

# Sporadic Ulcerative Mutilating Pseudosyringomyelic Acropathy: A Sort of “Melting Toe Disease”

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## Introduction

The term distal arthropathy comprises a group of conditions that includes neuropathic damage of joints, infections, inflammatory disorders as well as metastatic tumors of the peripheral bones and joints. Neuropathic arthropathy (Charcot's joint) was first described by Jean-Martin Charcot in 1868 in patients with tabes dorsalis and it was characterized by progressive destructive arthritis which associates loss of pain and deep sensitivity. In addition, normal muscular reflexes that modulate joint movement are decreased. Without these protective mechanisms, joints are subjected to repeated trauma, resulting in progressive cartilage and bone damage [1]. Nowadays, diabetes mellitus is the most frequent cause of neuropathic joint disease, but leprosy, yaws, syringomyelia, meningomyelocoele, peroneal muscular atrophy (Charcot-Marie-Tooth) or amyloidosis are occasionally seen and should be considered as a potential cause of destructive neuropathic arthritis.

We show herein two cases of sporadic ulcerative mutilating acropathy, an unusual form of neuropathic arthropathy, in heavy drinkers.

## Cases Report

### Case 1

A 55 year-old man was admitted to the hospital because of an ulcerative and destructive lesion in the right first during the last three months. He had a long history of alcohol abuse and was repeatedly brought to the emergency ward because of acute alcoholic intoxication.

An X-ray film showed osteonecrosis of the right first toe. He was treated with ciprofloxacin and clindamicin during 4 weeks, without a clinical response, so amputation of the distal phalanx of the right big toe was performed.

Four weeks later a similar ulcerative, painless lesion appeared on the left big toe. Local treatment with antibiotics did not determine any improvement and finally he was brought to the hospital because the restless and progressive destruction of the toe.



**Figure 1:** Ulcerating, necrotizing lesion of the left toe.

On admission, edema and redness of both feet was noted. An ulcerating, necrotizing lesion of the left toe was found (Figure 1). Patellar reflexes were exalted, whereas Achilles deep tendon reflex was bilaterally negative. Examination of sensitivity showed absence of the thermo-analgesic sensitivity in the distal parts of the lower limbs. Vibration sense was absent as well. Femoral, popliteal, and distal pulses were present in both extremities. Physical examination was otherwise unremarkable.

New X-ray films showed osteonecrosis of metatarsal and phalanges of the left first toe, as well as second, third and fourth metatarsal fractures.

Laboratory studies revealed a mild leukocytosis of 12,500/mm<sup>3</sup>; hemoglobin was 10.5 g/dl with platelet count within normal limits. The erythrocyte sedimentation rate was elevated at 105 mm/h and the C-reactive protein was 22.6 mg/dl (normal, 0-0.5 mg/dl). The patient had normal liver and renal function studies. There was no folic acid neither vitamin B12 deficiency.

Serologic studies for HIV and hepatotropic viruses were negative. The electromyogram showed signs of both motor-sensitive polyneuropathy, with axonal and sensitive predominance, which was severe in lower limbs and mild to moderate in upper extremities.

MR imaging was consistent with osteonecrosis of the first right metatarsian and phalanges of the first toe with soft tissue edema and bone rarefaction of the rest metatarsians (Figure 2). Because of the thermo-analgesic sensory deficit observed on physical examination, a complete spine MR imaging was performed in order to rule out a syringomyelic disorder. Chronic, osteoporotic vertebral fractures of D8 and L1 and degenerative discopathy were observed but signs of syringomyelia were absent.

Skin biopsies showed intense, succulent vascular proliferation in and around necrotic areas. No microorganisms were observed by means of Ziehl-Nielsen, Giemsa, Gram, PAS or Grocott stains.

### Case 2

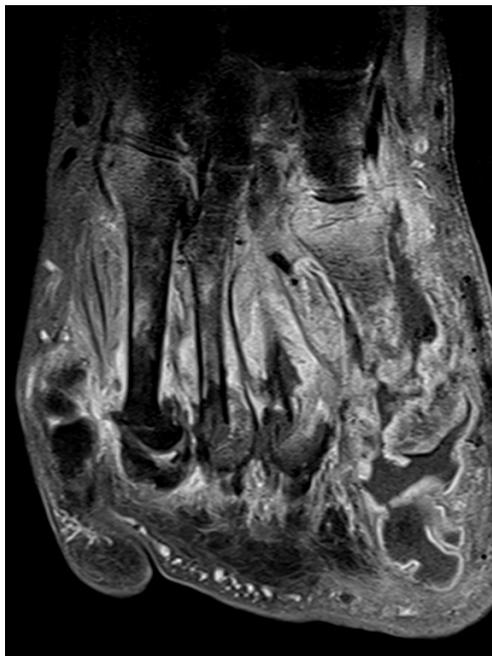
A 54 year-old man was admitted because of necrosis of the great right toe. He was a heavy drinker with frequent visits to the emergency room because of alcohol intoxication. Fever and pain

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**Figure 2:** MR imaging shows osteonecrosis of the first right metatarsian and phalanges of the first toe with soft tissue edema and bone rarefaction of the rest metatarsian bones.



**Figure 3:** Notice the swollen and cyanotic toe and areas of distal necrosis.

were absent.

Examination showed complete necrosis of the right great toe, edema and redness of the foot. Femoral, popliteal and peripheral pulses were present in both extremities. Examination of sensitivity showed complete absence of the thermo-analgesic sensitivity in the distal parts of the lower limbs.

Laboratory studies showed a normal leukocyte and platelet cell counts; hemoglobin was 12.5 g/dl. Red cell macrocytosis was observed in peripheral smears. C-reactive protein was 13.3 mg/dl. The patient had normal liver and renal function test. There was no folic acid or

vitamin B12 deficiency. Serologic studies for HIV and hepatotropic viruses were negative.

The electromyogram showed signs of severe motor-sensitive polyneuropathy, with axonal and sensitive predominance. Stimulation of both peroneal and tibial nerves did not arise any response. A complete amputation of the great toe was performed and the patient was dismissed. Few days later the patient was brought to the emergency room because of tumefaction and discoloration of the 2<sup>o</sup> right toe. Pain was absent. The toe was swollen, cyanotic with areas of distal necrosis (Figure 3). Peripheral pulses were once again felt.

A diagnosis of ulcerative mutilating pseudosyringomyelic acropathy was established. Local care and antibiotics did not improve the patient and he was dismissed to the outpatient clinic.

## Discussion

Sporadic ulcerative mutilating acropathy was first described by Bureau and Barrière in 1953 [2]. It consists on a rare trophic complication of alcoholic neuropathy which results in a chronic disease characterized by analgesic neuropathy, dysautonomic vasomotor disturbance and destruction of distal joints. It is also called “Bureau and Barrière ulceromutilant acropathy” or “vagabond’s disease”, and usually occurs in individuals with a debilitated condition, chronic alcoholism and unfavourable socioeconomic conditions [3]. Previously, in 1942, Thévenard described a similar clinical syndrome with familial distribution, well as an autosomal dominating inheritance, or as a recessive form. Since the first description, some sporadic cases have been reported, where the main differences from the familial form were a later clinical onset, the predominance of male gender and the concomitant history of malnutrition, liver disease and/or addiction to alcohol [4].

The sporadic ulcerative mutilating pseudosyringomyelic acropathy is a very rare neurogenic osteoarthropathy defined by the coexistence of a sensitive polyneuropathy of lower extremities, painless mutilant ulcers and destructive osteolysis of feet bones [4,5]. It is usually observed in males in the fourth or fifth decade of life, with a history of alcohol indulgence and malnutrition [4].

The most common presentation is the formation of a callus on pressure areas, usually in the head of the first and fifth metatarsal or heel, or areas of shoe friction that finally ulcer either spontaneously or after minor trauma [3,5]. Bone deformities with decreased bone calcium and osteolytic areas may trigger pathological fractures or osteomyelitis [6]. Painless ulcer has no spontaneous tendency to healing, with restless, progressive destruction of the toes that finally results in “melting” of the toes.

Remarkable findings on physical examination are a conspicuous absence of pain in the lesion coupled with a “shaped-sock” thermo-analgesic dissociation that is often accompanied by hypo or analgesia with preserved tactile and proprioceptive sensitivity. This affected area may develop trophic changes such as dark pigmentation, smooth and shiny skin, nail alterations or even excessive sweating or cyanotic tone of teguments [5].

A careful history and physical examination are of paramount importance for accurate diagnosis. Pathological findings consists of almost complete sensory denervation, with loss of myelinated fibres, axon deficiency, Schwann cell proliferation and increase of epi- and perineural tissue [7]. Some of these changes may be translated into an abnormal electromyogram. Wallerian degeneration with predominant axonal involvement resulting in a decrease of the response amplitude with normal conduction speed is characteristic [3,6]. As shown by

case 1, increased microvasculature with the presence of arterio-venous shunts is a remarkable finding [8,9].

There is no specific treatment. Alcohol withdrawal, hygiene measures and improving nutritional status are strongly recommended, as well as avoiding overweight and humidity. The use of orthopedic shoes that relief or minimize hyper-pressure areas are also recommended. Letting the lower limbs rest for several hours a day is also associated with a more favorable outcome. Antibiotic should be reserved for cases with documented infection of bone and soft tissues.

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