

Polyarteritis nodosa developing after discoid lupus erythematosus

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ABSTRACT

We describe a patient with discoid lupus erythematosus whose pattern of disease evolved into a systemic vasculitis polyarteritis nodosa.

Introduction

It is very rare for one connective tissue disease to evolve into another. We describe a patient with discoid lupus erythematosus (DLE) which evolved into polyarteritis nodosa (PAN).

Case report

A 36-year-old Caucasian woman developed DLE in 1987, with typical lesions in the ears, pre-auricular areas, face, nose and fingers. Antinuclear antibodies (ANA) were positive (1: 160) and smooth muscle antibodies were weakly positive. Thyroid thyroglobulin and microsome antibodies were strongly positive. She responded well to antimalarial treatment with Mepacrine, alternating with hydroxychloroquine, with a short course of prednisolone (10 mg) initially.

She was taking Mepacrine during the summer as preventative treatment when she developed in 1999, at the age of 47, high blood pressure (180/110), livedo reticularis on the knees and the elbows,

fevers and sweats but no weight loss. On the lower legs she had some nodular lesions and a stocking sensory neuropathy. Her DLE at this time had resolved and her Schirmer's test was normal.

Biopsy of a nodule from the left lower leg showed fibrinoid necrosis and neutrophilic vasculitis of the large and medium arteries (Fig. 1). Antineutrophil cytoplasmic antibodies (ANCA) were weakly positive with a perinuclear pattern (but were negative for proteinase 3 and myeloperoxidase antibodies).

A coeliac axis angiogram was normal as was the 24-hour protein excretion and creatinine clearance. Anti-DNA antibodies, ANA on Hep2 cell, antibodies to extractable nuclear antigens (ENA), complement (C3 and C4), cryoglobulin, hepatitis B serology, lupus anti-coagulant and antiphospholipid antibodies were all negative or normal on at least three separate occasions over the subsequent one year.

A diagnosis of polyarteritis nodosa (PAN) was established. As the patient was clinically getting worse, we started prednisolone 12.5 mg and azathioprine 150 mg daily. This was later modified to six fortnightly low dose intravenous cyclophosphamide pulses followed by

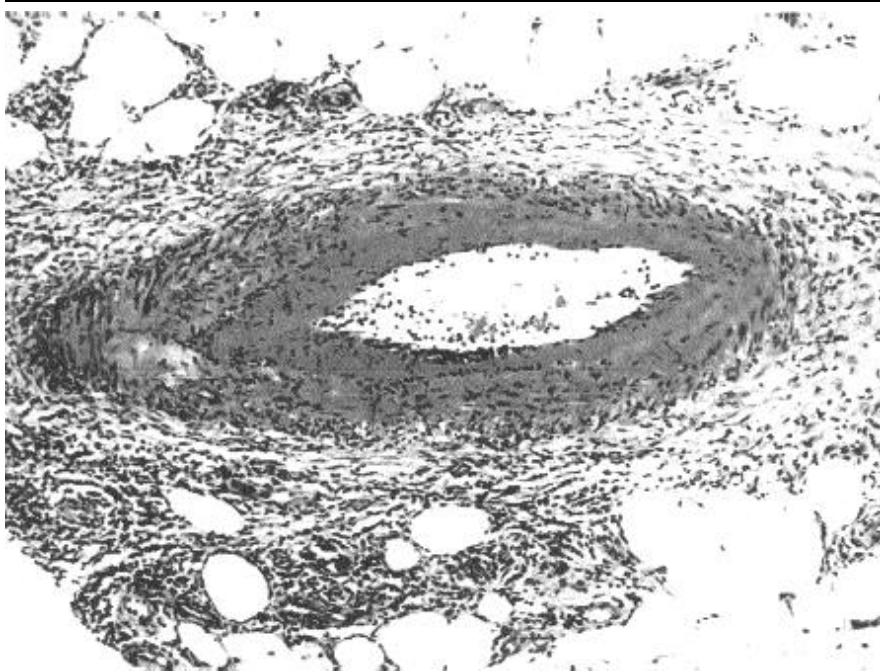


Fig. 1. Skin biopsy of the nodule on the left lower leg. Blood vessel showing neutrophilic inflammatory infiltrate with fibrinoid necrosis (hematoxylin-eosin, 100x).

Table I. Clinical features and American College of Rheumatology criteria.

Livedo reticularis	Leg tenderness	Biopsy	Neuropathy	Testicular pain	Kidney function	Blood pressure	Arteriographic abnormality	Hepatitis B virus	Weight loss
Yes	Yes	+	Yes	No	Normal	High	No	Negative	No

methotrexate, to which she has responded.

Discussion

Previous cases of systemic lupus erythematosus (SLE) evolving into systemic vasculitis in later life have been described (1,2). Only 5% of DLE develop into SLE, however (3). Our patient went from a pure stable cutaneous discoid lesion over ten years to a rapidly progressive systemic vasculitis. She was classified as having PAN based on the skin biopsy and on clinical criteria established by the American College of Rheumatology for the classification of systemic vasculitis (4) (Table I).

This patient again raises interesting questions as to how connective tissue disease should be classified. She had uncomplicated discoid LE responding to long-term anti-malarial therapy, and

showed no signs or symptoms to suggest either SLE or systemic vasculitis until eleven years later. The systemic illness that she developed best fits into a classification of PAN, since she was repeatedly pANCA positive (5 times) and ANA, DNA and Hep2 cell negative (more than 3 times). The unusual evolution of her DLE poses the question as to whether the systemic vasculitis was present subclinically at the beginning of the illness with an unusual clinical pattern or, alternatively, if the patient had both but the approach of menopause resulted in changing hormonal patterns that influenced the disease course towards vasculitis (5, 6). The vasculitides are generally seen in older postmenopausal patients.

To our knowledge this is the first report of a patient with DLE evolving into PAN.

References

- VIVANCOS J, SOLER-CARRILLO J, ARA-DEL REY J, FONT J: Development of polyarteritis nodosa in the course of inactive systemic lupus erythematosus. *Lupus* 1995; 4: 494-5.
- D'CRUZ D, CERVERA R, OLCAY AA, AHMED T, FONT J, HUGHES GR: Systemic lupus erythematosus evolving into systemic vasculitis: A report of five cases. *Br J Rheumatol* 1993; 32: 154-7.
- LAHITA RG: Discoid lupus erythematosus. In LAHITA RG (Ed.): *Systemic Lupus Erythematosus*. San Diego, London, Boston, New York, Sydney, Tokyo, Toronto, Academic Press, 1998: 631.
- HUNDER GG, AREND WP, BLOCH DA et al.: The American College of Rheumatology 1990 criteria for the classification of vasculitis. Introduction. *Arthritis Rheum* 1990; 33:1065-7.
- FONT J, PALLARES L, CERVERA R et al.: Systemic lupus erythematosus in the elderly: clinical and immunological characteristics. *Ann Rheum Dis* 1991; 50: 702-5.
- MADDISON PJ: Systemic lupus erythematosus in the elderly. *J Rheumatol* 1987; 14 (Suppl. 13): 182-7.