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Charcot osteo-arthropathy

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KEYWORDS
Charcot osteo-arthropathy;
Neuropathic;
Diagnosis;
Treatment

Summary
Charcot osteo-arthropathy is a potentially catastrophic complication of neuropathy. It can occur in any joint and has been associated with many of the causes of sensory deprivation; however it most commonly presents in the foot and ankle in the diabetic population. This article stresses early recognition and prevention of deformity. The key topics are: **Pathophysiology:** unrecognised microtrauma versus osseous hyperaemia as likely aetiologies, leading to fragmentation of the affected joint or bone. **Assessment:** diagnosis of neuropathy, x-ray changes, alternative imaging and its limitations and investigation to rule out other causes. **Classification and staging:** current staging of Charcot osteo-arthropathy and anatomical classification of foot and ankle Charcot osteo-arthropathy. **Treatment:** Current recommended treatment modalities including conservative, operative and pharmacological.

Introduction

In 1868 Jean-Martin Charcot, an eminent neurologist at the Salpêtrière Hospital in Paris, described a painless destructive joint osteo-arthropathy associated with tertiary syphilis (tabes dorsalis),[1] a common malady of the time. Although the painless destruction of joints had been previously described by other physicians, it was Charcot who concisely described the neurological element of the disease and the phrase ''Charcot joint'' or ''Charcot osteo-arthropathy'' has subsequently been used to describe any joint arthropathy resulting from a neurological condition with sensory loss. Peripheral neuropathy (including metabolic nerve injury, compressive neuropathies, infective nerve injury and re-implantation/limb transfer procedures) or a central nerve lesion (with associated inability to receive or interpret painful stimuli) may lead to Charcot osteo-arthropathy. Many of the identified neurological conditions associated with Charcot osteo-arthropathy are summarised in Table 1.

The terms 'neuropathic arthropathy' and 'Charcot joint disease' have become interchangeable. The precise semantics of the nomenclature should however consider all the skeletal changes secondary to a sensory neuropathy as bone distant to a joint can be affected. It is thus more properly described as being an osteo-arthropathy and hence the term Charcot osteo-arthropathy is more accurate. Indeed it is probable that the osteopathy precedes any joint changes and will be discussed within the pathophysiology of the disease.

Charcot osteo-arthropathy predominantly affects the foot, ankle and the knee, and is most commonly seen in the

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Table 1 Neurological conditions known to be associated with Charcot Osteo-Arthropathy

Metabolic nerve injury
- Diabetes mellitus
- Chronic alcoholism
- Steroid use (including intra-articular injection)
- Renal dialysis
- Amyloidosis
- Haemochromatosis

Infective nerve injury
- Poliomyelitis
- Guillain-Barré syndrome (post-infective auto-immune neuropathy)
- Syphilis (Tabes Dorsalis) (*Treponemia pallidum*)
- Hansen's disease (Leprosy) (*Mycobacterium leprae*)
- Lyme disease (*Borrelia*)
- Yaws (*Treponemia pertenue*)

Compressive neuropathies
- Rheumatoid arthritis
- Extreme gigantism
- CREST syndrome
- Klippel-Trenaunay-Weber syndrome
- Spondyloepiphyseal dysplasia (short-trunk disproportionate dwarfism)
- Pseudogout (calcium pyrophosphate dihydrate deposition)
- Tophaceous gout

Central nerve lesion
- Spinal cord lesions
- Charcot-Marie-Tooth syndrome
- Meningomyelocele/Spina bifida
- Syringomyelia
- Congenital insensitivity to pain (asymbolia)
- Riley-Day syndrome (hereditary sensory and autonomic neuropathy)
- Paraneoplastic sensory neuropathy
- Multiple sclerosis

Miscellaneous
- Trauma to peripheral nerves
- Allograft and re-implantation/limb transfer procedures
- POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes)
- Neurofibromatosis
- Thalidomide embryopathy

feet of patients with neuropathy associated with diabetes mellitus. Charcot osteo-arthropathy affects approximately 7.5% of all diabetic patients. Around 29% of diabetic patients with a peripheral neuropathy will be diagnosed with Charcot joint disease.[2] It is thought that an increase in inadvertent trauma from walking with a greater degree of distal sensory loss are factors contributing to increased lower limb problems over upper limb problems. Reports of knee and upper limb Charcot osteo-arthropathy have been recorded in syringomyelia when a quarter of these patients will suffer from a Charcot osteo-arthropathy, 80% with upper limb involvement (Fig. 1). Syphilis is also becoming an increasingly causative agent as the rate of sexual transmission of the spirochete *Treponemia pallidum* increases. The disease is often unrecognised or partially treated by the empirical use of antibiotics and so patients with tertiary or neurological syphilis presenting with non-classic signs and symptoms. A high index of suspicion is required and appropriate serological tests instigated including a VDRL screen in patients with a Charcot osteo-arthropathy of unknown aetiology.

In the presence of a neuropathy, even the most minor traumatic injury can lead to progressive joint destruction. A careful history and examination may elicit the cause although the history is often non-specific and investigations, including roentograms, may mimic many other causes of bone and joint destruction. It is important to understand the pathophysiology of this disease in order to understand its natural history and to enable the treating surgeon to instigate the appropriate investigations in the management of this debilitating disease. As the understanding of bone metabolism and its relation with neurological pathology advances, the potential for medical treatment becomes ever larger.

Pathophysiology

Although Charcot believed that the joint destruction was secondary to unrecognized traumatic events as a result of sensory deficit, contemporaries of the time had observed that fractures of the metatarsals in insensate feet healed without complication with exuberant bone formation. In 1917, Eloesser confirmed the direct relationship between nerve injury and joint destruction by sectioning the posterior (sensory) nerve roots to the forelimbs of 38 cats.[3] Following a period of activity Eloesser noted neuropathic bony changes in 71% of the animals. It is thought that the loss of proprioception and deep sensation ultimately leads to progressive joint degeneration, destruction, and disorganization secondary to repetitive unrecognized trauma. Six decades later, however, Finsterbush and Friedman sectioned the posterior nerve roots to the hind limbs of rabbits[4] in a similar fashion. In this study the denervated animals and a control group of normal animals were then immobilized in casts. Joint destruction differed between normal and denervated groups despite both being protected from traumatic injury. Finsterbush and Friedman concluded that trauma was an important but not the primary factor leading to the deterioration of insensate joints.

It was thus suggested that there is both a neurological and a neurovascular element in the pathophysiology of neuropathic osteo-arthropathy. Using scintigraphy, it has been shown that in patients with diagnosed neuropathy there is increased blood flow within bone[5] thought to be due to an autonomic, neurally-mediated vascular reflex ultimately resulting in a hyperaemia. In addition to repetitive unrecognized trauma hyperaemia may cause an osteopenia secondary to a mismatch in bone destruction and synthesis[6] making bone more susceptible to repeated minor trauma. The increased blood flow in active Charcot osteo-arthropathy can be demonstrated within the outpatient setting with the simple use of portable infrared dermal thermometry. This is a sensitive but non-specific non-invasive test and can only be used to monitor the disease once the diagnosis of Charcot osteo-arthropathy has been made.