et al and Bhat et al in their studies.^{13,18} Two patients (7%) had pain, which is an unexpected symptom for acquired perforating dermatoses. Saray et al reported pain in 9 percent of patients in his study.¹¹

Histopathology

As previously stated in the literature, we observed histopathological features consistent with each of the perforating dermatoses. Kyrle like histopathological features were the most common (54%), followed by reactive perforating collagenosis (43%) which is in concordance with the study done by Saray et al.¹¹ Perforating calcific elastosis like histopathological feature was noted in one patient. Patterson documented the lesions with histological features of perforating folliculitis, reactive perforating collagenosis and Kyrle's disease in the same patient with acquired perforating dermatoses.²⁰ Moreover, Rapini et al reported combined transepidermal elimination of both collagen and elastin in their four patients with acquired perforating dermatoses.¹⁰ They proposed that varying histological findings in this disease may represent the different stages or different types of lesions in the same pathological process. However, we did not observe such overlapping features in our patient group. Saray et al in their study also recorded the similar findings like our study.¹¹ Moreover, it is also possible that we have been unable to observe the different stages of the histopathological evaluation because of the fact that we have taken only one biopsy from each patient. However, on the basis of distinctive histopathological findings with the absence of overlapping histopathological features in our study, we think that acquired perforating dermatoses consists of four different types of perforating disorders, namely, Kyrle's disease, reactive perforating collagenosis, perforating folliculitis and elastosis perforans serpiginosa like perforating disorders. Patterson, in his study noted both collagen and elastin in the early, non umbilicated lesions, whereas collagen was observed in the umbilicated ones. Fibrous component was the only material eliminated in late lesions, probably due to the degeneration of both collagen and elastin. In contrast to these early findings, we observed neither collagen nor

elastin elimination in most of the non umbilicated lesions. However, collagen elimination was observed in the umbilicated ones, which was similar to the findings in the above mentioned study.

Associations

In our study, majority (67%) of our patients had at least one systemic disease, and diabetes mellitus (76%) and chronic renal failure (52%) were the most common diseases associated with acquired perforating dermatoses. Two of our patients had insulin dependent diabetes mellitus (IDDM), and most (14) had noninsulin dependent diabetes mellitus (NIDDM) similar to study by Saray et al.¹¹ Other investigators have also found that NIDDM is more frequent in patients with acquired perforating dermatoses.^{4,21} In contrast, Morton and coworkers observed that acquired perforating dermatoses is more often associated with IDDM compared with NIDDM.¹⁷

In diabetes mellitus, the proposed mechanism is that, trauma from scratching may cause dermal necrosis due to poor blood supply that results from vasculopathy, with necrotic material then being extruded through the epidermis. Chronic renal failure was the second most frequent (52%) disease associated with acquired perforating dermatoses in the present study. In 9 of eleven patients (82%) with chronic renal failure, the cause of kidney disease was diabetes mellitus, and in 2 (18%) glomerulonephritis, findings similar to studies done by Saray et al and Joseph et al were seen. In the literature, epidermal or dermal abnormalities such as alterations in collagen or elastic fibres due to metabolic disturbances related to chronic renal failure have been suggested as underlying factors in acquired perforating dermatoses patients with chronic renal failure.^{11,14} In patients with chronic renal failure, acquired perforating dermatoses often occurs following dialysis.^{17,21} However, the lesions may also start in the pretreatment period.^{17,22} In our study, acquired perforating dermatoses developed after the initiation of dialysis treatment in five patients with chronic renal failure. Hypothyroidism was associated in one patient with acquired perforating dermatoses. Association of acquired perforating dermatoses with hypothyroidism has been described in earlier studies by Faver et al.⁴ Hypertension was seen in one patient with periumbilical perforating calcific elastosis, who was an obese, multiparous woman with no characteristic lesions and family history of pseudoxanthoma elasticum.

There was no systemic association seen in any of our patients with familial reactive perforating collagenosis as described in earlier studies by Bhat et al.¹²

Interesting observation

An unusual finding of this study was that Kyrle's disease was detected in one patient in whom neither a systemic disease nor any other dermatological problem was

observed. There are only few reports of acquired perforating dermatoses occurring in otherwise healthy people. Kalla et al and Saray et al have reported similar cases in their studies.^{11,25}

CONCLUSION

Perforating dermatoses is a rare skin disease comprising only a very small percentage of patients attending the skin OPD. Kyrle's disease is the most common perforating dermatoses. Male preponderance is noted. Clinicopathological examination is important in arriving at the diagnosis. Non-insulin dependent diabetes mellitus is the most frequent association seen with acquired perforating dermatoses. An interesting case of acquired perforating dermatoses with no systemic association is also noted.

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Conflict of interest: None declared

Ethical approval: The study was approved by the institutional ethics committee

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