RESUMO – A variante ulcero-mutilante da síndrome do túnel cárpico ocorre em casos de doença prolongada não tratada. Os doentes apresentam-se com úlceras indolores do segundo e terceiro dedos da mão, acompanhadas por outras alterações cutâneas e sensitivas. Estes doentes são frequentemente diagnosticados erroneamente como tendo doença de Raynaud ou esclerose sistémica. A avaliação clínica é o gold-standard no diagnóstico do síndrome do túnel cárpico, mas a radiografia da mão e a eletromiografia do membro superior contribuem no diagnóstico. Os autores apresentam dois casos desta variante ulcerante da síndrome do túnel cárpico.

PALAVRAS-CHAVE – Síndrome do Túnel Cárpico; Úlcera da Pele.

ABSTRACT – Ulcerating and mutilating variant of carpal tunnel syndrome occurs in longstanding cases of untreated disease. Patients present with painless ulcers of the second and third fingers, accompanied with other cutaneous and sensory changes. These patients are often misdiagnosed as having a Raynaud disease or systemic sclerosis. Clinical assessment is the gold standard for the diagnosis of carpal tunnel syndrome, but hand radiography and electromyography help supporting the diagnosis. The authors present two cases of this ulcerating variant of carpal tunnel syndrome.

KEYWORDS – Carpal Tunnel Syndrome; Skin Ulcer.

INTRODUCTION
Carpal tunnel syndrome (CTS), is the most common nerve entrapment disorder, and results from the localized compression of median nerve at the wrist.1,2 It usually presents with the classic triad of nocturnal pain, hypoesthesia and thenar atrophy, but in longstanding cases it can present as an ulcerating and mutilating variant.3 Ulcerating CTS is characterized by ulcerative skin lesions, skin atrophy and acro-osteolysis in the sensory zones of the median nerve.4

The authors describe two cases of CTS presenting with cutaneous ulcers of the digits.

CASE REPORTS
Case 1
A 83-years-old male patient presented with a digital ulceration of left hand, that appeared 4 months before. The patient complained of paresthesia of the second, third and fourth finger of the left hand for 30 years, without pain. At physical examination, there was an ulceration on the pulp of the left third finger, with an erosion on the lateral aspect of the same finger (Fig. 1) and another erosion on the pulp of second finger. Complete blood count and C-reactive protein were normal. Electromyography of upper left limb revealed absence of motor and sensory response of the median nerve, compatible with a severe CTS. The responses of radial and ulnar nerve were normal.

With the diagnosis of ulcerating and mutilating CTS the patient was referred to surgical release of carpal tunnel content. Four months after surgery, there was a complete resolution of the ulcers and a partial improvement of paresthesia (Fig. 2).
A 78-years-old woman was referred to our Department for digital ulcerations on both hands present for 8 months. She also complained of arthralgia of the proximal and distal interphalangeal joints, with no associated Raynaud phenomenon. Her past medical history included osteoarthritis of the interphalangeal joints and CTS previously proposed for surgery, that the patient refused. At physical examination, there was sclerodactyly and ulcerations on the fingertips of the second and third fingers of both hands (Fig. 3). Complete blood count was normal. Anti-nuclear antibodies were negative. Electromyography revealed complete absence of motor and sensory response of median nerve, compatible with severe CTS. Cubital and radial nerve studies were normal.

The patient was diagnosed as an ulcerating and mutilating bilateral CTS. Carpal tunnel surgery was proposed again but the patient refused.

DISCUSSION

The ulcerating and mutilating variant of CTS was first described by Bouvier et al in 1979. Cutaneous involvement can be detected in 20% of patients with severe CTS. Although CTS is more frequent in women, this variant is more prevalent in males. Usually, these patients have been suffering from classical CTS for years, without seeking medical advice.

Cutaneous lesions appear as painless ulcerations on the palmar surface or sub-ungual region of the third phalanx.
of the forefinger and second finger. Median nerve branches supply the sensory innervation of the palmar aspect of the first, second, third and radial half of the fourth fingers and of the dorsal surface of the second and third fingers distal to the proximal interphalangeal joint. The first finger is usually unaffected due to its mixed innervation from both median and radial nerves. Other cutaneous signs may include erythema, edema, peri and sub-ungual abscesses, hemorrhagic blisters, adjacent hyperkeratosis and sclerodactyly. Nails may also be affected, showing discoloration, onycholysis and hyperkeratosis of the nail plate and cuticle hypertrophy.

Cutaneous involvement occurs in patients with severe damage of motor, sensory and, above all, autonomic fibers of median nerve. Compression and ischemia are the most important pathogenic factors in nerve damage. The carpal tunnel compression causes obstruction of venous return in the epineural venules and veins. This leads to infranunical edema, further increase in pressure and, ultimately, infranunical anoxia. The defective sensory nerve favors physical and thermal trauma by increasing the thermo-algesic perception threshold. The compression of autonomic fibers results in hypohidrosis, swelling and increases skin temperature.

Clinical assessment is considered the gold standard for CTS diagnosis. Therefore, it is essential to take an accurate clinical history. Neurologic examination may show hypoesthesia and dysesthesia in the median nerve area, atrophy of the thenar muscles and positivity of Tinel’s sign and Phalen’s test. Electromyography and hand radiography can support the diagnosis. A hand radiography may show acral osteolysis with bone reabsorption of the distal phalanx of the affected fingers.

Hydrocolloid and hyaluronic acid dressings may improve the ulcers, but the most effective treatment is surgery. Ulcerations usually improve with the surgical treatment of CTS, but osteolysis is permanent. Surgical treatment consists in releasing carpal tunnel content by transection of the transverse carpal ligament.

CONCLUSION

Patients with digital ulcers are often referred with a presumed diagnosis of systemic sclerosis or Raynaud phenomenon. Therefore dermatologists should suspect this mutilating variant of CTS in patients who present with ulcerations of the second and third fingers accompanied with sensory changes.

Presentations/Awards: The current work was presented as a poster at “Reunião da Primavera da SPDV 2019” and was awarded as 1st prize in poster category.
Caso Clínico


