Livedo Reticularis on the Lower Limbs in a Patient with Lupus Erythematosus: A Quiz

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A 47-year-old woman presented with reticulate skin lesions on her knees (Fig. 1) at the end of December 2016. She had a long history of systemic lupus erythematosus (SLE) that had started 26 years ago with malar rash and arthritis. Routine blood test results were within the normal range. The patient was positive for antinuclear and double-strand DNA antibodies, antiphospholipid antibodies and lupus anticoagulant. She was given topical corticosteroids and hydroxychloroquine. Three years later, the patient presented with renal involvement and was given systemic corticosteroids and an immunosuppressive drug. Thrombophlebitis and pulmonary embolism occurred in 1995, and was treated with warfarin (Coumadin®) with international normalized ratio (INR) between 2.5 and 3.5. Laboratory blood test results in May 2016 showed a moderate increase in antinuclear antibodies titre (1:200 dilution) and anti-DNA antibodies (43 IU, normal < 14 IU). The dermatologist promptly referred the patient to a tertiary care hospital for complementary investigations. A skin biopsy was taken (Fig. 2) and blood tests were performed.

What is your diagnosis? See next page for answer.
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Diagnosis: Erythema ab igne

The diagnosis was suspected clinically because of some epidermal changes visible on clinical examination, with mild desquamation; the lesions were not infiltrated on palpation; the fishnet livedo pattern was very regular and associated with brown-yellowish pigmentation of the lesions on the left knee (1, 2). Moreover, the patient received warfarin with target INR approximately 2.5, therefore the diagnosis of thrombosis was less likely. Questioning regarding a source of heating of the lower limbs at work or at home revealed that the patient had moved house in May 2016 and enjoyed sitting close to her new fireplace.

Livedo reticularis is permanent livedo that does not disappear with warming of the involved skin. It is termed regular when the annular lesions form complete unbroken circles, as on the left knee of the current patient, and termed livedo racemosa when the annular lesions form incomplete circles, as on her right knee (3). The occurrence of livedo reticularis in a patient with SLE is potentially life-threatening, and clinical and biological signs of venous or arterial thrombosis need to be checked, and tests performed for lupus anticoagulant, antiphospholipid and anti-beta-2 glycoprotein-I antibodies. Approximately 40% of patients with SLE are positive for lupus anticoagulants and approximately 45% of those with antiphospholipid syndrome also have SLE (4, 5). Biological tests are required, including testing for lupus anticoagulant activity. Such tests in the current patient ruled out disease flare as a cause of the skin lesions. A skin biopsy is useful to demonstrate capillary thrombosis or, eventually, vasculitis. In our case, skin biopsy of a recent lesion on the right knee showed dilated dermal capillary vessels (see Fig. 2), with no extravascular blood red cells and no dermal hemosiderin deposits, which was consistent with erythema ab igne (6). The lesion on the left knee was not biopsied. It was older than the lesion on the right knee and showed yellow-brown pigmentation, which was probably due to extravasation of red blood cells and hemosiderin deposits (6).

Erythema ab igne results from prolonged exposure to moderate heat, generally between 40 and 47°C. Some well-known causes include heaters, boilers and radiators. Other causes, linked to modern technology, explain the resurgence of cases and unusual skin locations on the abdomen or thighs: heating pads (7), slimming heaters, heated blankets, heated car seats (8, 9), and, mostly, laptops placed on the knees for hours (10). Treatment is to stop exposure to the heat source, and then the livedo should slowly fade.

REFERENCES