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CONCISE REPORT

Cutaneous lupus erythematosus on the elbows: a peculiar localization

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Background: The presence of lesions limited to the elbows as expression of a cutaneous lupus erythematosus (CLE) is very unusual.
Objectives: To describe the clinical and microscopic characteristics of these lesions, as well as their relationship with the different cutaneous lupus erythematosus subsets.
Methods: Seven cases of CLE with lesions on the elbows, collected from 1998 to 2009, were retrospectively analysed. All patients had a previous or concomitant diagnosis of lupus erythematosus based on other typical skin lesions for each subtype of lupus erythematosus, and they all met clinical and microscopic criteria.
Results: Six patients were women with a mean age of 33.8 years. Five cases had been previously diagnosed with CLE (three lupus erythematosus [LE] tumidus, one subacute CLE and one acute CLE). In the other two cases, these lesions were the first manifestation of the disease. The lesions consisted of pruriginous, erythematous papules and plaques with a slightly scaly surface, located on both elbows. Microscopically, in addition to the typical features of CLE, other changes were observed, including alterations in the texture and the staining properties of collagen fibres (five cases), an interstitial histiocytic infiltrate (four cases), eosinophils (one case) and a Churg–Strauss granuloma (one case).
Conclusions: These peculiar lesions located on the elbows should be included in the spectrum of cutaneous manifestations of LE. Their histopathology combines changes of both LE and interstitial granulomatous dermatitis. *Lupus* (2012) **21,** 84–88.

Key words: cutaneous lupus erythematosus; discoid lupus erythematosus; lupus tumidus; subacute cutaneous lupus erythematosus

Introduction

The spectrum of cutaneous lesions that can be seen in the context of a lupus erythematosus (LE) is clinically and microscopically very rich and includes specific and nonspecific lesions.[1] Specific skin lesions of cutaneous LE (CLE) are classified as acute CLE (ACLE), subacute CLE (SCLE) and chronic CLE (CCLE), according to their clinical characteristics.[2] In the last few years lupus erythematosus tumidus (LET) has been recognized as a new entity, probably related to the spectrum of CLE, although its exact position in some of the previously defined CLE subsets is still a matter of debate in the literature.[3–7] Cutaneous lesions of LE, regardless of type, are mainly located on the face, neckline, back and dorsal aspects of the limbs. The presence of lesions limited to the elbows, with or without other manifestations of CLE, is very unusual. In the literature only one case of CLE with lesions limited to the elbows, considered as a LET, has been reported. We describe seven patients with CLE who developed these peculiar lesions on the elbows.

Material and methods

From 1998 to 2009, seven cases of CLE with lesions on the elbows were collected. The clinical and microscopic data of these lesions, as well as other

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clinical and immunological characteristics of the accompanying CLE, were retrospectively analysed. All the skin biopsy specimens were fixed in a 10% solution of formaldehyde and embedded in paraffin. Four-micrometre thick sections were stained with haematoxylin–eosin and processed for histological examination and evaluation. Furthermore, antinuclear antibodies (ANAs) were assayed using a standard indirect immunofluorescence technique with commercially available Hep-2 cells.

Results

Most of the patients were women (85.7%), with a mean age of 33.8 years (range 20–54 years old). Three of the patients were sisters (cases 2, 3 and 5) with two of them being twins (cases 2 and 3). Five cases of CLE had been diagnosed (three LET, one SCLE and one ACLE) between 2 and 8 years previously (mean 5.5). In the other two cases, simultaneous lesions on both elbows and the face were the first manifestation of the disease. In these cases clinical and histopathological features of the facial lesions were compatible with LET. Two cases presented systemic involvement, consisting of leukopenia and arthritis, and had positive ANAs (cases 3 and 7). In addition, one of them (case 7) had positive anti-Sm and anti-ribonucleoproteins (RNP) antibodies and delivered a baby who developed neonatal LE.

In all cases, the lesions located on the elbows consisted of erythematous papules and plaques with a slightly scaly surface. Both elbows were symmetrically affected (Figure 1). Pruritus was a constant symptom in all cases. Lesions were persistent over weeks or months with some clearing during winter and worsening during summer time.

With the exception of cases 4 and 6, in whom the lesions on the elbows were seen simultaneously with LET lesions on the face, in the rest of the cases the recurrent breakout of lesions on the elbows was not accompanied by lesions in other areas.

Histopathological findings of all patients are summarized in Table 1. In all biopsies a mild to moderate perivascular lymphocytic infiltrate was observed (Figure 2a). In three cases, the perivascular infiltrate was well demarcated and had a 'coat-sleeve' distribution (Figure 2b). In three cases a moderate lymphocytic infiltrate around the appendages was also found. Vacuolar change of the basal layer was focal in six cases (Figure 2d) and extensive in another. A variable degree of mucin deposition in the dermis and around the adnexae appeared in all specimens. In five cases, alterations in the texture and the staining properties of collagen fibres were evident (Figure 2c). In six cases, these changes were associated with an interstitial histiocytic infiltrate, with many eosinophils in one of them (case 3). Furthermore, one of the biopsies (case 7) showed a granulomatous infiltrate forming a ring around basophilic degenerated collagen (Churg–Strauss granuloma).

The lesions showed no response to topical steroids. In three cases, hydroxychloroquine sulphate treatment was started because of intense pruritus. The lesions improved or healed without scarring, with rapid recurrence after discontinuation of oral treatment.

Discussion

The presence of lesions on the elbows as the unique cutaneous manifestation of the flare of LE is very infrequent. The only reference found in the literature is a woman with a 10-year history of recurrent erythematous papules and plaques on the elbows during the summer.[8] The microscopic changes were compatible with LET and the lesions resolved with topic steroids and sunscreens, without recurrences after a follow-up of 12 months. In our cases the clinical picture was quite similar, consisting of some itchy erythematous papules and plaques with a slightly scaly surface, which settled on both elbows in a perfectly symmetrical pattern. The lesions showed a trend to persist over time, with alternating periods of improvement and worsening during winter and summer time, respectively. When these are compared with other reported cases, two facts are worthy of mention. Our patients, in addition to these similar lesions, had previously or at the same time had other cutaneous lesions with typical features of CLE, classified as SCLE in one patient, ACLE in another and LET in the rest. Despite this, in most cases when lesions on the elbows appeared they constituted at this time the only cutaneous manifestation of the disease. Moreover, the fact that three of the patients in the series are related point to specific genetic factors that can account for the rarity of this as a presentation.

Regarding systemic involvement, two patients fulfilled systemic lupus erythematosus (SLE) criteria. Both presented mild haematological disturbances and arthritis and one of them delivered a child with neonatal LE. Therefore, it seems that this type of lesion on the elbows can develop not exclusively in LE patients with only cutaneous

Figure 1 Erythematous papules and plaques with a slightly scaly surface on the elbows: (a) case 2, (b) case 3, (c) case 4, (d) case 6.

Table 1 Histopathological features

Cases	*Vacuolar change of the basal layer*	*Lymphocytic infiltrate*		*Mucin deposition*	*Interstitial histiocytic infiltrate*	*Changes in the texture and the staining properties of collagen fibres*
		Perivascular	*Periadnexial*			
1	+	+++, CS	–	+	+++	++
2	+	+++, CS		+	++	++
3	+	+++	++	+	+++, and many eosinophils	++
4	+	++	–	++	+	–
5	+	++, CS	–	++	–	–
6	+	++	++	+	++	+
7	+++	++	++	++	+	++, Churg–Strauss granulomas

CS: 'Coat sleeve' distribution

lesions, but also in patients with systemic involvement.

Histologically, in addition to the characteristic findings of CLE, represented by a perivascular lymphocytic infiltrate in the upper and mid-dermis, focal vacuolar change of the basal layer and some dermal mucinosis, there were necrobiotic changes within the collagen fibres similar to those reported in interstitial granulomatous dermatitis or in the interstitial form of granuloma annulare. Interstitial granuloma annulare could be rejected because of the clinical features and the presence of focal vacuolar degeneration of the basal layer,[9] while the lack of palisading necrobiotic granuloma

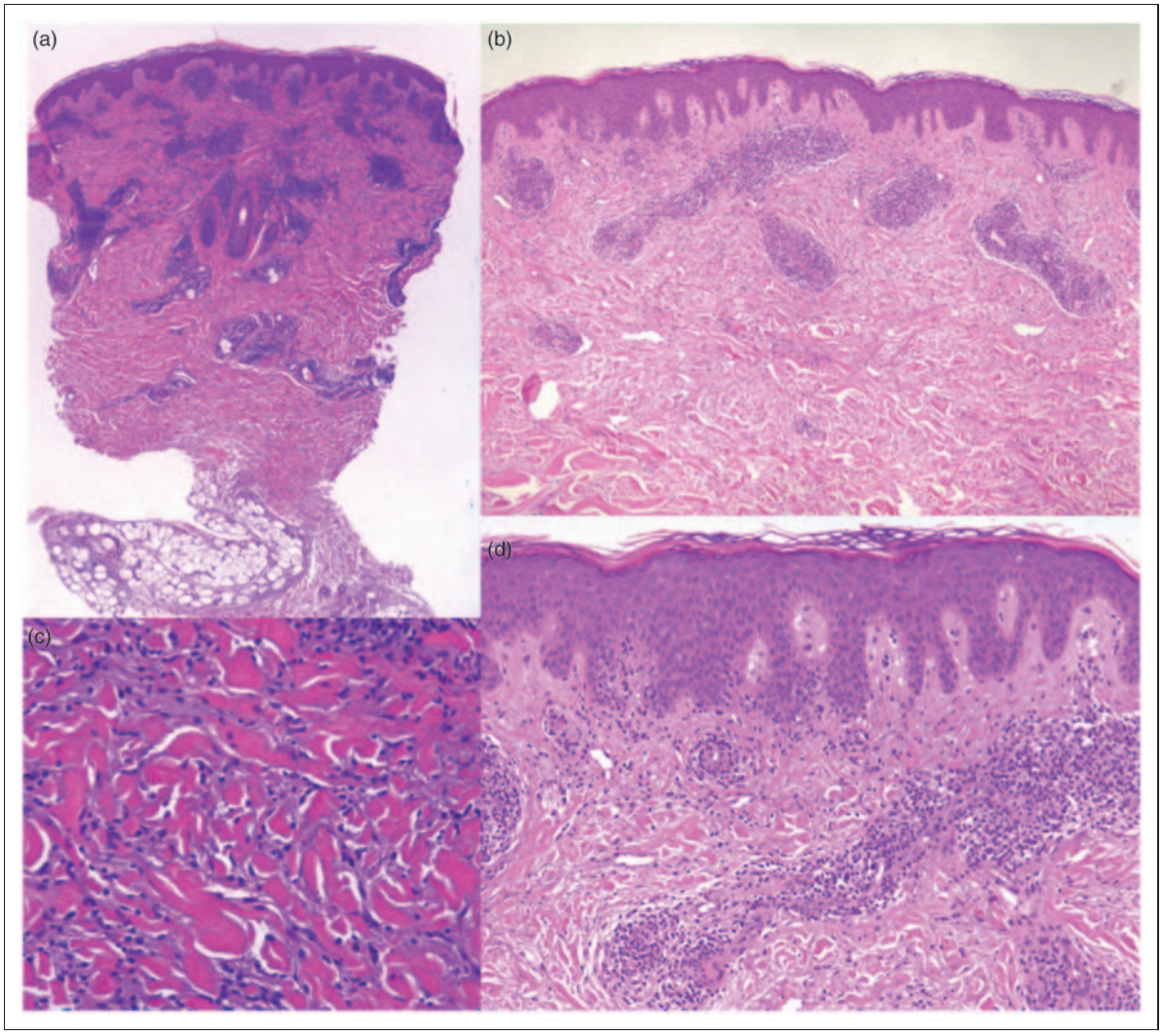

Figure 2 (a) Moderate perivascular lymphocytic infiltrate (HE × 4) (b) Well demarcated perivascular infiltrate with a 'coat-sleeve' distribution (HE × 10) (c) Interstitial histiocytic infiltrate with alterations in the staining properties of collagen fibres (HE × 40) (d) Focal vacuolization changes of the basal layer (HE × 20).

ruled out the diagnosis of classic granuloma annulare. Interstitial granulomatous dermatitis is considered a reaction pattern of necrobiotic and granulomatous inflammation encountered in the setting of systemic diseases, including LE, with varied clinical expression and often located on the elbows.[10–12] In our cases the most remarkable microscopic changes of interstitial granulomatous dermatitis were seen in those with more systemic involvement. However, the presence of focal vacuolar degeneration of basal layer and perivascular lymphocytic infiltrate make this diagnosis unlikely.

In conclusion, we believe that these peculiar lesions located on the elbows should be included in the spectrum of cutaneous manifestations of LE. The histopathology combines changes of both LE and interstitial granulomatous dermatitis. From the therapeutic point of view they show a good response to hydroxychloroquine, with rapid recurrence after discontinuation of treatment.

Conflict of interest

None declared.

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