Motor Branch Biopsy of the Pronator Teres Muscle in a Patient with Painful Forearm Neuropathy

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Key Words

Motor nerve branch biopsy · Multiple mononeuropathy · Muscle biopsy · Nerve biopsy · Painful neuropathy · Pronator teres

Abstract

Histological evaluation of a peripheral nerve is often the final diagnostic work-up for a neuropathy of unknown origin, and a distal sensory nerve is usually biopsied. Here, we report the case of a female patient with painful unilateral neuropathy in the upper arm. According to the histological evaluation of the pronator teres motor branch, vasculitis seemed to be the most probable cause of the condition, and steroid therapy improved the patients' symptoms. A biopsy of the motor branch of the pronator teres muscle nerve may be considered a valuable diagnostic option in selected cases with neuropathy affecting the upper limb, when performed in cooperation with neurologists and orthopedic surgeons.

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Introduction

Histological evaluation of a peripheral nerve is often the final diagnostic work-up for a neuropathy of unknown origin, and is most informative when the nerve is clinically affected [1]. A distal sensory nerve, including the sural, superficial peroneal, or superficial radial nerve, is usually biopsied, and a motor nerve branch is rarely considered [2–4]. Here, we report a case with painful unilateral neuropathy in the upper arm. In this case, a biopsy of a

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pronator teres motor branch provided useful information in consideration of the pathogenesis.

Case Report

A previously healthy 19-year-old Japanese woman noticed a skin rash on her lower limbs. After 4 months, the rash spread to her hands and forearms. In February 2013 (9 months from the onset of the exanthema), she was admitted to our hospital because of sudden paresis and pain in her left upper limb. At the first presentation, she showed livedo reticularis in all four limbs, with a temperature of 36.5°C, a pulse of 65 beats per minute, and a blood pressure of 112/62 mm Hg. Examination showed decreased sensation in the radial side of her left forearm and dorsum of the hand. Manual muscle testing was uncompleted in her left forearm because of severe neuralgia, although it was normal in the left proximal arm and lower limbs.

Laboratory findings were normal for sedimentation rate (8.0 mm/h), leukocyte counts (5,880/mm³), serum C-reactive protein (0.01 mg/dl), serum creatinine (0.49 mg/dl), and urinalysis. Tests for serum antibodies, including antinuclear, anti-DNA, anti-RNP, anti-SSA and anti-SSB, IgG anticardiolipin, and anti-beta-2-glycoprotein I, were negative. Lupus anticoagulant was negative. Serum antineutrophil cytoplasmic antibody was negative in both perinuclear and cytoplasmic staining patterns, and antimyeloperoxidase and proteinase 3 specificity were not detected. Thyroid function was normal, while antithyroid peroxidase and antithyroglobulin antibodies were positive. The results of the serological tests for acute infection with Epstein-Barr virus, herpes simplex virus, varicella-zoster virus, cytomegalovirus, human parvovirus B19, and *Mycoplasma pneumoniae* were all negative. Antibodies to human immunodeficiency virus (HIV) and hepatitis B and C viruses were also negative. Cerebrospinal fluid analysis showed a normal cell count of 1/mm³ (mononuclear cells) with normal total protein concentration (26 mg/dl, normal <40 mg/dl). MRI scans of the brain, whole spine, left upper limb, cervical nerve roots, and brachial plexus and whole-body computed tomography scans showed no abnormal findings. Skin biopsy specimens from the livedo reticularis in her left lower limb showed no specific findings of vasculitis or thrombophlebitis.

In neurophysiological studies, the patient's motor nerve conduction in the median, ulnar, and radial nerves was within the normal range; however, the compound muscle action potential and conduction velocity in the left radial nerve (8.4 mV, normal >7.0 mV; 64.4 m/s, normal >60 m/s, respectively) were decreased in comparison with the right radial nerve (10.6 mV and 91 m/s, respectively). The result of the sensory nerve conduction study was normal. Whilst we were unable to record the motor unit potentials during voluntary contraction of the left forearm muscles because of the patient's severe pain, needle electromyography showed fibrillation potentials in her left extensor carpi radialis muscle. Ultrasound examination of the ulnar, median, and radial nerve in the left forearm showed no abnormalities.

Livedo reticularis and severe neuralgia pointed to vasculitic neuropathy as the most likely pathological condition in this patient. Due to the acute progression of the motor impairment, a single course of intravenous immunoglobulin (400 mg/kg body weight daily for 5 consecutive days) with methylprednisolone pulse therapy (intravenous methylprednisolone 1 g daily for 3 consecutive days) was commenced. Improvement of the patient's neuralgic pain and motor dysfunctions was observed within a few days of commencement of the therapy. However, severe tenderness along the median nerve in her left forearm and

muscle weakness mainly in the left median nerve territory remained. Manual muscle testing showed muscle weakness in the left median nerve territory: flexor digitorum profundus (second finger: 3/5; third finger: 4/5; intact in fourth and fifth finger), flexor digitorum superficialis muscle (second finger: 2/5; third finger: 2/5; fourth finger: 4/5; intact in fifth finger), and pronator teres muscle (2/5). A second MRI of the left upper limb showed new abnormal signal intensities in the median nerve at the forearm and muscles innervated by the radial and median nerve, including flexor pollicis longus, flexor digitorum profundus, flexor digitorum superficialis, extensor carpi ulnaris, and extensor digitorum (fig. 1). These findings were suggestive of the presence of mononeuropathy multiplex involving the left median and radial nerves (predominant in the former).

Despite vasculitic neuropathy being the most likely causative pathological condition in this patient, histological confirmation was required in order to justify the length of her treatment with potentially cytotoxic medications. A superficial radial nerve biopsy was not encouraged because there were no apparent findings of its involvement. We performed a combined biopsy of the left pronator teres muscle and a motor branch of the median nerve to the pronator teres. The procedures were performed in the operating room under conduction anesthesia. We found that her left pronator teres motor nerve had two branches, one proximal branch going to the superficial (humeral) head and the other distal branch going to the deep (ulnar) head of the pronator teres muscle. We biopsied the latter, which was a very thin nerve branch, to reduce potential motor complications of the procedure. There were no complications during or after the biopsy.

In our case, the specimens from a motor branch, that contained only one nerve bundle, showed severe loss of myelinated fibers with a sectional distribution, myelin ovoids with many foamy macrophages, and marked edema in the perineurium and subperineurium (fig. 2A). The specimens from the pronator teres muscle showed typical neurogenic features including small group atrophy with small-angulated muscle fibers (fig. 2B). These findings indicated ischemic nerve injury. As expected, vasculitis seemed to be the most probable cause of the condition, although direct findings (e.g., fibrinoid necrosis of wall vessels) were not seen in these specimens.

As a result of these histological findings, two cycles of a 3-day regimen of high-dose intravenous methylprednisolone (1 g/day) were initiated, followed by oral prednisolone at 0.5 mg/kg/day. The patient's neuralgia remitted and muscle weakness was gradually alleviated. The patient was discharged in April 2013, and returned to full-time housekeeping with no notable neurological deficits. She is currently continuing treatment with low-dose oral prednisolone under active follow-up.

Discussion

The original technique of diagnostic biopsy of the pronator teres and a motor branch of the median nerve was described at academic meetings [5, 6] and reported in a retrospective review of 20 patients as a safe and useful procedure to differentiate between motor neuropathy and motor neuron disease in patients with weakness of the upper limbs [4]. A biopsy of the motor branch of the pronator teres muscle nerve may be considered a valuable diagnostic option in selected cases with neuropathy affecting the upper limb, when performed in cooperation with neurologists and orthopedic surgeons.

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Disclosure Statement

The authors have no conflicts of interest to disclose.

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Fig. 1. Axial short TI inversion recovery (1.5T, TR 7,300 ms, TE 71 ms) MRI of the left forearm. Median nerve (b), flexor digitorum superficialis (c), flexor digitorum profundus (d), pronator teres (e), extensor pollicis longus (f), spinator (g), extensor carpi ulnaris (h), and extensor digitorum (i) showed abnormal high signal intensity. The ulnar nerve (a) showed normal signal intensity.

Fig. 2. Specimens from a motor branch (a single nerve bundle) of the pronator teres muscle (toluidine blue) disclosed sectional loss of myelinated fibers and marked edema in the perineurium and subperineurium (A-a, bar = 100 μm). At higher magnification, severe loss of myelinated fibers with a sectional distribution and foamy macrophages (arrowhead) are visible (A-b, bar = 50 μm). The specimens from the pronator teres muscle showed typical neurogenic features including small group atrophy with small angulated muscle fibers (B, bar = 100 μm). Direct findings of vasculitis (e.g., fibrinoid necrosis of wall vessels) or thrombosis were not observed in these specimens.