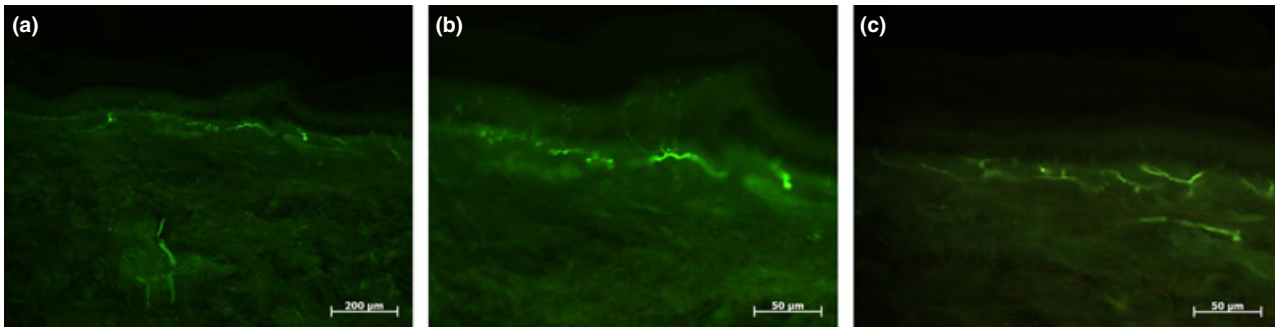


**Figure 1** PGP 9,5 immunolabelling on a skin biopsy showing small-fibre neuropathy. (a) Subepidermal neural plexus underlines the basal membrane, in the deep dermal innervation of a sweat gland. (b) Proximal thigh. Numerous intraepidermal fibres running towards the surface and fibre branching in a case of preserved proximal fibre density. (c) Distal leg. The density of intraepidermal fibres is reduced when compared with the proximal biopsy.

The density of IENF in distal biopsies was decreased (Fig. 1) in eight patients (not in the case of Mrs.A). It was decreased only two times in proximal biopsies (not in Mrs. A).

The limited number of cases and the absence of controls in our study does not allow for the calculation of correlations. However, the evaluation of cold sensitivity threshold appears more sensitive than other thresholds in our hands and the study of IENF density appears as a very reliable tool for diagnosis.

In the case of Mrs A., QST showed abnormalities in cold sensitivity thresholds that were similar to those observed in SFN, but the final diagnosis was not SFN but rather simulation or psychogenic pruritus thanks to the measurement of IENF density.

QST remains a good tool for SFN diagnosis but is subjective.[6] In contrast, the analysis of IENF is a better method for diagnosis of SFN because it is faster, more objective and less expensive than QST.

## Acknowledgement

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# Anti-CCP antibody in a patient with generalized discoid lupus erythematosus, vasculitis and acute interstitial lung disease but not rheumatoid arthritis

*Editor,*

Presence of the rheumatoid arthritis (RA)-specific anti-cyclic peptide (anti-CCP) has been shown in two studies to be associated with interstitial lung disease (ILD) in patients without clinically apparent articular RA.[1,2] The lung phenotypic characteristics described in these patients resembled those of established RA.[1] Furthermore, only a few of these patients had developed articular RA over a short duration of follow-up.[1] However, none of these patients had cutaneous manifestations or acute fulminant ILD.[1,2] Herein, we describe a gentleman who developed generalized discoid lupus erythematosus (DLE) of mixed morphologies, nail fold infarction and acute ILD associated with high titres of anti-CCP and RF (rheumatoid factor) without articular RA.

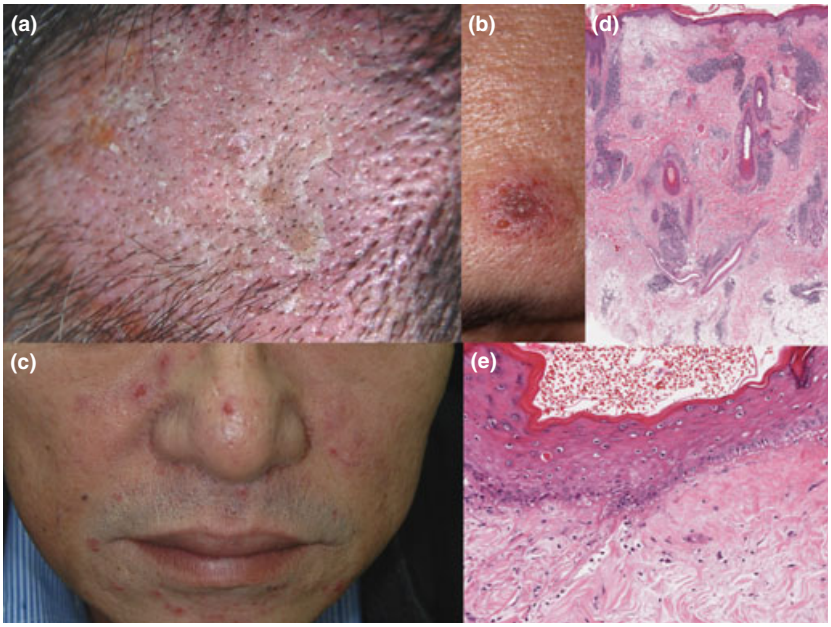


**Figure 1** (a) Large, tender, erythematous and scaly plaque on his scalp without alopecia. (b, c) Non-tender, erythematous, coin-shaped plaque and acneiform papules on his face. (d) Biopsy of the scalp lesion showed parakeratosis, diffuse periappendage lymphocytic infiltrations, mucin deposition, (e) basal vacuolation, dyskeratotic cell and thickened basement membrane.

A 62-year-old business man who was a carrier of hepatitis B and had a 20-pack-year history of smoking presented with a 1-month history of facial and scalp eruptions. Examination revealed multiple, tender, erythematous and scaly plaques on his scalp without alopecia. Coin-shaped plaques and acneiform papules were also found on his face (Fig. 1a–c). Biopsy of the scalp lesion was diagnostic of DLE (Fig. 1d and e). His regular health check-up 4 months prior including a chest radiograph was unremarkable. Rest of the examination and history were non-contributory. His complete blood count, liver and renal function tests, serum creatinine kinase level and urine routine were unremarkable. His anti-nuclear, anti-dsDNA, anti-Smith, anti-Ro, anti-La, anti-Jo-1, anti-Scl-70, anti-RNP antibodies and hepatitis B DNA viral load were within normal limits. He had minimal response to 20 mg of daily oral prednisolone and topical corticosteroid for 3 weeks. At 4th week he returned with multiple DLE on his back and buttock (two biopsies with direct immunofluorescence studies), periungual pain of several fingers and nail fold infarction of his left thumb (Fig. 2). He also had mild fever, sore throat, hoarseness, rhinorrhea, dry cough and significant pain of both helices of the ears without any rash. The RF (364 IU/mL) and anti-CCP (>340 IU/mL) were highly elevated with mild reduction of complement three level. He was admitted 4 days later with severe respiratory distress and subsequently received a combination of antibiotics and corticosteroid (up to 500 mg methylprednisolone QD for 2 days). Rest of the extensive investigations and infectious screen was notable for a mild elevation of C-reactive protein. Computer tomography scan of the chest (Fig. 3) and the bronchoalveolar larvage results were compatible with ILD. The bronchoalveolar lavage fluid was tested negative for atypical microbes including pneumocystis jiroveci. His blood and sputum cultures were negative. He was tested negative for influenza viral and urinary legionella antigens. His lung function deteriorated despite treatment, his hoarseness never improved, he had no further cutaneous or articular manifestations. He died of respiratory failure 4 weeks after medical treatment.

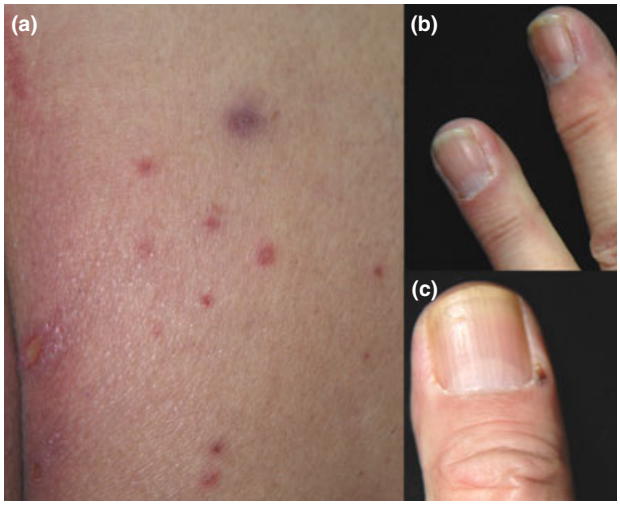


**Figure 2** (a) Multiple erythematous, purpuric and ulcerated papules and plaques on his Buttock. (b) Periungual erythema, swelling and tenderness. (c) Nail fold infarction of his left thumb.

Rheumatoid arthritis and DLE may coexist; however, no definitive link has been established.[3] The authors are unaware of any association between DLE and ILD or DLE and vasculitis.[3–5] The recent hypothesis that RA-specific autoimmunity may be

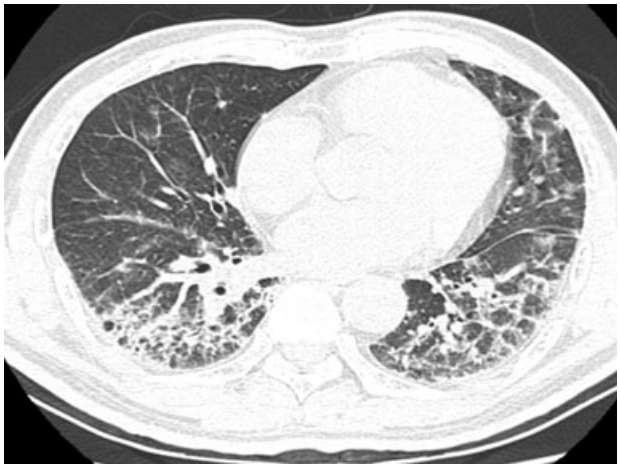

**Figure 3** Computed tomography scan of the chest showed interlobular septa and interstitial thickening with mild peribronchial infiltration, more in bilateral lower lungs, which were compatible with interstitial lung disease with pulmonary fibrosis.

generated due to immunologic interactions in the lung related to smoking may provide a clue to the cause of the medical problem of this patient.[1,2] While ILD may rarely precede articular RA in patients with high levels of anti-CCP antibody, acute fulminant ILD had not been described.[1,2,4] To the best of our knowledge, this is the first report of a patient who developed acute fulminant ILD following acute generalized DLE with highly elevated levels of Anti-CCP antibody and RFs without apparent RA. Further studies are required to elucidate the observations reported here.

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# Extragenital subcutaneous cellular angiofibroma of the elbow

*Editor,*

Cellular angiofibroma (CAF) is an uncommon well-circumscribed benign mesenchymal tumour occurring mainly in the genital region of both genders.[1,2] Extragenital CAF is rare and to the best of our knowledge, there has been no report of it occurring in the upper extremities. Herein, we describe the first case of extragenital subcutaneous CAF of the elbow.

A 43-year-old Japanese man presented with a skin coloured, slightly elastic hard, well-circumscribed, dome-shaped subcutaneous nodule measuring 32 × 28 mm in diameter without pain or tenderness on the flexural side of his left elbow that had developed over 2 years (Fig. 1a). T2-weighted magnetic resonance image disclosed a well-defined, solid subcutaneous mass indicating a non-specific soft tissue tumour (Fig. 1b). The lesion was completely excised with a rim of normal tissue. The subcutaneous tumour was encapsulated and easily detached from surrounding tissue (Fig. 1c). Microscopically, a spheroid well-circumscribed tumour measuring 35 × 30 mm was observed in the subcutis (Fig. 1d). The tumour was composed of fusiform cells forming short interlacing bundles, a vascular network and interstitial fibrous tissues. The abundant vessels were small to medium in size, some with thick hyalinized walls or fibrous stroma (Fig. 2a). The spindle-shaped cells showed ovoid uniform bland nuclei with palely eosinophilic cytoplasm. Mitoses, haemorrhage, or necrosis were not observed (Fig. 2b). An immunohistochemical study disclosed that CD34 (Fig. 2c) and CD31 were positive for blood endothelial cells, but negative for spindle cells, and α-smooth muscle actin (α-SMA) was positive for blood smooth muscle cells, but negative for spindle cells. CD99, S-100 protein and desmin (Fig. 2d) were all negative for spindle cells. We diagnosed this case as extragenital subcutaneous CAF. There has been no recurrence for 4 months.

Cellular angiofibroma was first described by Nucci *et al.* in 1997 as a distinctive mesenchymal neoplasm composed of spindle cells and the prominent vasculature.[1] Iwasa and Fletcher described a series of 51 cases of CAF to characterize its clinicopathologic and immunohistochemical features; these were 26 women and 25 men, aged from 22 to 78 years (mean 53.5).[2] Most common sites were the vulvovaginal and inguinoscrotal regions. All tumours consisted of bland, spindle-shaped cells, short bundles of wispy collagen and numerous small- to medium-sized thick-walled vessels. By immunohistochemistry, 29 of 48 tumours (60%) expressed CD34, 21% SMA and 8% desmin, but none expressed S-100 protein. We ruled out spindle cell lipoma because of the lack of adipocytic component, solitary fibrous tumour due to the absence of alternating hypercellular