

# Necrotic plaques in an elderly male

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## Lesson

Reactive perforating collagenosis is commonly recognised as an unusual form of transepithelial elimination of collagen and elastin fibres which are extruded through the epidermis in patients with a genetic predisposition or underlying diseases, such as diabetes mellitus or renal diseases. We present the unusual case of an 87-year-old diabetic male with a giant form of reactive perforating collagenosis and review the available literature.

## Keywords

dermatology, diabetes, drugs and medicines, endocrinology, geriatric medicine, other geriatric medicine

## Introduction

Reactive perforating collagenosis may present in a giant form in elderly patients in the absence of end stage renal failure.

## Case report

An 87-year-old male was admitted to the medical ward with sepsis secondary to a lower respiratory tract infection. He was subsequently referred to dermatology for a review of generalised pruritus associated with development of increasing numbers of generalised purulent and necrotic plaques (Figure 1). History was unobtainable due to sepsis-related delirium but the lesions were noted to have started after admission two months ago. He had a history of hypertension, atrial fibrillation, ischemic heart disease, treated pulmonary tuberculosis, type 2 diabetes and diabetic nephropathy (Chronic Kidney Disease stage 3a) with a pacemaker in situ. His medications included Apixaban, Amiloride, Nifedipine and Bumetanide.

Examination revealed well-defined disseminated symmetrical large round and oval plaques with areas of exposed dermis and subcutis ranging from 10 mm to 45 mm on erythematous bases. The older plaques had necrotic centres with evidence of purulent exudate from newer lesions over the back (Figure 2).

Blood tests showed raised inflammatory markers, stable renal function and a normal autoimmune screen. An echocardiogram ruled out infective endocarditis as a cause of the skin changes.

A 4 mm punch biopsy demonstrated mild hyperkeratosis with necroinflammatory exudate, erosion and subepidermal clefting, neutrophil extravasation in the upper dermis and evidence of perforating elastic fibres by Elastic van Gieson and trichrome stains (Figure 2).

## What is your diagnosis?

Giant perforating collagenosis and elastosis.

## Summary

Reactive perforating collagenosis is commonly recognised as an unusual form of transepithelial elimination of collagen and elastin fibres which are extruded through the epidermis in patients with a genetic predisposition or underlying diseases, such as diabetes mellitus or renal diseases.

Giant Acquired Reactive Perforating Collagenosis should be considered in the differential diagnosis of ulcerative plaque lesions of larger sizes compared to the usual size ranging between 2 and 8 mm and the largest lesions have been noted to be 2 cm.<sup>1–3</sup>

## Definition and classification

Due to the complexity of perforating disorders, it was suggested to classify the disease as follows: (i) perforation as an incidental histological finding; (ii) secondary perforation (e.g. in granuloma annulare, pseudoxanthoma elasticum and chondrodermatitis nodularis chronica helices) and (iii) primary perforating dermatoses.<sup>1</sup>

Primary perforating dermatosis is used as an umbrella term for four different entities: Elastosis Perforans Serpiginosa, Acquired Reactive Perforating Collagenosis, Perforating Folliculitis and Kyrle's disease.<sup>1</sup>

## Causes and genetics

An interaction of keratinocytes with altered structural proteins is likely to be the triggering factor to develop reactive perforating dermatosis. Elastin receptor 67-kDa overexpression in Elastosis Perforans Serpiginosa and Transforming growth factor beta (TGF- $\beta$ 3), Matrix metalloproteinase 1 (MMP-1) and Tissue inhibitor of metalloproteinase 1 (TIMP-1) overexpression in Acquired Reactive Perforating Collagenosis, indicate changes in extracellular matrix composition.<sup>1</sup>

There have been reports of Acquired Reactive Perforating Collagenosis in a paraneoplastic form secondary to underlying solid organ and haematological malignancies.<sup>2,4</sup>

Acquired Reactive Perforating Collagenosis may be associated with use of immunotherapeutic agents including epidermal growth factor receptor inhibitors (tyrosine kinase-i) including Erlotinib, Sorafenib and Lenvatinib and use of Ranibizumab (anti-angiogenic monoclonal antibody used in wet age-related macular degeneration).<sup>5,6</sup>

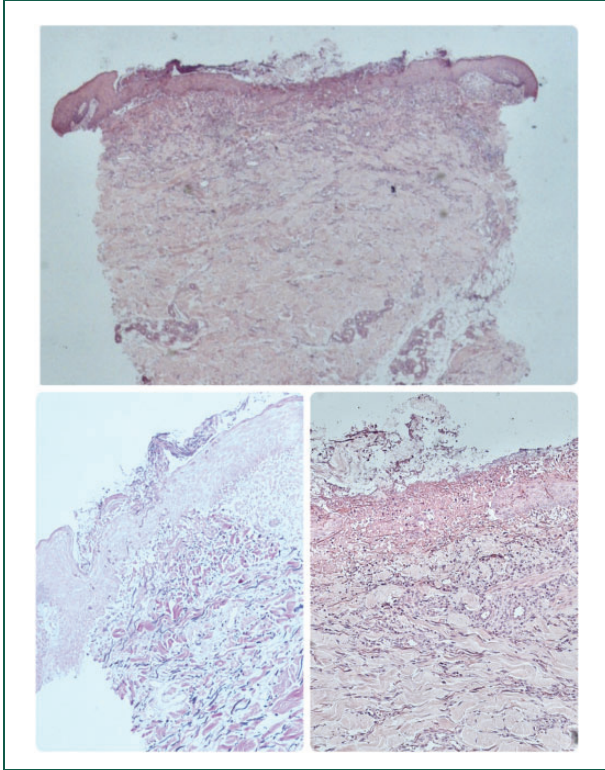
## Dermatoscopic features

Ormerod et al. analysed some dermatoscopic features of reactive perforating collagenosis which included a hyperkeratotic plug appearing as yellow brown structureless areas surrounded by an erythematous rim of inflammation.<sup>7</sup>

**Figure 1.** Clinical photography during admission. Suggestive of generalised large well defined plaques ranging from 10-45 mm in size with erythematous borders and purulent centres in newer lesions over the back and necrotic centres in older lesions over the scalp, fingertips, thighs, and abdominal wall.



**Figure 2.** Histopathological appearances of a lesion from the back on which a 4 mm punch biopsy was performed (a – Elastic Van Gieson stain showing perforating elastic and collagenous material through the epidermis, b – H&E stain at 20× magnification, c – H&E stain at 100× magnification).



## Treatment options

Reactive perforating collagenosis may be self-limiting with spontaneous resolution over months. However, when significantly symptomatic and leading to a poor quality of life or complications, treatments have been trialled. It is a general understanding that underlying disease should be controlled alongside dermatological treatment for successful management. Topical potent steroids are the first line treatment options but in case of failure successful treatment with phototherapy with Narrow Band UVB/PUVA has been reported. Topical benzoyl peroxide, topical isotretinoin gel 0.1% OD and improvement with oral allopurinol and oral acitretin (suggestive of follicular plugging being the initial pathogenesis) have been reported. In case of biologics causing the disease, when these have been stopped, the disease shows complete reversal.

## Learning points

- Giant acquired reactive perforating collagenosis should be considered in the differentials of all

hyperkeratotic plaques or in disseminated ulceration in patients with diabetes with renal impairment.

- With the advent of newer immunotherapy agents, it may be possible that more of these cases may be seen over time as a side-effect of treatment.
- It should also be considered as a paraneoplastic phenomenon in the absence of a history of diabetes or renal impairment.

## MCQs

1. What are the 2 most common associations with Acquired reactive perforating collagenosis?
  - (a) Diabetes and renal impairment
  - (b) Hypothyroidism and diabetes
  - (c) Hypertension and hypothyroidism
  - (d) Infective endocarditis
2. Which one of these drugs can cause Acquired Reactive Perforating Collagenosis?
  - (a) Warfarin
  - (b) Tyrosine kinase inhibitors
  - (c) Metformin
  - (d) Topical steroids

## Declarations

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**Guarantor:** BG

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