Sir, Scleroderma is a rare connective tissue disease characterized by fibrosis and microvascular occlusion. The hallmark of scleroderma is tight, thickened skin brought about by excessive collagen deposition in the dermal and subdermal layers. Generalized scleroderma (systemic sclerosis) is associated with internal organ involvement, autoantibodies and a poor prognosis [1–4]. Patients with localized scleroderma also carry antibodies but their projected lifespan is not foreshortened because sclerotic involvement is largely confined to the skin and musculoskeletal systems.

Three variants of localized scleroderma exist: morphoea, generalized morphoea and linear scleroderma [4–7].

In linear scleroderma, sclerotic areas occur in a linear, band-like distribution, often crossing joint lines [3, 4]. The lesions follow the embryological lines of Blaschko. Local inflammation and fibrosis may affect the dermis, connective tissue, muscle and bone, giving rise to arthralgias, tenosynovitis, constriction, undergrowth of the limb, and nodulosis. There may be associated Raynaud’s phenomenon and carpal tunnel syndrome, but linear scleroderma usually affects the lower limbs of young females [2–4, 6, 7].

Linear scleroderma may cause substantial disabiity where one or more limbs are severely affected and there is the potential for marked cosmetic morbidity. Medical management may include non-steroidal anti-inflammatory drugs, penicillamine aimed at skin softening, and camouflage creams.

Local and systemic corticosteroids, methotrexate and interferon may also play a role [8]. Surgical intervention is rarely indicated and the outcome of surgery must be considered uncertain, giving the patient’s predisposition to excessive local collagen deposition and that taut skin might significantly impair wound healing.

We present the case of a 19-yr-old, left-handed motor mechanic who complained of a 2-yr history of arthralgia involving the fingers of both hands. There was an ill-defined history of Raynaud’s phenomenon, but no symptoms of systemic disease. Examination revealed symmetrical, linear scleroderma arising around the mid-scalpula region and radiating down the posterior aspect of the arms to the dorsum of the hands (Fig. 1). Anti-nuclear antibody was present at a titre of 1:160 IgG. Rheumatoid factor, ESR, anti-ScI70, anticentromere antibody and complement levels were normal or negative. The diagnosis of linear scleroderma was made.

Over the next 7 yr he continued to complain of palmar pain and swelling as well as ‘catching’ of his fingers with straightening, especially in the right hand. This interfered with his ability to grip tools and had a significant impact on his ability to work.

Examination revealed nodularity of the flexor tendons in the right hand proximal to the flexor retinaculum and within the palm, with palpable triggering of all digits as the fingers were actively flexed and extended. Similar findings were found in the left hand to a lesser degree. There were no signs or symptoms of carpal tunnel syndrome. Magnetic resonance imaging (MRI) demonstrated marked thickening around all flexor tendons.

The patient was managed with oral D-penicillamine and local corticosteroid injections, but his principal symptoms persisted unchanged. In view of his continuing intrusive symptoms, tenosynovectomy was considered as a means of reducing symptoms. In the absence of any information in the literature concerning the results of surgery in linear scleroderma, the patient was informed of the possibility of poor wound healing, and of recurrence and excessive fibrotic reaction.

Under a general anaesthetic and with tourniquet control, an approach from the distal forearm to the distal palmar crease was made. The median nerve was protected. There was nodular, fibrotic synovial thickening, most prominent around the profundus tendons. The synovial thickening extended from the musculo-tendinous junction to the entrance of the fibrous flexor sheath distally, confirming the findings on MRI.

A complete flexor tendon synovectomy was performed, combined with release of the A1 pulley of the index and middle finger (Fig. 2). Histological examination of the synovium showed minimal synovitis with marked fibrotic and fibrinoid changes,
probably due to scleroderma. Postoperatively, the patient made an excellent recovery with a full range of pain-free movements in all digits of the right hand. At review 2 yr later, there was no evidence of recurrence. Symptoms in the left hand were not sufficient to warrant surgery.

Carpal tunnel triggering is rare and has not been reported in association with linear scleroderma. There is also no literature on the surgical management of tenosynovitis in this condition. We present a patient with a symmetrical linear scleroderma suffering with bilateral synovitis and triggering of the flexor tendons of the fingers at the entrance to the carpal tunnel. Having been resistant to medical therapy, this was successfully treated with flexor synovectomy and release of selected A1 pulleys.

The surgical treatment of the synovitis in patients suffering with scleroderma causes some concern in view of the risk of postoperative fibrosis resulting in worsening contractures and deformities.

This case demonstrates that good results may be achieved with surgical management and that prolonged medical treatment may be avoided.

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Systemic nocardiosis mimicking an ocular relapse of giant-cell arteritis

We describe a patient treated for temporal arteritis (TA) who experienced recurrent ocular events from different aetiologies, clearly showing that a high degree of suspicion against infection is permanently required in such patients. In addition it seems to be the first reported case of ocular and systemic nocardiosis in that context.

A 83-yr-old man was diagnosed with TA in September 2000, on account of new headaches, tender superficial temporal arteries, polymyalgia rheumatica, a C-reactive protein (CRP) level of 39 mg/l and granulomatous giant-cell arteritis on temporal artery biopsy. Prednisone 40 mg/day (0.5 mg per kg of body weight) led to prompt clinical improvement and CRP normalization. After 3 weeks, however, the left visual acuity suddenly decreased to 6/20 due to choroidal ischaemia attributed to TA (Fig. 1A,B), and progressively returned to 20/20 after four pulses of methylprednisolone 250 mg/day followed by oral prednisone 80 mg/day. At the beginning of May 2001, on prednisone 25 mg/day with the CRP level being 34 mg/l, a painless visual loss of the right eye occurred, leading to a vision of ‘counting fingers at 50 centimetres’. Retinal angiography showed a delayed retinal artery filling time of 35 s and marked localized subretinal oedema without choroidal defect or papillary oedema (Fig. 1C,D). The patient had no signs or symptoms of infection and a negative work-up including chest radiography, echocardiography and microbiological analysis of urine and blood, and we concluded there was TA progression involving the right central retinal artery. A 3-day course of 120 mg/day pulsed methylprednisolone was started on 14 May. During the following days, the CRP level increased rapidly to 240 mg/l and no visual improvement occurred. At the end of May, a pustular lesion was noticed on the patient’s abdomen, and he died on 4 June from septic shock and multi-organ failure. Autopsy showed multiple abscesses varying in size from 1–2 mm to 4 cm in the heart, lungs, pancreas, peritoneum, kidneys, thyroid and in the whole brain (cerebral hemispheres and brain stem, oculomotor nerves and choroid plexus, associated with a purulent meningitis). An accumulation of 1 μm-wide filamentous gram-positive bacilli, Grocott-positive and Ziehl-positive, randomly oriented and