Clinical Reasoning: A 58-year-old man with distal hand weakness

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Section 1

A 58-year-old man presented to our neurologic clinic with progressive distal weakness in the left hand. Six months prior, he started noticing weakness in the hand. This progressively worsened and led to difficulty in manipulating objects. The patient denied sensory symptoms.

The patient's family history was unremarkable. His medical history included hypertension and a peripheral facial nerve palsy at age 15. At 57 years of age, he had undergone total thyroidectomy for a mixed papillary and follicular carcinoma.

Neurologic examination revealed weakness of the abductor digiti minimi (ADM) muscle and interosseous muscles (Medical Research Council [MRC] grade 3/5) and flexor muscles of the third, fourth, and fifth fingers (MRC 4/5) of the left hand, in the absence of atrophy. Tendon reflexes were normal except for slightly increased left cubito-pronator; the Hoffmann reflex was absent; plantar responses were flexor. The rest of the examination was normal.

The patient had had blood tests performed elsewhere including hematocrit, kidney and liver function, vitamin B_{12}, folic acid, homocysteine, Hb1Ac, inflammatory markers, creatine phosphokinase, thyroid hormones, and autoimmune and onconeural screening, which had normal results.

Questions for consideration:

1. What is the localization?
2. What is the differential diagnosis?

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Section 2

The clinical signs could suggest a C8-T1 radiculopathy limited to the motor roots, due to the absence of sensitive symptoms. A medial cord injury could be considered due to the involvement of ADM, interosseous muscles, and flexor ulnaris carpi, although abductor brevis pollicis was normal and no sensory signs were detected.

Another diagnostic hypothesis is an ulnar neuropathy at the elbow, although considered less likely due to the clinical involvement of the flexor muscle of the third finger (innervated by the median nerve) and the absence of sensory symptoms, usually expected in an ulnar entrapment. Moreover, an ulnar neuropathy at the wrist could be clinically excluded, due to the weakness of the flexor muscles (which are not affected in this condition).

Finally, the increased left cubito-pronator reflex is not consistent with a peripheral nerve disorder, suggesting the possibility of an upper motor neuron disorder.

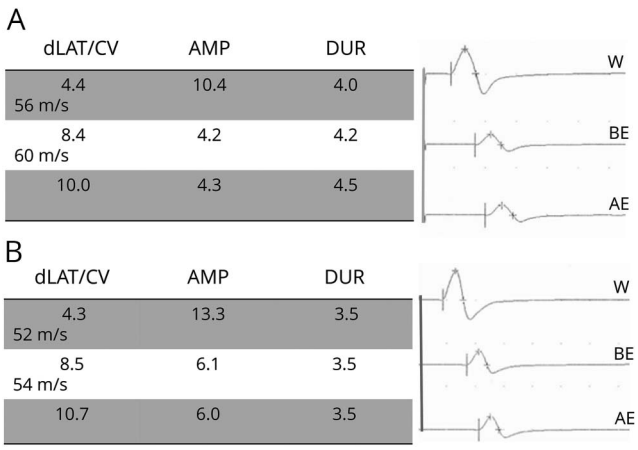
The patient underwent cervical spine MRI and ultrasound of the ulnar nerve at the elbow, which were unremarkable.

Moreover, the patient had also previously undergone MRI of the left wrist elsewhere, which turned out normal.

In order to explore a possible peripheral disorder, a nerve conduction study (NCS) should be performed.

The sensory and motor conduction studies of the median nerve bilaterally showed normal findings as well as the motor conduction studies of the ulnar nerve recording from ADM. Conversely, the study of the ulnar nerve recording from the first dorsal interosseous muscle (FDI) bilaterally disclosed a partial conduction block (CB) in the forearm (figure 1).

Figure 1 Nerve conduction study



Bilateral ulnar partial conduction block in the forearm recording from the first dorsal interosseous muscle. (A) Left ulnar nerve. (B) Right ulnar nerve. AE = above elbow; AMP = amplitude; BE = below elbow; CV = conduction velocity; dLAT = distal latency; DUR = duration; W = wrist.

Questions for consideration:

1. How would you interpret the electrodiagnostic findings?
2. Are neurophysiologic studies sufficient to make a diagnosis?

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Section 3

Focal CB is identified by showing a compound muscle action potential (CMAP) drop across the site of block: the CMAP after stimulation proximal to the site of block is smaller than the CMAP after stimulation distal to the site of block, with approximately the same duration.

When the CB does not occur across a compression site (the wrist or the elbow), it could suggest the presence of an acquired demyelinating peripheral neuropathy, such as motor neuropathy with conduction block, also called multifocal motor neuropathy (MMN).

This entity is a rare neuropathy usually presenting with asymmetrical weakness of distal upper limbs, fitting a specific nerve territory.

According to the European Federation of Neurologic Societies/Peripheral Nerve Society guideline,[1] our patient could be diagnosed with possible MMN, due to the clinical involvement of 1 nerve and the presence of motor conduction block in 2 nerves on electrophysiologic testing.

Therefore, this diagnostic hypothesis was considered.

However, the electrodiagnostic findings did not justify the clinical signs and symptoms, since the CBs were bilateral despite unilateral clinical disturbances and neurologic signs (weakness of left hand, normal strength in right hand).

Question for consideration:

1. Is there another explanation for these electrophysiologic findings?

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Section 4

Another explanation of these electrophysiologic findings is a Martin-Gruber anastomosis (MGA), a known neuroanatomic variant.

Indeed, a short segment study of bilateral ulnar motor nerve recording from FDI confirmed a drop in amplitude between the wrist and below-elbow sites; the stimulation of the median nerve (at the same level of the drop in the ulnar CMAP amplitude) recording from FDI evoked a CMAP approximately equal to the drop in amplitude in ulnar studies (figure 2A). These findings were bilaterally consistent with the presence of a crossover from median nerve or its branches to the motor fibers of ulnar nerve going to the first interosseous muscle, known as MGA.

The ultrasound of the median and ulnar nerves in the forearm confirmed the presence of the anastomosis: a nervous branch arose from the median nerve just below the pronator teres muscle and crossed over to join the ulnar nerve in the middle third of the forearm.

MGA is the most common nerve anatomic variant found in the upper extremities and is seen in around 11%–39% of normal individuals.[2] It involves only motor fibers, sparing the sensory fibers.

The 3 most common sites of MGA origin are the branches of the median nerve supplying the superficial forearm flexor muscles, the anterior interosseous nerve, or the main median nerve.

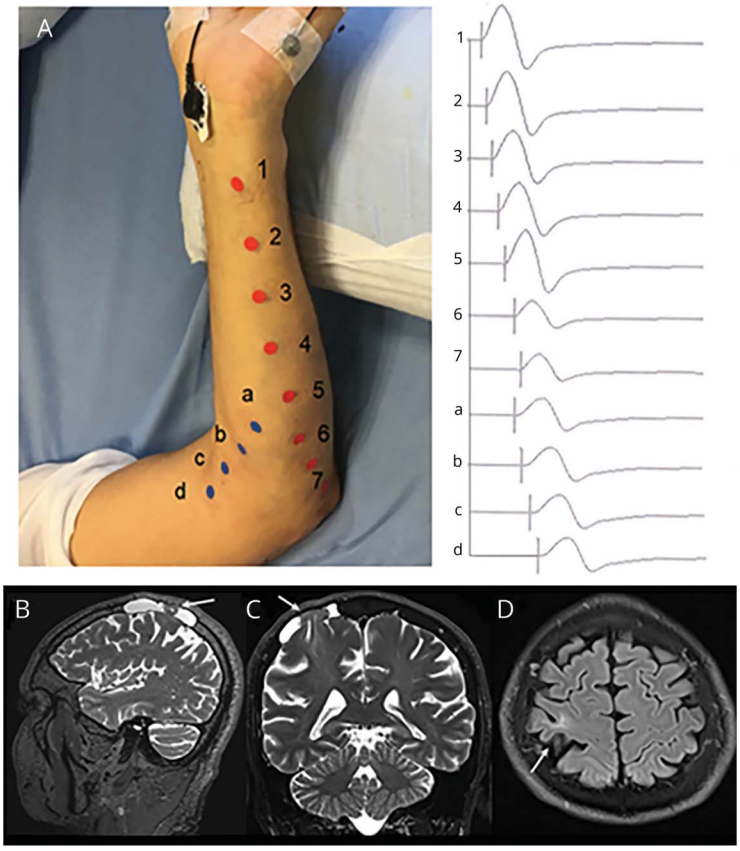
In electrophysiologic studies, the type of MGA is classified into 3 groups according to the muscles innervated by the crossover fibers: in type I, the hypothenar muscles (mainly, ADM); in type II, the FDI; and in type III, the thenar muscles. Type II is the most common form, even if not often recognized during routine electrodiagnostic studies, the ADM being the muscle most often recorded for ulnar motor studies.

In our patient, the discovery of this anatomical variant should be considered only a normal finding, which did not explain the clinical presentation of hand weakness.

Question for consideration:

1. How would you continue the diagnostic work-up?

Figure 2 Martin-Gruber anastomosis



(A) Short segmental study along the course of the left ulnar nerve at 3-cm interval with first dorsal interosseous muscle (FDI) recording shows a conduction block at point 6. The stimulation of the median nerve (at the point d) recording from FDI evokes a compound muscle action potential (CMAP) amplitude equal to the difference between CMAP 6 and 5, confirming a Martin-Gruber anastomosis (a–c). (B–D) Cerebral 3T MRI. Meningoencephalocele of the right rolandic region. (B) Sagittal T2-weighted image and (C) coronal T2-weighted image: defect of the inner table of the parietal bone containing CSF and a mass of herniated brain tissue including rolandic gyrus (arrows). (D) T2 fluid-attenuated inversion recovery sequence: hyperintensity of rolandic gyrus (arrow).

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Section 5

At this point, a cerebral CT to exclude central lesions was performed.

CT showed a lytic lesion of the parietal bone on the right side characterized by an intact but remolded outer table and an interrupted inner table with a large defect.

Thus the patient underwent a cerebral 3T MRI that confirmed the presence of a right parasagittal defect of the inner table of the parietal bone containing CSF and a mass of herniated brain tissue including rolandic gyrus, which was hypointense on T1-weighted images and hyperintense on T2-weighted images, as gliosis (figure 2, B–D).

A diagnosis of symptomatic meningoencephalocele of the right rolandic region was finally achieved.

After a few months, the patient underwent neurosurgical treatment consisting of craniotomy, dural opening, and reduction of the herniated brain tissue, followed by duraplasty and replacement of the outer table of the skull. At the 2-month follow-up, the weakness of the hand was improved.

Discussion

This case highlights the importance of the clinical context when interpreting electrodiagnostic studies. Our patient presented with left hand weakness, carrying a symptomatic meningoencephalocele of the right rolandic region and a bilateral MGA, which could mislead the diagnosis.

The intradiploic meningoencephalocele is an extremely rare condition caused by a defect of the inner table of the calvarium and subsequent herniation of the meninges and cerebral parenchyma into the intradiploic space.[3] Since 1976, very few cases of intradiploic encephaloceles have been reported in the literature. Most of them occurred within the parietal bone and were related to a trauma or neurosurgery; one case within the frontal bone was caused by an accidental tear of the dura during a craniosynostosis repair.[4–8]

More recently, a spontaneous frontal intradiploic meningoencephalocele has been reported in a 60-year-old woman with a history of multiple traumatic head injuries during her childhood.[9]

The authors hypothesized that "spontaneous" meningoencephaloceles may be caused by a traumatic event not recalled or believed not relevant by the patient, even in the distant past.

When we asked the patient about any head injury, he reported a head trauma a few months before the onset of his symptoms.

The diagnostic delay in this case could be due to the presence of a drop in the ulnar CMAP amplitude in the forearm, mimicking an ulnar conduction block consistent with an acquired demyelinating peripheral neuropathy.

Moreover, the clinical symptoms of the patient could be confusing, mimicking an ulnar nerve territory, even if unilateral instead of bilateral neurophysiologic abnormal findings.

Therefore, a deep clinical reasoning allowed us to consider the diagnosis of MMN unlikely, and led us to perform another focused NCS, achieving the diagnosis of MGA.

MGA is an under-recognized entity in routine electrodiagnostic studies, since it could mimic an ulnar neuropathy or a peripheral neuropathy with CB.

Neurophysiologists and clinicians should be aware of screening for the presence of MGA in case of apparent CB across the forearm, especially when discrepancy between neurophysiologic and clinical findings exists.

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Name	Location	Role	Contribution
Veria Vacchiano, MD	Department of Biomedical and Neuromotor Sciences, University of Bologna, Italy	Author	Drafted/revised the manuscript and created the figures, involved in the clinical care of the patient
Vitantonio Di Stasi, MD	IRCCS Istituto delle Scienze Neurologiche di Bologna, Italy	Author	Revised the manuscript, conducted/interpreted the electrodiagnostic studies
Vincenzo Donadio, MD	IRCCS Istituto delle Scienze Neurologiche di Bologna, Italy	Author	Revised the manuscript, conducted/interpreted the electrodiagnostic studies, involved in clinical care of the patient

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Name	Location	Role	Contribution
Carmelo Sturiale, MD	IRCCS Istituto delle Scienze Neurologiche di Bologna, Italy	Author	Involved in surgical treatment of the patient, revised the manuscript
Rocco Liguori, MD	Department of Biomedical and Neuromotor Sciences, University of Bologna, Italy	Author	Conceptualized the study and revised the manuscript, the electrodiagnostic studies and the figures; involved in clinical care of the patient

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