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Clinical Short Communication

Ultrasonographic findings of proximal median neuropathy: A case series of suspected distal neuralgic amyotrophy

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ABSTRACT

Spontaneous anterior interosseous nerve (AIN) palsy develops following the resolution of nerve pain, which may be considered as distal neuralgic amyotrophy. NA is assumed to have a complex etiology, but an autoimmune mechanism is likely involved. However, precise assessment of the lesion is challenging. We examined five consecutive patients with suspected spontaneous AIN palsy using ultrasonography. On electromyography, all patients exhibited denervation potentials in the muscles, not only in the AIN territory, but also in the proximal median nerve territory (e.g., the flexor carpi radialis or pronator teres). Ultrasonography of the median nerve demonstrated neural swelling at the proximal side of the medial epicondyle in four patients and an hourglass-like constriction of the nerve fascicle in three patients. Four patients were diagnosed with distal neuralgic amyotrophy; of these, three received intravenous immunoglobulin administration, but only limited beneficial effect was achieved in one patient with early stage disease. One patient showed significant median nerve hypertrophy on ultrasonography and was diagnosed with neurolymphomatosis following the detection of malignant lymphoma during a systemic survey. Our experience demonstrates that ultrasonography for proximal median neuropathy presenting as AIN palsy may be useful for the accurate lesion assessment.

1. Introduction

The anterior interosseous nerve (AIN) branches from the median nerve at the elbow and typically innervates the flexor pollicis longus, flexor digitorum profundus (including the index and sometimes the middle fingers), and pronator quadratus muscles. Weakness of these muscles leads to defective flexion of the distal phalanges of the thumb and index finger when the patient is asked to make the "OK" sign; this is known as the "tear drop sign." Because there are many anatomical variations of the AIN, patients do not always present with typical palsy [1].

Known causes of non-traumatic AIN palsy include space-occupying lesions, compression neuropathy, and pronator teres syndrome; "spontaneous" AIN palsy is considered if medical history, physical examination, and imaging fail to reveal a clear cause. Spontaneous AIN palsy often manifests approximately 1–2 weeks after resolution of idiopathic nerve pain in the affected limb, which is a distal subtype of neuralgic amyotrophy (NA) [2]. NA is assumed to have a complex etiology involving autoimmune, mechanical, and genetic factors. Observations that symptoms are often preceded by an antecedent infection, vaccination, or immunomodulating treatment support the hypothesis that attacks are immunomediated, explaining the term "immune-mediated brachial plexopathy" preferred by some, but no clear pathological mechanism has been identified [3]. Given the many uncertainties surrounding AIN palsy, there is no current consensus on an appropriate therapeutic protocol. Neurolysis has been reported as an effective treatment option, and given the potential involvement of inflammatory processes, immunotherapies such as corticosteroids or intravenous immunoglobulin (IVIG) are often administered [4,5].

In recent years, ultrasonography has been described as useful for diagnosing peripheral neuropathy. To further investigate these possible treatment modalities, we evaluated ultrasonographic findings of the peripheral nerve and clinical courses of patients with AIN palsy [6].

2. Methods

The subjects in this study were referred to our department between 2013 and 2015 for suspected spontaneous AIN palsy. We included five patients experiencing idiopathic focal pain in an upper limb and

Abbreviations: AIN, anterior interosseous nerve; NA, neuralgic amyotrophy; IVIG, intravenous immunoglobulin; CMAPs, compound muscle action potentials; CSA, cross sectional area.

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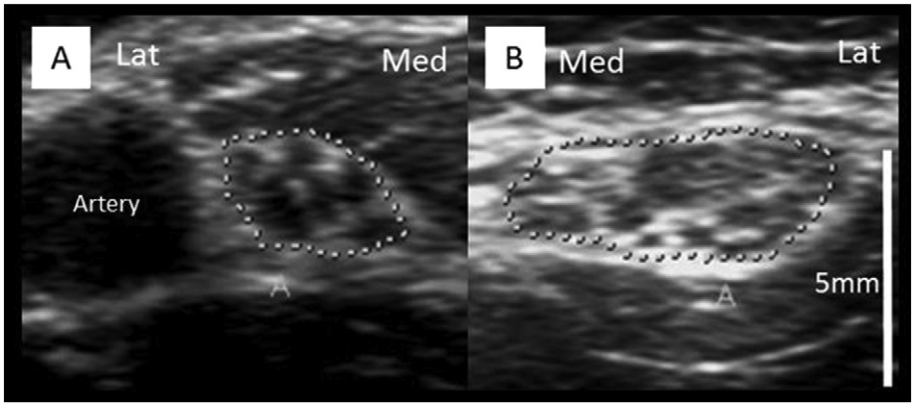

Fig. 1. Ultrasonograms of patient 1 showing side-to-side comparisons of the median nerve (circled). A: The 9-mm^2 non-affected side (right arm) vs. B: cross-sectional area of the 18-mm^2 affected side (left arm), in the region 5 cm proximal to the elbow. Lat: lateral, med: median.

subsequently decreased muscle strength in the fingers of the same limb (predominantly the flexor muscles of the thumb and index finger). The diagnosis was based on medical history, clinical examination, and ancillary investigations, such as magnetic resonance imaging of the cervical spine or lumbar puncture, to exclude other causes. All patients provided informed consent. Ultrasonography was performed using an APLIO™ 500 (Toshiba Medical Systems), a prosound α10 (Hitachi-Aloka Medical), and a 12–18 MHz linear probe. We identified the main trunk of the median nerve (including the branch of the AIN with the proximal part from the elbow) from the forearm to the upper arm region, and searched for the characteristic findings: neural swelling or hourglass-like constriction of the nerve branch. When the probe is lowered to the distal part of the constricted branch, it separates from the trunk of the median nerve in the pronator teres muscle and branches as the anterior interosseous nerve. To study nerve conduction, we applied surface electrodes to the abductor pollicis brevis, pronator quadratus, and flexor hallucis longus muscles. We then applied a supramaximal stimulus to the wrist and elbow regions of the median nerve and compared the bilateral compound muscle action potentials (CMAPs). The study met the guidelines of the Ethics Committee of Kobe University and was performed after informed consent was obtained.

3. Results

3.1. Patient 1

A 30-year-old man presented with pain extending from the left shoulder to the fingertips. Clinical examination revealed distal muscle weakness and atrophy (pronator teres, pronator quadratus, flexor digitorum superficialis, flexor digitorum profundus, flexor carpi radialis, opponens pollicis, and abductor pollicis brevis). He did not report any shoulder region symptoms other than pain. There was no weakness or electromyographic abnormality in the proximal muscles (e.g., anterior serratus and supraspinatus muscles). Ultrasonography revealed neural swelling 5 cm proximal to the elbow (maximum cross sectional area [CSA]: 18 mm^2; healthy side: 9 mm^2), but there were no signs indicating hourglass-like constriction of the nerve fascicle (Fig. 1). Magnetic resonance imaging (MRI) of the left brachial plexus indicated normal signals. IVIG was administered 12 months after onset but had no effect. Subsequent neurolysis was somewhat effective in decreasing the patient's clinical symptoms.

3.2. Patient 2

A 46-year-old man presented with left shoulder pain. Clinical examination revealed distal muscle weakness (pronator quadratus, flexor digitorum profundus, and flexor pollicis longus). He did not report any shoulder region symptoms besides pain. There was no weakness or electromyographic abnormality in the proximal muscles. Ultrasonography revealed neural swelling 3 cm proximal to the elbow (maximum CSA: 20 mm^2; healthy side: 9 mm^2), and we suspected hourglass-like constriction of the nerve fascicle (Fig. 2). MRI of the left brachial plexus indicated normal signals. IVIG was administered 5 months after disease onset, and slight recovery of strength was observed in the flexor pollicis longus muscle 2 weeks later. Further gradual improvement of muscle

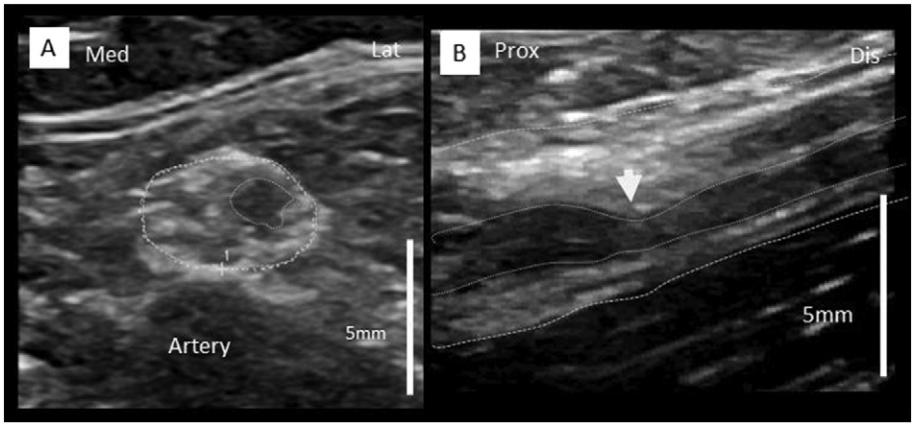

Fig. 2. Ultrasonograms of patient 2. A: Cross-sectional scans of the median nerve in the region 3 cm proximal to the elbow (cross-sectional area: 20 mm^2). B: A longitudinal scan showing constriction of a fascicle of the median nerve at the elbow. Nerves with hourglass-like, incomplete constrictions and enlargements are shown. Dashed line: nerve epineurium, dotted line: nerve fascicle, arrowhead: hourglass-like constrictions, lat: lateral, med: median, dis: distal, prox: proximal.