Acute-onset painful upper limb multifocal demyelinating motor neuropathy

Abstract We report three patients who presented with acute onset of shoulder and upper arm pain followed within a few days by predominantly distal upper limb weakness. Nerve conduction studies showed severe and unequivocal focal motor conduction block in the forearm and/or upper arm along with slowing of motor conduction and prolonged F wave responses. Only very mild changes in sensory nerve conduction were found. One patient made partial clinical improvement after 17 months, and there was a significant improvement in the degree of motor conduction block and the motor conduction velocities. A second patient remained unchanged after 5 months. Idiopathic brachial neuritis (IBN) typically presents acutely with brachialgia and acute or subacute non-progressive weakness. Multifocal motor conduction block in nerves in the arm or forearm has not been described in patients with IBN. Multifocal motor conduction block restricted to the upper limbs has been described in focal chronic inflammatory demyelinating polyneuropathy (CIDP) and in multifocal motor neuropathy with multifocal motor conduction block (MMNCB). However, both these conditions have hitherto usually been described as largely painless chronic progressive disorders with a subacute onset. Our patients, with features overlapping MMNCB/CIDP and IBN, represent an as yet unreported clinical variant.

Key words Idiopathic brachial plexopathy · Multifocal motor neuropathy with conduction block · Chronic inflammatory demyelinating polyneuropathy

Introduction

Idiopathic brachial neuritis (IBN) is a fairly common disorder which presents acutely with severe brachialgia followed by upper limb weakness. The weakness is usually predominantly proximal, affecting mainly the muscles around the shoulder girdle, although more widespread or predominantly distal muscle weakness has also been described [7]. In addition, selective involvement of muscles supplied by the anterior interosseous nerve has long been recognised [15]. Electromyography (EMG) and nerve conduction studies have mainly suggested a process of axonal degeneration at the brachial plexus [4] although recent studies have reported motor conduction block across the brachial plexus [1, 11]. Motor conduction block in nerves distal to the brachial plexus has not been reported. Late recovery occurs.

Multifocal motor neuropathy with multifocal motor conduction block (MMNCB) is a relatively recently recognised rare disorder [10] which usually presents with painless, persistent limb weakness which may be progressive. Focal upper limb involvement is common in MMNCB [13] and has also been reported less commonly in chronic inflammatory demyelinating polyneuropathy [19]. Both conditions are believed to be clinically and pathologically related, although this view is still somewhat controversial [3, 13, 14, 16]. It is generally agreed that an autoimmune ba-
sis is likely, and treatment of patients with MMNCB and CIDP by immunosuppression or immunomodulation has been shown to result in variable improvement in both diseases [14]. The mainstay in the diagnosis of both these conditions are the changes in the nerve conduction studies. Multifocal motor conduction block and slowing of motor nerve conduction are seen in both conditions while additional marked abnormalities of sensory nerve conduction are common in patients with CIDP.

Here we describe three patients whose upper limb weakness began acutely with severe pain, and who had evidence of motor conduction block and slowing of motor conduction in peripheral nerves. Following the generally accepted convention [2], we considered motor conduction block to be present when supramaximal electrical stimulation of a nerve at a proximal site showed a greater than 50% reduction in the compound muscle action potential amplitude compared to stimulation at a distal site.

**Case reports**

**Case 1**

A 30-year-old man developed severe pain in his left upper arm, with radiation to the shoulder and to the forearm and hand. It resolved over a few days at which time he became aware of weakness affecting his left thumb and index finger. He noticed some pins and needles in the left hand. Six years previously he had suffered a painless partial right oculomotor nerve lesion, with full recovery in less than 2 months. Investigations at that time had not revealed a cause. Four years previously he had presented with a scotoma in the visual field of the left eye, caused by a localised sub retinal inflammatory lesion of undiagnosed aetiology treated with a short course of prednisolone, with resolution of his symptoms.

On examination he had very slight weakness of left infraspinatus and deltoid, severe weakness of the left flexor pollicis longus (MRC grade 1) and of flexor digitorum profundus to the left index and middle fingers (MRC grade 3). In addition, there was minimal weakness of left wrist flexion. In both hands he had mild weakness of dorsal interosseous muscles and abductor digitii minimi, and in the right hand of abductor pollicis brevis. All upper limb reflexes were absent, but lower limb reflexes were normal. There were no sensory signs. Cranial nerve and general medical examination were normal.

Nerve conduction studies (Table 1) showed severe motor conduction block in the forearm and upper arm sections of the median and ulnar nerves bilaterally (Fig. 1). Motor conduction velocities were considerably reduced and F wave latencies were markedly prolonged. Sensory nerve conduction studies showed very minor abnormalities of the right median and ulnar nerve sensory action potential (SAP) amplitude. Lower limb motor conduction was normal. EMG examination of the right abductor pollicis brevis showed fibrillations and positive sharp waves and discrete motor units firing rapidly but with a reduced interference pattern. Routine blood tests (haematology and biochemistry) were normal. Anti-ganglioside antibodies were not measured.

Although the patient had significant impairment and disability, his management was complicated by his reluctance to have further investigations. Intravenous immunoglobulin G was advised as the treatment of choice, but the patient declined. He was treated with prednisolone for 4 months building up to a maximum dose of 80 mg on alternate days. Prior to commencing prednisolone the minimal weakness in left deltoid and infraspinatus had resolved spontaneously. After 2 months of prednisolone upper limb reflexes had returned, but profound weakness in muscles supplied by the anterior interosseous nerve remained, as did minor weakness in median and ulnar nerve supplied muscles in the hands. When last reviewed some months after stopping prednisolone, the only weakness was in the left anterior interosseous nerve supplied muscles. Repeat neurophysiological testing (Table 1) showed persisting, but less severe, conduction block in both median and ulnar nerves, improvement in motor conduction velocities and the F wave latencies, and also improvement in median nerve sensory conduction.

**Case 2**

A 47-year-old man presented with acute onset of severe left shoulder and upper arm pain which lasted for 3 or 4 days. During that time he noticed transient paraesthesia of his left little and ring finger. As the pain was subsiding he not

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**Fig. 1** Nerve conduction in the left median nerve of patient 1. Upper tracing: Motor response from the abductor pollicis brevis to stimulation at the wrist; lower tracing: response to stimulation at the elbow. Note the profound conduction block and dispersion of the response to stimulation at the elbow. Conduction velocity in the forearm 23.5 m/s