Systemic scleroderma associated with bilateral ulnar nerve entrapment at the elbow

Sir, Peripheral nervous system involvement in systemic sclerosis (SSc) is rare, and is usually limited to carpal tunnel syndrome [1]. Ulnar nerve involvement secondary to compression of Guyon’s canal has been reported in two patients [2, 3]. Herein, we describe for the first time an SSc patient who developed bilateral ulnar nerve sensorimotor compression at the elbow and improved markedly after neurolysis and surgery.

A 36-yr-old woman was referred to our unit in March 1998 for investigation of bilateral sensorimotor ulnar nerve deficit. She had a history of obesity, allergy to tiaprofenic acid, and nervous breakdown. There was no family history of neuropathy. She had two children, worked as a waitress in a restaurant and had a history of smoking one packet of cigarettes/yr for 10 yr. There was no history of alcohol consumption. In 1995, she presented with dysphagia, sclerodactyly, Raynaud’s phenomenon, digital ulcers and telangiectasia; roentgenograms of her hands were normal, anticientromeric antibodies were present at 1/500 and CREST syndrome (calcinosis, Raynaud’s syndrome, oesophageal involvement, sclerodactyly and telangiectasia) was diagnosed. At that time, no other clinical abnormality was noted. In 1996, dyspnoea led to the diagnosis of pulmonary fibrosis, as assessed by high-resolution CT scanning which evidenced interstitial fibrosis on both lung basis and altered function tests (vital capacity 30% diminished, carbon monoxide diffusion capacity 50% diminished). There was no evidence for diffuse scleroderma and renal function was normal. Because pulmonary fibrosis progressed, she received a monthly (0.6 g/m²) intravenous cyclophosphamide pulse from February to August 1996, which stabilized the pulmonary fibrosis, and oral azathioprine was started (2 mg/kg/day).

In August 1997, she complained of paraesthesia involving the fourth and fifth fingers of both hands. At the end of 1997 hypoaesthesia was noted in the same areas. In February 1998, she reported weakness of the left hand. Two months later, clinical examination demonstrated bilateral hypoaesthesia of the fourth and fifth fingers and on the ulnar side of the hand and forearm; on the left side, amyotrophy in the first intersosseous space and bilateral motor deficit was characterized by diminished thumb abduction, reduced pinch strength and diminished extension of the interphalangeal joints. No evidence for involvement of other nerves was found. Deep tendon reflexes were normal. There was no diminution of extension of the interphalangeal joints in the fingers. There was no evidence for elbow synovitis or limitation of the elbow joints.

Biological tests, including sedimentation rate, CRP and fibrinogen, were normal. Anti-SC170 antibodies were negative and anti-centromere antibodies were positive at 1/800. Electromyography confirmed the existence of bilateral ulnar nerve involvement, with left and right diminished motor conduction velocities at 16 and 30 m/s, respectively and abolished and diminished sensory nerve action potentials, indicative of bilateral ulnar nerve entrapment. There was no evidence for median nerve involvement. X-ray films of both elbows and lumbar puncture were normal. Chest CT scan was unchanged. Echocardiography evidenced 10 mm circumferential pericardial effusion. The patient underwent left and right elbow surgery in May and July 1998, respectively. Oedema of the entrapped nerve was noted during surgery and it declined after ulnar arcade resection: the
Ulnar nerve was relocated in front of the epitrochlea. Histological examinations of the epitrochlear–olecranon aponeurotic strip, anterior ulnar arcade and perineural tissue were normal. Complete recovery of muscular strength and sensitivity in the ulnar regions was observed 3 months after surgery.

Ulnar neuropathies at the elbow are caused by direct compression in the retrocondylar groove or entrapment as the nerve passes through the cubital tunnel. Classical causes, such as post-trauma palsy, repetitive chronic trauma and arthritis of the elbow joint, were ruled out in the present case.

For many years, peripheral nervous system (PNS) involvement in SSC has been considered uncommon; it consists of trigeminal neuropathy [4] or peripheral nerve entrapment (e.g. carpal tunnel syndrome) [5, 6] or mononeuritis multiplex [1]. However, subclinical PNS involvement documented by electrophysiological evidence is not rare in SSC; it has been reported in a selected sample of 17 SSC patients without clinical evidence of PNS involvement [7], in 14 of 28 patients [8] and in six of 27 SSC patients without evidence of clinical abnormality [9]. The pathophysiological mechanism(s) at the origin of such involvement is not clearly understood; it may involve both low-grade distal nerve trunk ischaemia and compression due to oedema [5, 10].

Although few cases of symptomatic or asymptomatic ulnar nerve entrapment have been reported [7, 9], only two cases of ulnar nerve paralysis due to nerve compression in Guyon’s canal by calcinosis necessitating surgery have been published [2, 3]. Removal of calcium deposits in both cases and the pisiform in combination with external neurolysis of the ulnar nerve in one case resulted in complete relief of symptoms.

In our patient, SSC was evolutive, as demonstrated by pericarditis and anti-centromere antibodies, and bilateral nerve entrapment was observed in the absence of an aetiological event. Thus, ulnar nerve entrapment at the elbow may be caused by SSC.

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