CASE REPORT

Piso-hamate hiatus syndrome in a patient with Riche-Cannieu anastomosis

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SUMMARY

Various nerve anastomoses and anatomic variants in the nervous system have been described. It is important to be familiar with these anastomoses because they can mimic several clinical conditions, possibly leading to misdiagnosis. We report the case of a patient who experienced progressive loss of strength in his left hand without sensory complaints. On neurological examination atrophy was seen in the thenar and first dorsal interosseous muscles of the left hand. Serial electrophysiological studies ruled out motor neuron disease. Electromyography and nerve conduction studies revealed a Riche-Cannieu anastomosis in the left hand. In combination with piso-hamate hiatus syndrome, this anastomosis explained the clinical condition of the patient. It is important to be aware of this anastomosis and this syndrome because it can mimic motor neuron disease.

BACKGROUND

Motor weakness of the hand without sensory complaint has a wide differential diagnosis including motor neuron disease, multifocal motor neuropathy, myelopathy (syringomyelia) and deep ulnar branch neuropathy. Most of these diagnoses have a clear history with typical findings on physical examination, electrodiagnostic testing (EDT) and needle electromyography (EMG). However, in the case of an anomalous innervation, such as Riche-Cannieu anastomosis, electrodiagnostic findings can be confusing, easily leading to the wrong diagnosis.

We report the clinical and EDT features of a young Dutch man with piso-hamate hiatus syndrome in combination with an underlying Riche-Cannieu anastomosis.

CASE PRESENTATION

A 24-year-old, healthy, Caucasian Dutch man was admitted to our hospital because of progressive loss of strength in his left hand. He had no history of neurological or other disease. He was left-handed and had experienced 9 months of difficulties in his work as a mason. The complaints were slowly progressive without an underlying cause. He did not mention any sensory complaints in the left hand, weakness of the left forearm muscles or problems in the upper right extremity or lower extremities.

On neurological examination, atrophy of the thenar muscle and first dorsal interosseous muscle (FDI) in the left hand was noticed (figure 1). The patient also had much callus formation on his palms. In the left hand, there was weakness of the abductor pollicis brevis muscle (APB; Medical

Research Council Scale for Muscle Strength (MRC) 3) and the FDI (MRC 3). No other abnormalities were found at neurological examination. Sensory tests and reflexes were normal, and there were no bulbar signs or fasciculations.

INVESTIGATIONS

Electrophysiological findings

The EDT findings are summarised in table 1. All measurement were carried out with a Viking Select EMG NCS system, V.12 (CareFusion). Sensory nerve conduction studies were normal at initial EDT. Motor conduction studies of the ulnar nerve, recorded from the abductor digiti minimi muscle (ADM) and FDI, fell within the normal range on both sides. However, the compound muscle action potential (CMAP) amplitude of the FDI was obviously lower on the left side (table 1). Motor conduction study of the median nerve, recorded from the APB, demonstrated a normal CMAP amplitude and a normal distal motor latency (DML) on the left side. Late responses (F waves) were normal. EMG demonstrated many fibrillation potentials and positive sharp waves in the FDI on the left with a single voluntary motor unit potential (MUP) pattern on maximal exertion. No abnormalities were seen in the FDI on the right or in the ADM on both sides. Needle examination of the APB on the left side demonstrated increased MUPs with a poor recruitment pattern on exertion.

At a second EDT (4 weeks later), the sensory nerve action potentials (SNAPs) of the ulnar nerve and median nerve were again within normal ranges on both sides and there were no signs of abnormal sensory innervation. Motor conduction study of the ulnar nerve, recorded from the ADM with stimulation at the wrist and the elbow, revealed no abnormalities on either side. Motor conduction study of the ulnar nerve, recorded from the FDI with stimulation at the wrist and the elbow, demonstrated a normal DML and a normal CMAP on both sides. Stimulation of the median nerve at the wrist and the elbow elicited a CMAP of 1.2 mV recorded from the FDI on the left side; no abnormalities were seen on the right side. Recording from the ADM, stimulation of the median nerve on both sides did not elicit CMAP (figure 2). Stimulation of the ulnar nerve at the wrist and the elbow on the left side induced a CMAP of 2.6 mV recorded from the APB; no abnormalities were seen on the right side (figure 3).

Additional studies

MRI of the cervical spine revealed no abnormalities. Ultrasonography of the arm, wrist and hand

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Figure 1 (A) Atrophy of the first dorsal interosseous muscle (FDI) of the left hand. (B) Detail showing atrophy of the FDI of the left hand. (C) Atrophy of the abductor pollicis brevis muscle (APB) of the left hand. Note the extensive callus formation on both palms. (D) Detail showing atrophy of the APB of the left hand.

demonstrated no abnormalities on either side. The cross-sectional area and longitudinal diameter of the ulnar and median nerves were within normal ranges, without increased intraneural vascularisation.^{1 2} An anastomosis could not be demonstrated. MRI of the left hand and wrist demonstrated that a branch of the ulnar nerve which normally runs on the

ulnar side of the palm, was localised on the radial side of the palm. No further details could be obtained because of the small size of this branch. No other abnormalities were seen. EDT and EMG findings suggested that the described aberrant branch of the ulnar nerve seen on MRI was a Riche-Cannieu anastomosis.

TREATMENT

It was likely that the patient's complaints had arisen as a result of his work as a mason due to repeated compression when carrying heavy bricks and the callus formation on his palms. A brace was prescribed and rehabilitation therapy was started.

OUTCOME AND FOLLOW-UP

The patient had fewer complaints eight months later and there was recovery of hand function.

DISCUSSION

Weakness and atrophy of the intrinsic muscles of the hand, without sensory complaints, often suggest motor neuron disease. Therefore, it is very important to distinguish motor neuron disease from less severe conditions such as multifocal motor neuropathy, myelopathy (syringomyelia) and deep ulnar branch neuropathy. EDT is essential in the diagnostic process, but abnormal innervation and anatomical variations can cause difficulty in interpretation of the results.

We describe a patient with an anatomic variation of the innervation of hand muscles (Riche-Cannieu syndrome), who also had piso-hamate hiatus syndrome.

Table 1 Nerve conduction study findings

Stimulate (record)	Amplitude (mV or μ V)			Conduction velocity (m/s)			Distal latency (ms)		
	L	R	NL	L	R	NL	L	R	NL
Inar, motor (FDI)	7.2	17.4	≥ 7	56.9	58.0	≥ 49	3.5	3.7	≤ 4.5
Inar, motor (ADM)	16.7	14.4	≥ 6	56.9	58.0	≥ 49	2.9	2.9	≤ 3.3
Median, motor (APB)	5.7	8.4	≥ 4	54.6	56.6	≥ 49	4.1	4.0	≤ 4.4
Ulnar, sensory (fifth)	17.2	—	≥ 17	—	—	≥ 50	2.7	—	≤ 3.1
Median, sensory (index)	21.7	—	≥ 20	—	—	≥ 50	3.0	—	≤ 3.5
Ulnar, sensory (fourth)	14.1	12.5	≥ 10	—	—	≥ 50	2.9	2.7	—
Median, sensory (fourth)	19.5	14.9	≥ 10	—	—	≥ 50	3.2	3.1	—
Digit 4 difference	—	—	—	—	—	—	0.3	0.4	≤ 0.4
Ulnar, motor (APB)	2.6	—	—	—	—	—	3.6	—	—
Median, motor (FDI)	1.2	—	—	—	—	—	3.8	—	—
Median, motor (ADM)	NR	—	—	—	—	—	NR	—	—
Ulnar, sensory (index)	NR	—	—	—	—	—	NR	—	—
Median, sensory (fifth)	NR	—	—	—	—	—	NR	—	—

ADM, abductor digiti minimi muscle; APB, abductor pollicis brevis muscle; FDI, first dorsal interosseous muscle; L, left; NL, normal; NR, no response; R, right.

Figure 2 Motor nerve conduction study, recorded from the first dorsal interosseus muscle (FDI; A1–A4) and from the abductor digiti minimi muscle (ADM; B1–B4) on the left side. Stimulation of the ulnar nerve generates a compound muscle action potential (CMAP), recording from the FDI (A1–A2) and from the ADM (B1–B2). Stimulation of the median nerve generates a CMAP, recording from the FDI (A3–A4). Stimulation of the median nerve did not generate a CMAP, recording from the ADM (B3–B4).

Atrophy of the FDI without atrophy of the ADM suggests ulnar nerve pathology distal to the bifurcation of the ulnar nerve in the palm. The combination of atrophy of the FDI without atrophy of the ADM and without sensory complaints has been described in patients with piso-hamate hiatus syndrome.^{3 4} This syndrome is a variation of Guyon's canal syndrome. Because the ulnar nerve divides into sensory and motor branches within Guyon's canal, varying clinical conditions can be seen in compression at this level. Five different clinical syndromes have been described.⁴ Ulnar nerve lesions distal to the bifurcation of the superficial sensory branch result in purely motor deficits. Superficial to the deep motor branch of the ulnar nerve lies a ligament band which runs from the pisiform bone to the hook of the hamate.⁴ At this level the ulnar nerve can be compressed by the piso-hamate ligament or the superficial volar carpal ligament of Guyon's canal itself (figure 4).^{4 5}

After exiting Guyon's canal, the deep motor branch of the ulnar nerve gives off a branch to the hypothenar muscles. The motor branch supplying the ADM most commonly arises from the main motor branch just proximal to the hiatus.⁴ Compression or a lesion of the ulnar nerve distal to this bifurcation will involve the flexor pollicis brevis muscle, adductor pollicis muscle and the interossei muscles, but spares the hypothenar muscles.^{4 6} Repeated trauma to the palm (often occupational, for example when using a hammer or gripping strongly) is a frequent cause.^{4 5} Other causes are accessory muscles, soft tissue

Figure 3 Motor nerve conduction study recorded from the abductor pollicis brevis muscle in the left hand. Stimulation of the median nerve generates a compound muscle action potential (CMAP; C1–C2). Stimulation of the ulnar nerve also generates a CMAP (C3–C4).

tumours, haematoma, compression by a synovial cyst, callus formation surrounding the hook of the hamate bone after a fracture and arteriovenous malformation.⁶

However, this syndrome could not explain the atrophy of the APB in the left hand of our patient. The only possible explanation was the coexistence of a connecting nerve branch between the ulnar nerve and the median nerve in the palm, beyond the bifurcation of the ulnar nerve. In our patient, EDT confirmed the presence of this communication which is known as a Riche-Cannieu anastomosis (figure 4). While many clinical neurophysiologists are familiar with a Martin-Gruber anastomosis, Riche-Cannieu syndrome is often unrecognised and not well understood. In Riche-Cannieu syndrome, the anomaly is thought to involve a crossover from the deep branch (profundus ramus) of the ulnar nerve to the recurrent branch of the median nerve in the palm of the hand.^{7 8} Cannieu described the anastomosis in 1896 in three out of 20 hands he dissected, and Riche described the anastomosis in 1897 in three out of 12 hands he dissected.^{9 10} Riche-Cannieu anastomosis is considered to be a random developmental variation, although the exact cause is not known. A familial predisposition with autosomal dominant inheritance has been postulated.¹¹ An incidence of up to 83% has been described in a reference population, in which there is a functional connection between the ulnar and median nerves in the hand.¹² Riche-Cannieu syndrome can have several clinical presentations, resulting in a hand that: (a) is completely supplied by the ulnar nerve (the all ulnar hand); (b) has a proportion of (typically) median nerve innervated muscles supplied by the ulnar nerve; and (c) has motor innervation solely supplied by the ulnar nerve.^{7 11 13–16} Because of the existence of a communication between the ulnar nerve and the median nerve in the hand, double motor innervation of specific muscles is

Figure 4 (A) Schematic illustration of the pisohamate ligament and Riche-Cannieu anastomosis in the left hand of our patient. (B) The red oval areas represent the atrophic muscles (the upper indicates the abductor pollicis brevis muscle, and the lower the first dorsal interosseous muscle). The blue oval area indicates the unaffected abductor digiti minimi muscle. The black star indicates the site of the lesion. H, hook of hamate; MN, median nerve; P, pisiforme; PL, pisohamate ligament; RCA, Riche-Cannieu anastomosis; S, tuberosity of scaphoid; UN, ulnar nerve.

possible.^{3 8 11 16–20} As a result, for example, it is possible to have preservation of thenar function (APB and opponens pollicis muscle function) in patients with median nerve transection in the forearm or in patients with advanced carpal tunnel syndrome.^{17 21} However, in the case of Riche-Cannieu anastomosis, damage to the ulnar nerve can lead to loss of all hand intrinsic muscle function. Eight types of Riche-Cannieu anastomosis have been described involving several muscles.^{8 18 19} The main issue is to demonstrate the (partial) ulnar innervation of the thenar eminence. Because this communication takes an ansiform course through the thenar eminence, the term thenar ansa has also been proposed.^{8 18 20} As can be seen in figure 3, we obtained a CMAP recording over the APB by ulnar nerve stimulation. It is important to be aware that such a result can also be observed from CMAP recording from the adductor pollicis muscle by ulnar nerve stimulation, which is a normal finding. We believe this is less likely given the localisation of the atrophy and the site of recording. In our patient we also obtained a CMAP recorded over the FDI by median nerve stimulation. This is not a typical finding in a Riche-Cannieu anastomosis, and it could be a manifestation of a volume-conducted response from the thenar eminence because of the atrophic FDI. However, we have been able to reproduce this measurement. In our opinion it should be possible to generate a CMAP recorded over the FDI by stimulation of the median nerve because of the crossing fibres of the Riche-Cannieu anastomosis.

MRI of the hand could not visualise an anastomosis, thus corroborating the low diagnostic value of MRI for demonstrating a Riche-Cannieu anastomosis.¹¹ To our knowledge there has been only one report of a similar case in the literature, which described a patient with thenar and FDI wasting in combination with sparing of the hypothenar muscles and sensory function, because of an underlying Riche-Cannieu anastomosis. A fourth metacarpal fracture was the likely cause in this case.³ Knowledge of this condition and resulting anomalous innervation patterns remains critical for correct diagnosis and management.

Learning points

- Consider anomalous innervation patterns and/or nerve anastomosis in atypical muscle wasting.
- Riche-Cannieu anastomosis in combination with pisohamate hiatus syndrome can mimic motor neuron disease.

Competing interests None.

Patient consent Obtained.

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