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L. Padua • C. Pazzaglia • A. Insola • I. Aprile • P. Caliandro • M. Rampoldi • C. Bertolini • P. Tonali

Schwannoma of the median nerve (even outside the wrist) may mimic carpal tunnel syndrome

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Abstract Over the last 3 years we have observed 5 cases of median nerve schwannoma that clinically simulated carpal tunnel syndrome (CTS). We describe the atypical clinical-neurophysiological picture indicating to perform ultrasonography (US). We retrospectively re-evaluated 5 cases of schwannoma that clinically simulated CTS. Five consecutive patients were referred to the neurophysiopathology laboratory. All patients complained of symptoms and had a neurophysiological examination that might have indicated CTS. Nevertheless we performed US because of some incongruous aspects. In cases of atypical abnormalities at neurophysiological and clinical examination, or dissociation between neurophysiological and clinical findings, physicians should consider the presence of a median nerve tumour. Here, US evaluation is very useful as supporting diagnostic methodology to assess the anatomopathological condition of the nerve lesion and must not be limited to the wrist.

Key words Schwannoma • Carpal tunnel syndrome • Ultrasonography

L. Padua (✉) • C. Pazzaglia • I. Aprile • P. Caliandro • P. Tonali
Department of Neuroscience
Institute of Neurology
Università Cattolica
Largo F. Vito 1, I-00168, Rome, Italy
e-mail: lpadua@rm.unicatt.it

L. Padua • I. Aprile
Fondazione Don C. Gnocchi
Centro Santa Maria della Pace, Rome, Italy

A. Insola
U.O. Neurofisiopatologia
C.T.O. Roma, Rome, Italy

M. Rampoldi
U.O. Chirurgia della mano C.T.O. Roma
Rome, Italy

C. Bertolini
Department of Rehabilitation
Università Cattolica, Rome, Italy

Introduction

The presence of a schwannoma in upper limb nerves may determine neurological symptoms in the hands [1–3]. Carpal tunnel syndrome (CTS) is caused by the entrapment of the median nerve at wrist; it is a common disease with a 10% lifetime risk, according to the American Academy of Neurology [4]. A recent study on the Swedish general population demonstrated that the prevalence is 3.8% [5].

The CTS clinical picture is typically characterised by nocturnal algo-paraesthesia at the hand; in more severe cases hypoaesthesia in the hand region innervated by median nerve and hypotrophy of the thenar eminence.

Neurophysiological evaluation is considered very useful for the diagnosis of CTS, as it shows the focal reduction of the conduction velocity of the median nerve at wrist, because median nerve abnormalities associated with CTS are focal and, at least in the initial phases, restricted to the proximal segment that lies within the carpal tunnel [6, 7].

Recently, detail resolution has increasingly been used to aid the diagnosis of compression and entrapment neuropathies. Advances in ultrasound technology have made it possible to gain higher spatial resolution and even depict nerves with excellent visual quality [8]. The importance of the ultrasonography (US) technique in CTS (and more generally in focal nerve involvement) is that we can see, from an anatomic point of view, the nerve along its course.

During the last 3 years we have observed 5 cases of schwannoma of the median nerve that clinically simulated CTS. Diagnosis was made by using neurophysiological evaluation and US.

Materials and methods

The 5 cases reported here were enrolled in two neurophysiological outpatient services specialising in upper limb nerve lesions; over the time period (3 years) in which we observed these 5 cases, we evaluated 830 patients with diagnosis of CTS. Table 1 shows the time of onset of symptoms, diagnosis and treatment of the studied patients.

Usually we diagnose CTS on the basis of the clinical picture and neurophysiological evaluation. The neurophysiological approach and the normal values have been previously extensively reported [9].

According to recommendations made by the American Association of Electrodiagnostic Medicine (AAEM) and of the Italian CTS Study group, the following electrodiagnostic protocol was adopted [9, 10]:

- median sensory nerve conduction velocity (SNCV) in two digits-wrist segment (thumb and third digit-wrist);
- median motor conduction velocity of arm and forearm;
- median distal motor latency (DML) from the wrist to thenar eminence;
- radial or ulnar SNCV for the digits-wrist segment.

Moreover, when standard tests bring normal results ("standard negative" hands), more sensitive tests (segmental or comparative studies) have to be performed [10].

Sometimes we ask to perform a neuroimaging examination to have a more comprehensive nerve assessment. In each of the 5 reported cases, we completed the diagnostic pathway by using US because of an atypical neurophysiological or clinical pattern. The instrument used to perform US was a Falcon 2101 EXL (BK Medical, Herlev, Denmark) equipped with a broadband (frequency band, 12–5 MHz) linear transducer.

Case 1

A 75-year-old woman was referred to our neurophysiological department complaining of nocturnal paraesthesia and pain in the right palmar side of the first three digits, which radiated to the forearm. Symptoms had started two years before. CTS had been previously diagnosed and she had been operated on for carpal tunnel syndrome one year before. Because of persisting symptoms she was operated again 6 months later without benefit. At clinical evaluation she had positive Tinel signs at the wrist and at the distal third of the forearm, but no mass was palpable. Mild deficit of median thenar eminence muscles was observed.

At conventional neurophysiological evaluation, we observed a focal median motor slowing at the distal third of the forearm with conduction block (*see* Fig. 1a).

By using near nerve conduction evaluation we confirmed focal slowing of sensory and motor median nerve conduction velocity in the distal third of the forearm; by using the motor "inching" test evaluation we confirmed the focal slowing and an abnormal difficulty in stimulating the nerve in a specific area, suggesting that an anatomopathological structure was interposed between the stimulation site and the nerve.

Because of the atypical pattern of median nerve abnormalities, suggesting focal damage in the forearm, we completed the diagnostic assessment by using US.

US showed a hypoechoic ovalar structure (1.5x5 cm) in continuity of median nerve in the distal third of the forearm (see Fig. 1c). This was confirmed by magnetic resonance imaging (MRI): after contrast medium injection the image showed a widespread enhancement of the structure with internal inhomogeneous features and a lobate border (see Fig. 1b). The patient underwent surgical excision (see Fig. 1d).

At histopathological evaluation the lesion was diagnosed as a cellular schwannoma. After surgical excision the patient had complete resolution of symptoms.

Case 2

A 77-year-old woman came to our department complaining of pain and dysaesthesia in the right palmar side of the thumb. She also reported awakening due to numbness.

At neurophysiological evaluation we did not detect sensory action potential (SAP) of median nerve in the thumb-wrist segment. While normal findings were observed at median nerve in II, III, and IV digit to wrist. Motor nerve conduction tests were always normal.

We diagnosed focal damage of the first sensory branch of the right median nerve. Because of the atypical pattern of median nerve abnormalities, suggesting isolated severe damage of a sensory branch of the median nerve, we completed the diagnostic assessment using US. US at the level of metacarpophalangeal joint showed the presence of a structure (0.20x0.23 cm) suggesting a schwannoma distal to the wrist in continuity with the sensory branch of the median nerve directed to the thumb (handling the region, the

Table 1 Timing of onset of symptoms, diagnosis and treatment

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Onset of symptoms	2001	2002	1998	2002	1999
Initial diagnosis	2001 diagnosis of CTS	2002	1998 diagnosis of CTS	2002	2000 diagnosis of CTS
Previous treatment	Surgical treatment for CTS in 2001		Laser therapy for CTS in 1998		Surgical treatment for CTS in 1999
Definitive diagnosis	2003 diagnosis and treatment of schwannoma	2002 diagnosis and treatment of schwannoma	2002 diagnosis and treatment of schwannoma	2002 diagnosis and treatment of schwannoma	2002 diagnosis and treatment of schwannoma

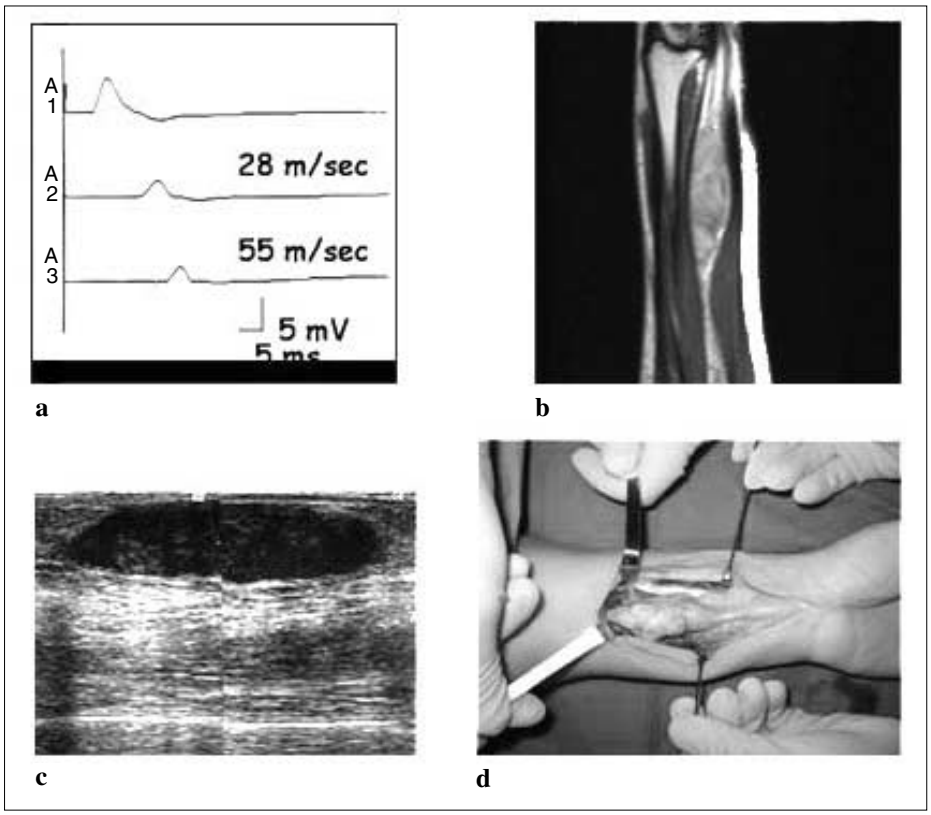

Fig. 1 *Case 1.* **a** Motor nerve conduction study of the median nerve: focal median motor slowing at the distal third of the forearm with conduction block. **b** T1-weighted gadolinium-enhanced MRI of forearm: the image shows an ovalar structure along the median nerve. **c** Ultrasonography of forearm: hypoechoic ovalar structure along the course of median nerve. **d** Median nerve surgical findings

mass was not distinguishable from the other rigid anatomical structures). She underwent surgical excision and at histopathological evaluation the structure showed to be schwannoma. After surgical excision the patient had complete resolution of symptoms.

Case 3

A 43-year-old man came to our department complaining of pain and paraesthesia in the right hand. At the initial interview he told us that he worked as a gardener and he told us that the symptoms worsened during heavy manual activities with a feeling of numbness. At the neurophysiological investigation we found a relative slowing of median nerve with respect to the radial nerve [10]. We diagnosed a "minimal" CTS [11]. One month after laser therapy and after stopping performing heavy manual work he came back for a second visit and we detected a slowing of sensory conduction velocity of the median nerve recording at the thumb. On a more accurate clinical examination of the hand we detected a palpable mass in the palm.

Considering that his conditions had worsened although he had stopped performing heavy work and also considering the presence of the palpable structure we requested a US examination. US showed the presence of a structure (0.3x0.5 cm) in the thenar branch at the palm. The patient was operated on for excision of the tumour and he improved rapidly. At anatomopathological evaluation the structure was shown to be a schwannoma. After surgical excision the patient had complete resolution of symptoms.

Case 4

A 59-year-old man came to our department because of pain or paraesthesia in the right hand. On clinical examination we observed a swelling on the right elbow in the median nerve region with a positive Tinel's sign in that region evoking radiating paraesthesia at the first finger. We did not find any sensory or motor deficits.

Neurophysiological evaluation showed normal sensory and motor conduction velocities of the median nerve also by using the motor inching evaluation in the swollen region.

Because of normal neurophysiological evaluation, sensory symptoms, Tinel sign and bump at elbow US was performed and it showed that the swelling originated from the median nerve.

He was operated on for excision of the tumour and he had transitory complications due to infection. Symptoms disappeared one month after surgery. At anatomopathological evaluation the structure showed to be a schwannoma.

Case 5

A 48-year-old woman came to our department complaining of paraesthesia in the median region of the hand exacerbated by some postures of the arm. One year before she had been operated on at the wrist for CTS on the basis of a clinical diagnosis (neurophysiological evaluation was not performed).

At clinical evaluation we observed weakness of thenar median innervated muscles but also uncertain weakness of flexor

longus pollicis. At neurophysiological examination we found no sensory action potential of the median nerve in digit-wrist segments associated with normal DML. This neurophysiological pattern suggested further proximal evaluation and we detected a mild relative slowing of motor conduction velocity at arm with respect to the forearm. Segmental evaluation showed focal slowing of the conduction velocity in the third middle of the arm; the other nerves we examined were normal.

By using US, a small ovalar structure was observed in the median nerve in the third middle of the arm. Excision of the tumour was particularly difficult because the tumour tightly adhered to the nerve structure. Anatomopathologically, the tumour was diagnosed as a schwannoma. After surgery the patient complained of pain and worsening of the weakness (score 3 at BMRC scale) of the median innervated muscles. At following EMG evaluations, we observed mild axonal damage in median innervated muscles, absence of median sensory responses and slowing of median motor conduction velocity in the region of the scar. One year later, normal strength was observed although we still detected absence of median nerve sensory responses, while motor conduction velocity at the site of the scar was almost normal. Two years after, median muscle strength was normal. At neurophysiological evaluation a small median sensory response was detectable (1 μV) and median motor nerve conduction was normal in all segments.

Discussion

CTS may be idiopathic but it may be associated with various conditions such as Colles' fracture, rheumatoid arthritis, diabetes mellitus, disthyroidism, pregnancy and jobs requiring excessive use of the hands [12]. Finally, CTS may be due to tumours at the wrist [2, 3].

Diagnosis is usually based on clinical findings and neurophysiological evaluation is really useful because it provides evidence of focal myelin impairment typical of the entrapment condition [13, 14].

Recently ultrasonographic measurements of the nerve, especially to assess focal lesions, have become more common, because they are non-invasive tests providing evidence on the anatomopathological condition [15–19].

At sonographic examination of the extremities, using high frequency "small-parts" equipment, peripheral nerves may be virtually identified in all patients. Peripheral nerves have a typical ultrasonographic pattern that correlates well with the histological structure and facilitates differentiation between nerves and tendons. The ability of this technique to depict peripheral nerves makes it possible, in many instances, to study nerve abnormalities in trauma, entrapment syndromes and tumours. Ultrasounds can differentiate endoneural from extraneural space-occupying lesions and evaluate the extent and consistency of the lesion [11, 20]. When evaluating abnormal masses, high-resolution US cannot clearly differentiate neurofibromas from schwannomas but it can clarify the relationship between tumour and neural trunk [21].

Because of their complementary perspectives we have started to add the US assessment to the neurophysiological evaluation.

In our sample, all patients complained of symptoms that could be due to CTS. In two cases they were operated on for CTS without benefit. At neurophysiological examination all patients presented a pattern that could be partly due to CTS. Nevertheless we also performed US assessment for of the following reasons: (1) electrodiagnostic studies were equivocal, (2) the clinical picture presented some aspects that could be not due to CTS, (3) there was dissociation between the clinical and neurophysiological pictures. Four out of five cases were negative at physical examination for a mass (non-palpable). In all these cases we observed abnormal nerve conduction findings. Obviously, particularly in these cases, the association of the neurophysiological examination and US had a more relevant impact.

Combining neurophysiological and US evaluations we diagnosed a tumour in different segments along the course of the median nerve (arm, elbow, forearm, wrist and palm) whose symptoms mimicked CTS. This study confirms the usefulness of US [8, 11, 15–21]. Note that US provided exhaustive data. As we had a lack of long experience in US of nerves, in some cases MRI was requested in order to confirm US findings prior to operative exploration (MRI always confirmed the US observation without adding further information).

In conclusion we know very well that CTS is a very common disease while schwannoma is an uncommon condition. There are few studies describing misdiagnosis of CTS due to median nerve tumour [2, 3, 22–24]. In our sample tumours outside the wrist produced symptoms similar to those provoked by CTS. Making a diagnosis of CTS in the presence of CTS symptoms is easy, but in cases of atypical abnormalities at neurophysiological examination, atypical clinical CTS or dissociation between neurophysiological and clinical findings, physicians should consider the presence of a median nerve tumour.

In these cases, US evaluation is very useful as a supporting diagnostic method to assess the anatomopathological conditions of the nerve lesion and must not be limited to the wrist. US and neurophysiological evaluation performed in the same session (or in a synergic collaboration with the US examiner) may increase the sensitivity of both tests because of a feedback mechanism obtained to localise and to identify the damage. We recommend US when the clinical–neurophysiological results are atypical or there is dissociation between the clinical and neurophysiological pictures.

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