A 72-year-old man was referred with suspected cutaneous metastases from a renal cell carcinoma treated by right partial nephrectomy. There was an 18-month history of persistent erythema and erosions of the right inguinal fold, which had failed to respond to all topical therapy. He had a 2-year history of immunosuppression with prednisolone and azathioprine for rheumatoid arthritis, and had had a transurethral resection of prostate for benign prostatic hyperplasia in 1992. There was no evidence of metastatic spread of his renal malignancy. He had multiple squamous proliferative lesions on his face and scalp consistent with nonmelanoma skin cancer (NMSC). There was marked erythema with superficial erosion, exudate and patchy scaling on the right inguinal fold extending on to the right hemiscrotum consistent with extramammary Paget’s disease (EMPD). A firm papule was palpable in the centre of the right inguinal fold. Full blood count was normal. There were no paraproteins or Bence–Jones proteins. Prostate-specific antigen (PSA) was elevated at 9.4 μg L⁻¹. Computed tomography of the chest, abdomen and pelvis showed no evidence of metastatic spread and no local underlying pathology. Scalp and face biopsies confirmed squamous and basal cell carcinomas. Biopsy of both the inguinal fold and right hemiscrotum showed features of EMPD and infiltration of the epidermis by clusters of adenocarcinoma cells. A biopsy from the papule showed invasive adenocarcinoma of primary appendageal origin which stained positive with MNF116, AE1, AE3, CK7, CK20 and CAM5.2 and negative with S100, PSA, PSAP and vimentin. The NMSCs were treated surgically. In view of the extent of skin involvement by EMPD, excision of the primary appendageal carcinoma was performed initially. Following excision of the appendageal tumour there was complete resolution of the remaining skin affected by EMPD. Four months following excision there is no evidence of recurrence of the EMPD or evidence of metastatic spread from his renal cell carcinoma. EMPD is a rare disorder occurring mainly in apocrine gland-bearing regions. The histogenesis is controversial, with several investigators proposing that it is a heterogeneous entity with some cases representing a de novo adenocarcinoma in situ arising in the epidermis and others being epidermotropic metastases or a direct extension of an associated internal malignancy. The association of subjacent apocrine carcinoma and EMPD is well recognized. Treatment of EMPD is often difficult due to the site and extent of cutaneous involvement. Regression of EMPD after excision of the proxim-
antiphospholipid antibodies should be recommended in all patients with anetoderma. To our knowledge, this is the first case of anetoderma secondary to lupus profundus with positive antiphospholipid antibodies.

DP-21

Coexistence of elastoma and lichen sclerosus on the vulva

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A 77-year-old woman presented with vulval pruritus and soreness. Clinical examination revealed ivory plaques on the inner aspects of both labia minora characteristic of lichen sclerosus (LS). She exhibited destruction of vulval architecture with fusion of the labia minora and majora.

Ten years later, she developed a slightly yellowish papular area with focal hyperpigmentation on her right labium minus. This was biopsied and histology showed mild basal hyperpigmentation but no melanocytic or keratinocyte atypia. The underlying connective tissue showed a nodular collection of elastic tissue confirmed by positive Verhoeff van Gieson stain.

Our patient showed a coexistence of two different diseases: LS – a condition commonly seen on the vulva and elastoma – a rare hamartomatous condition characterized by circumscribed defects in the development of the dermis. There is only one previous case report of an elastoma on the vulva. Interestingly, this patient also had LS (Sanchez-Yus E, Aguilar A, Requena L et al. Naevus elasticus and lichen sclerosus et atrophicus on the vulva. Cutis 1990; 45: 252–5).

A study of elastic fibres in the advanced stages of LS showed the presence of newly formed collagen fibres and elastin or elastin-like substances (elastocollagenous mass) in the dermal homogeneous zones (Mihara Y, Mihara M, Hagari Y, Shimao S. Isolation and partial characterization of an elastase-type protease in human vulva fibroblasts – its possible involvement in vulvar elastic tissue destruction of patients with lichen sclerosus et atrophicus. J Invest Dermatol 1982; 78: 270–5).

We describe the occurrence of an elastoma on a background of long-standing LS. We postulate that the remodelling of the elastic fibres in advanced LS may have contributed to the formation of the elastoma.

DP-22

Palisaded neutrophilic granulomatous dermatitis in rheumatoid arthritis

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We describe two patients with long-standing rheumatoid arthritis (RA) who developed skin lesions with histological features of palisaded neutrophilic granulomatous dermatitis (PNGD).

Patient 1, a 60-year-old woman, presented with a 5-year history of annular lesions consisting of peripheral papules and a few pustules with central clearing affecting the trunk, thighs and arms. She had a 55-year history of seropositive RA previously treated with a number of disease-modifying drugs. Histology revealed small granulomas composed of a palisade of histiocytes around collagen fibres with central necrobiosis, neutrophils and neutrophilic fibrin deposition. Leucocytoclastic vasculitis was seen in a further biopsy. Hydroxychloroquine 200 mg twice daily led to regression of the lesions after 6 weeks of treatment. However, she developed a recurrence when this drug was discontinued due to gastrointestinal symptoms.

Patient 2, a 55-year-old woman, presented with a 1-year history of multiple, tender violaceous papules over the buttocks and thighs. She had an 18-year history of severe seropositive RA treated with multiple disease-modifying drugs. Histology revealed macrophages present in a palisaded arrangement around degenerative collagen fibres and foci of neutrophil infiltration and nuclear dust. Due to the severity of her arthritis and lack of response to disease-modifying drugs she was treated with etanercept, which improved both her arthritis and skin eruption.

PNGD is recognized most in patients with RA but is also reported in lupus erythematosus, other connective tissue diseases, lymphoproliferative conditions and inflammatory bowel disease (Chu P, Connolly MK, LeBoit PE. The histopathologic spectrum of palisaded neutrophilic and granulomatous dermatitis in patients with collagen vascular disease. Arch Dermatol 1994; 130: 1278–83). PNGD has not been completely defined clinically or histologically, previously being described under a variety of terms including rheumatoid papules, interstitial granulomatous dermatitis with cutaneous cords and arthritis and Churg–Strauss granuloma. However, the literature supports evidence that this condition represents a spectrum of lesions with the clinical appearance ranging from asymptomatic nonspecific macular rash to painful, infiltrative papules and nodules found on the extensor surfaces of the limbs (Sangueza OP, Caudell MD, Mengesha YM et al. Palisaded neutrophilic granulomatous dermatitis in rheumatoid arthritis. J Am Acad Dermatol 2002; 47: 251–7). Histologically, early lesions may reveal a sparse mixed inflammatory infiltrate or leucocytoclastic vasculitis with evidence of collagen damage. Fully developed lesions show palisading granulomas surrounding degenerate collagen and neutrophilic debris with fibrin deposition within the granulomas. Fully developed lesions may resemble granuloma annulare (GA) clinically and histologically; however, the collagen bundles are thin in GA and thick in PNGD, and mucin is more commonly found in GA and fibrin in PNGD. In later lesions there are palisaded granulomas with dermal fibrosis. Immunoreactants within blood vessel walls and the dermis have been demonstrated by direct immunofluorescence, suggesting that this condition is immune complex-driven.

The clinical and histopathological findings in our patients reflect the diversity described above and add to the reported patients with this entity.