Painless Ulcers on the Fingers: An Unusual Presentation of Severe Bilateral Carpal Tunnel Syndrome

Úlceras digitales indoloras como presentación inusual de síndrome de túnel carpiano bilateral severo

To the Editor:

Carpal tunnel syndrome (CTS), the most frequent type of entrapment neuropathy, is caused by compression of the median nerve inside the tunnel formed by the wrist bones and the carpal annular ligament. This condition affects between 1% and 3% of the population, and it is 4 times more common in women than in men. Incidence peaks between the fourth and sixth decade of life, and more than 50% of cases are bilateral. Although most cases are idiopathic, the compression of the nerve is occasionally due to a combination of factors that increase pressure on this nerve. CTS commonly presents with a triad of nocturnal pain, hypoaesthesia, and thenar atrophy. We describe a case of CTS that was diagnosed based on the presence of painless cutaneous ulcers with a characteristic location.

An 86-year-old male nonsmoker with a history of cardiac arrhythmias and lower urinary tract symptoms was referred to our unit with painless ulcers on both hands that had appeared 1 month earlier. He was on treatment with furosemide, acenocoumarol, dutasteride, and tamsulosin. During the physical examination, we observed lesions on several fingers of both hands, bullous lesions on the thumbs, and ulcers on the tips of the index and middle fingers of the right hand and middle finger of the left hand. In addition, a short distal phalanx and a short, wide fingernail were observed in the right index finger (Figs. 1 and 2). Radial and brachial peripheral pulses were normal.

Based on the loss of sensation to pain and the distribution of the lesions, CTS was suspected and the patient was referred to the neurology department. A neurological examination revealed thenar atrophy and loss of pressure sensation in the median nerve territory, with preserved sensation in the forearms and in the ulnar and radial nerve territories of the hands. Electroneurography showed a bilateral loss of sensory and motor function in the median nerve distal to the carpal tunnel.

Blood tests revealed an elevated erythrocyte sedimentation rate and an immunoglobulin A gammopathy, but further testing by the hematology department ruled out multiple myeloma. Rheumatoid factor, cryoglobulin, cold agglutinin, and autoantibody tests were normal.

Hand radiographs showed bone resorption in the distal phalanx of the right index finger (Fig. 3).

The patient was advised to take measures to avoid mechanical and thermal injury, and the skin lesions healed after several weeks with the use of topical treatment. He refused surgical treatment.

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Skin ulcers in CTS are rare. They were first described by Bouvier et al.\textsuperscript{1} in 1979 and few cases have since been published. The ulcerative and disfiguring form of CTS is characterized by the appearance of painless skin ulcers on the fingertips and under the fingernails, sclerodactyly, and acroosteolysis in the sensory hand area innervated by the median nerve.\textsuperscript{2} Skin involvement is observed in 20\% of CTS cases\textsuperscript{3} and is caused by the compression of autonomic fibers in the median nerve. This explains why skin manifestations occur in patients with severe CTS.\textsuperscript{4} Vasomotor dysfunction can lead to Raynaud phenomenon and skin necrosis, and sensory dysfunction can result in mechanical or thermal injury, which may in turn cause necrosis to spread.\textsuperscript{5} Cutaneous lesions include erythema, edema, blisters, painless ulcers on the fingertips and under the fingernails, nail discoloration, onycholysis, gangrene, autoamputation, and acroosteolysis.\textsuperscript{6} In most patients, the second and third fingers are involved and the main area affected is the volar side of the distal phalanx.\textsuperscript{7} These fingers are affected most because the innervation of the fingers is mixed, originating from both median and radial nerve fibers, and because they are more vulnerable to injury.\textsuperscript{8,9} Acral ulcers and osteolysis are frequently unilateral, although some bilateral cases have been described. In lesions of this type, the differential diagnosis should include collagenopathies, autonomic neuropathies, hematologic diseases, injuries due to external trauma, metabolic diseases, and vascular pathologies.\textsuperscript{2,10}

Diagnosing CTS is simple if the classic manifestations are present; physical examination is therefore useful. Phalen and Hoffman-Tinel signs are highly suggestive of CTS. Imaging techniques are used to evaluate the strictures of the carpal tunnel and rule out the presence of fractures. However, to confirm the diagnosis and evaluate the degree of median nerve involvement, a neurophysiological examination is required.

It is important for the dermatologist to correctly evaluate these types of lesions because early diagnosis is essential to preventing bone lesions and irreversible deformities.

References

Localized Primary Cutaneous Nodular Amyloidosis in a Patient With Paraproteinemia

To the Editor:

The term primary cutaneous amyloidosis (PCA) refers to a group of diseases caused by the extracellular deposition of amyloid in the skin without the involvement of other organs. PCA has been divided into the following types: macular, lichen and nodular. The first 2 types are characterized by the deposition in the papillary dermis of amyloid derived from the degeneration of keratin filaments. Primary localized cutaneous nodular amyloidosis (PLCNA) is the rarest form of PCA and the only one in which the amyloid deposits are of the amyloid light-chain (AL) type, as in the primary and myeloma-associated systemic forms of amyloidosis. AL amyloidosis is due to monoclonal immunoglobulin (Ig) light-chain deposition.

We report the case of an 83-year-old man with a history of systemic hypertension, type 2 diabetes mellitus, and hyperuricemia, who was seen for a 2-year history of asymptomatic but progressive lesions that had appeared on the left lower limb. One year before consultation, he had been diagnosed with monoclonal gammapathy of undetermined significance (MGUS), after detecting an IgG(k) paraprotein in the serum with no evidence of multiple myeloma in the bone marrow study. Physical examination revealed a plaque with an area of ecchymotic appearance and several hard, reddish-orange nodules with an ulcerated surface in the pretibial region of the left leg (Fig. 1).

Histopathological examination showed deposits of an amorphous eosinophilic material diffusely distributed through the papillary and reticular dermis, extending into the subcutaneous tissue, accompanied by a dense infiltrate of plasma cells (Fig. 2). When stained with thioflavin, the deposits fluoresced under ultraviolet light. Immunohistochemical staining demonstrated a light-chain restriction in the majority of the plasma cells. These findings suggested a possible diagnosis of PLCNA, although systemic amyloidosis could not be ruled out.

Laboratory tests including complete blood count, biochemistry tests, and liver and kidney profiles were normal. Immunoglobulin concentrations were normal, although the IgG value was at the upper limit of normal: 1590 mg/dL (normal range, 700–1600 mg/dL). Immunelectrophoresis showed a monoclonal IgG(k) band with oligoclonal λ bands in the serum, but was normal in the urine. No amyloid deposits were observed in a biopsy of the abdominal fat. After 18 months of follow-up, the patient has not shown evidence of progression to myeloma or systemic amyloidosis.

PLCNA is a disease that occurs predominantly in women, with a mean age at diagnosis of 60 years. It has sometimes been described in association with systemic diseases such as Sjögren syndrome, diabetes mellitus, or CREST syndrome. It manifests as waxy nodules that are usually solitary or localized or, less commonly, disseminated. It tends to affect acral areas, the most common site being the lower limbs, followed by the face and trunk. Histopathological findings are indistinguishable from those of primary systemic amyloidosis or myeloma-associated amyloidosis. Immunohistochemical staining may show the presence of immunoglobulin light chains in the amyloid or in the cytoplasm of plasma cells.

Based on the demonstration of clonality of the plasma cell infiltrate in the skin using gene rearrangement techniques and on the absence of clonal rearrangement in bone marrow, some authors have suggested that PLCNA should be considered an extramedullary plasmacytoma that produces localized amyloid deposition. Extramedullary plasmacytomas are plasma cell neoplasms that arise in any organ except bone marrow; they are able to produce an M-component in the serum in up to 20% of cases. Extramedullary plasmacytomas that initially do not produce the M-component in serum progress to multiple myeloma in 20% of cases.

In our patient, there was a monoclonal infiltrate of plasma cells that synthesized λ chains and produced the amyloid deposits in the skin, and a finding of a monoclonal IgG(k) band and oligoclonal λ bands in the serum. This suggests a biclonal gammapathy (the presence of 2 monoclonal components in serum), which has been observed in approximately 3% of MGUS.

The appearance of paraproteinemia in cases of PLCNA has rarely been described in the literature, though it should always be investigated at the initial assessment. In the cases in which it has been observed, it was generally associated with an advanced stage of the disease, and may indicate progression to systemic amyloidosis.

The risk of progression to systemic amyloidosis in PLCNA has been well defined by several authors. Since 1970, the risk has been established at around 50%. However, in 2001, Woolons and Black observed only 1 case of progression to systemic amyloidosis in a series of 15 patients with PLCNA, in addition to high immunoglobulin concentrations in 40%.