## Remitting symmetrical pitting edema of hands and feet at onset of pediatric systemic lupus erythematosus: a case report

Sirs,

A 12-year-old female presenting with a one-month history of painful edema of the hands followed by edema of the feet and legs was admitted to our clinic. She had a history of occasional thrombocytopenia (83,000/mcl) since eleven years of age, without markers of autoimmune diseases.

Our physical examination revealed pitting edema of her hands and feet and edema of the pretibial region and of the forearm, as well as arthritis and tenosynovitis of her proximal interphalangeal and wrist joints (Fig. Ia and Ib). There were no other abnormal physical signs.

Laboratory tests showed hypocomplementemia, high titer ANA (homogeneous pattern) and anti-DNA antibodies, low leukocyte count (4,000/mcl) and low platelet count (139,000/mcl). No other haematological problems were found (a bone marrow biopsy revealed only poor cellularity). Erythrocite sedimentation rate and C-reactive protein were normal. Anti-cardiolipin antibodies and ENA profile were negative. Renal and liver function tests and the 24-hour urine test were normal. Infectious and hematological diseases were excluded. ECG and echocardiography were normal.

The diagnosis of pitting oedema was made by clinical and ultrasound (US) examination (1). US of her ankles and wrists showed tenosynovitis of flexor and extensor tendons and joint effusion, and US of the shoulders also showed joint effusion. Radiological findings were normal.

A diagnosis of systemic lupus erithematosus (SLE) was made on the basis of four of the eleven ACR criteria (2), namely: arthritis, leukopenia, thrombocytopenia, and anti-DNA antibodies and ANA positivity.

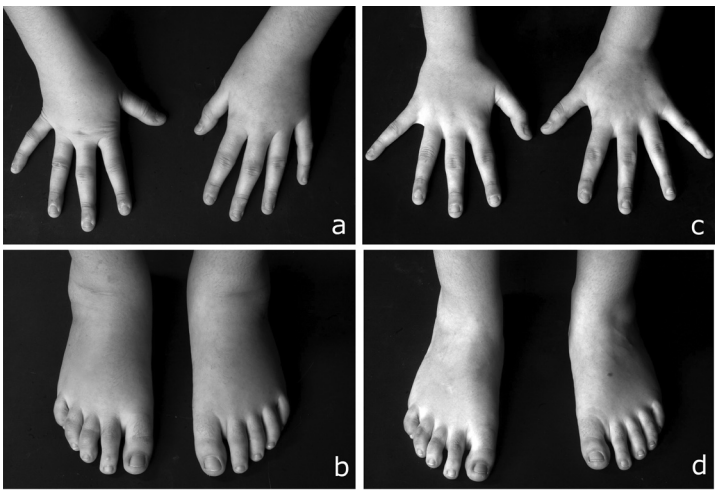
During hospitalization, the patient was treated with NSAIDs, hydroxychloroquine (5 mg/kg/d) and prednisone (initial dose 0.9 mg/kg/d, followed by tapering until withdrawal).

After one month of steroid treatment, pitting edema and joint symptoms disappeared (Fig. 1c and 1d).

Symmetrical pitting edema of the hands and feet is a non-specific finding that may be observed in several rheumatic and neoplastic disorders (3). In 1985, McCarty *et al.* (4) defined a new syndrome characterized by acute onset of symmetrical polysynovitis with pitting edema of the dorsum of the hands and/or feet and negative radiological findings as remitting seronegative symmetrical synovitis with pitting edema (RS3PE). Pitting edema has also been described as presenting symptom of rheumatoid arthritis, Sjögren's syndrome, psoriatic arthropathy and polymyalgia rheumatica (5).

**Fig. 1a, 1b.**
Edema of the dorsum of the hands and feet in a patient with SLE.

**Fig. 1c, 1d.**
The same patient, after 4 weeks of steroid treatment.




The nature of remitting pitting edema is unknown. Two main mechanisms have been suggested. One is delay in lymphatic drainage, probably due either to lymphangitis secondary to the extension of joint inflammation, or to a vasculitis-like syndrome involving the lymphatic vessels. Some authors suggested that arthritis-induced peripheral edema is only found in predisposed patients with pre-existing lymphatic defects (6). The other is a local alteration of permeability of capillaries, probably due to extensive tenosynovitis. Recent studies speculated that some humoral factors (*e.g.* VEGF) could facilitate this clinical manifestation (7).

Only a few cases of remitting pitting edema of the limb in adult SLE have been reported in the literature. In 1998, Pittau *et al.* (8) described a case of a 41-year-old female with SLE associated with steroid-responsive symmetrical distal pitting edema. In 1999, Gunaydin I *et al.* (9) reported on two patients with SLE and lower limb symmetrical pitting edema responsive to systemic steroids. In one case, edema was the first manifestation of the disease. In 2000, Pittau *et al.* (10), suggested that pitting edema in SLE may have an asymmetrical presentation, and described 2 cases.

To our knowledge, this is the first case of remitting symmetrical pitting edema as a presenting manifestation of SLE in a young patient.

Our case indicates that pitting edema can be found in the pediatric population and, when present, should prompt pediatricians to look for an underlying disease, including SLE.

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