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CASE REPORT

# Ankle Fusion in grossly deformed "Idiopathic Charcot's Neuroarthropathy"

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## ABSTRACT

This typical case aged 65 years is reported with history of injury in childhood, clinically has progressed to ankle fusion following surgery 7 years early with multiple screws. Anti-tumor necrosis factor / methotrexate is recommended early in management to prevent progression of inflammatory pathology.



**Keywords:** Arthrodesis, Charcot's neuroarthropathy, Diabetic neuropathy.

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## SUMMARY

This unique case presenting with typical Charcot's destructive hypertrophic osteoarthropathy is neither a case of diabetic neuropathy nor any other disease could be attributed, such as alcoholism, any chemotherapy, use of any heavy metal, toxins (ethanol, drug related), leprosy infection, tabes dorsalis, traumatic paraparesis, syringomyelia, Parkinson's disease, rheumatoid arthritis, psoriasis, sarcoidosis, and human immunodeficiency virus (HIV) infection. Current understanding of this neuroarthropathy postulates initiation of posttraumatic uncontrolled inflammation responsible for continue osteoarticular changes. This particular patient had only injury to the ankle during his childhood. It is for this atypical presentation that this condition has been christened "Idiopathic Charcot's Neuroarthropathy." Ankle arthrodesis using multiple screws after correction of the deformity seemed to give encouraging result, but after 7 years' follow-up, there are evidences of delay in union and fracture of screws but clinically the ankle is stable. Because of inflammatory pathogenesis, a treatment protocol with methotrexate or anti-tumor necrosis factor (TNF) alpha is recommended early in management of such condition.



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## INTRODUCTION

This disease is named after the famous French neurologist Jean-Martin Charcot. In 1968, he gave first in-depth description of such destructive hypertrophic osteoarthropathy affecting joints of tertiary syphilis.[1-3] Penicillin virtually eradicated tertiary syphilis.

Diabetes with increased survival due to insulin and long-standing peripheral developing neuropathy appears to be the precursor of this neuropathic osteoarthropathy.

A benchmark monograph by Eichenholtz in 1966 cataloged the clinical, radiographic, and pathological data from 68 consecutive patients. Based on his light microscopic photographs, pronounced osteoclastic activity was noticed.

Occasionally, patients with neuropathy of other etiologies, such as alcohol, chemotherapy, and heavy metal, were also present. More than half the patients cite a specific traumatic episode, often trivial, initiating the process. Some of the patients are obese. Patients typically present in sixth and seventh decade.

## CASE REPORT

A patient aged 65 years, weighing 68 kg and height 5.5 feet, presented with a deformed right foot walking in extreme varus and rocker bottom appearance, no swelling, and had no infection.

He was operated, the deformity corrected, and after bony contact was fixation with multiple screws was done. The operative wound healed uneventfully (Figs 1A to J).

The limb was protected in below-knee plaster cast for 4 months and then in ankle foot orthosis for further 6 months. He has been walking comfortably for the last 7 years. He walks even bare footed and use ordinary shoes, too.

X-rays are now clearly showing fracture of few screws, but the foot is clinically stable.

This patient's radiograph is typical of Charcot's osteoarthropathy showing destruction of tibia fibula and hindfoot bones of the foot. This patient is different as far as pathogenesis is concerned. He never had diabetes and also no evidence of tertiary syphilis could be pinpointed. The only history that could be articulated was a childhood injury of ankle which was not properly treated and the foot started gradually deforming from age 50 onward and finally presented as the foot became unshoeable and even barefoot walking became difficult. Hence, in this particular patient, the diagnosis has been labeled "Idiopathic Charcot's Neuroarthropathy."

## DISCUSSION

The ankle fusion, which seemed to be progressive in 2 years' follow-up, started showing fracture of screws and the condition warrants further follow-up and it requires aggressive surgical exercises to lead to ankle fusion.

This syndrome might also occur in patients with a spectrum of unrelated diseases complicated by nerve damage. These include distal neuropathies caused by toxins (ethanol, drug related) and infection (leprosy), as well as diseases of the spinal cord and nerve roots (tabes dorsalis, trauma, syringomyelia) and a number of other conditions (Parkinson's disease, HIV, sarcoidosis, rheumatoid disease, and psoriasis).

There is no singular cause for the development of the Charcot foot, but there are factors that predispose to its development, as well as a number of likely precipitating events. The current belief is that once the disease is triggered in a susceptible individual, it is mediated through a process of uncontrolled inflammation in the foot. This inflammation leads to osteolysis and is indirectly responsible for the progressive fracture and disintegration of joint characteristic of its presentation.[4] The evidence to support this hypothesis is largely circumstantial. A neurally mediated vascular reflex leading to increased peripheral blood flow and active bone resorption has been proposed as an etiological factor in the development of bone and joint destruction in neuropathic patients. However, the relationship between increased blood flow to bone and active bone resorption has not been conclusively defined.

## DOES TRAUMA INITIATE NEUROARTHROPATHY?

### Posttraumatic Uncontrolled Inflammation

After fracture of a bone, release of proinflammatory cytokines including tumor necrosis factor-$\alpha$ and interleukin-1$\beta$ leads to increased expression of the polypeptide receptor activator of nuclear factor-k$\beta$ ligand (RANKL) from any of a number of local cell types. RANKL triggers the synthesis of the nuclear transcription factor nuclear factor-k$\beta$ (NF-k$\beta$), and this in turn stimulates the maturation of osteoclasts from osteoclast precursor cells. At the same time, NF-k$\beta$ stimulates the production of the glycopeptide osteoprotegerin (OPG) from osteoblasts. This "decoy receptor" acts as an effective antagonist of RANKL.[5] The fracture will also be associated with pain, and this leads to splinting of the bone, and the rise in proinflammatory cytokines is usually relatively shortlived. In the person who develops an acute Charcot foot, however, the loss of pain sensation allows for uninterrupted ambulation, with repetitive trauma. It has been suggested that this results in continual production of proinflammatory cytokines,

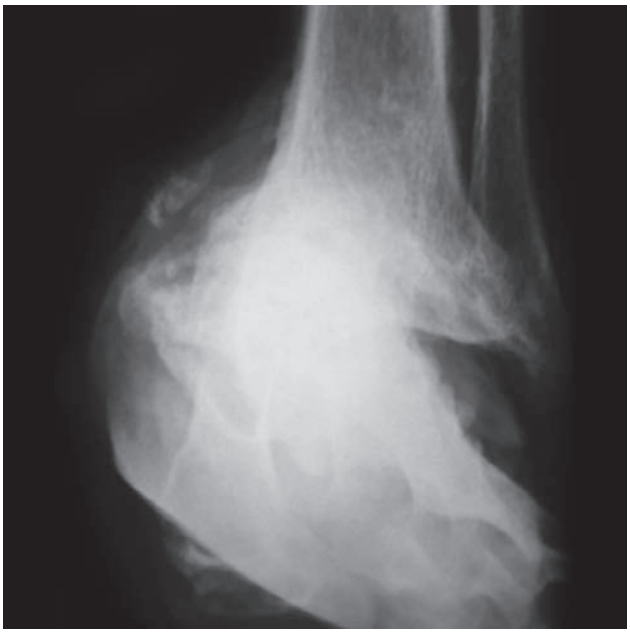

**Fig. 1A:** Preoperative anteroposterior ankle

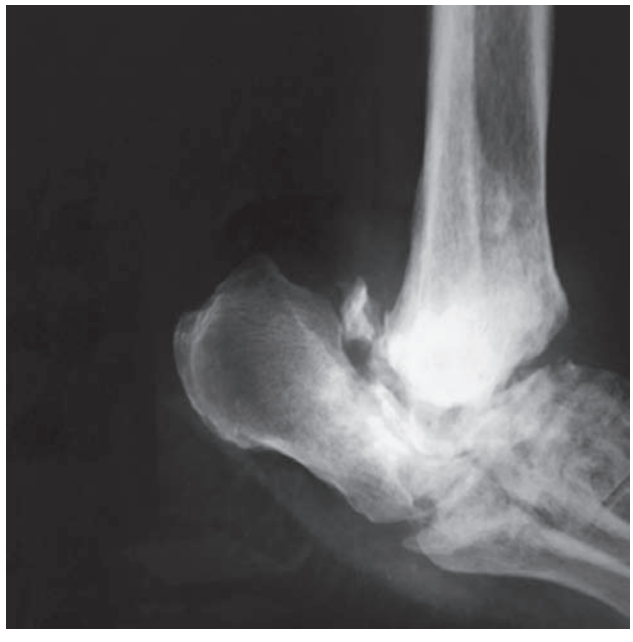

**Fig. 1B:** Preoperative lateral

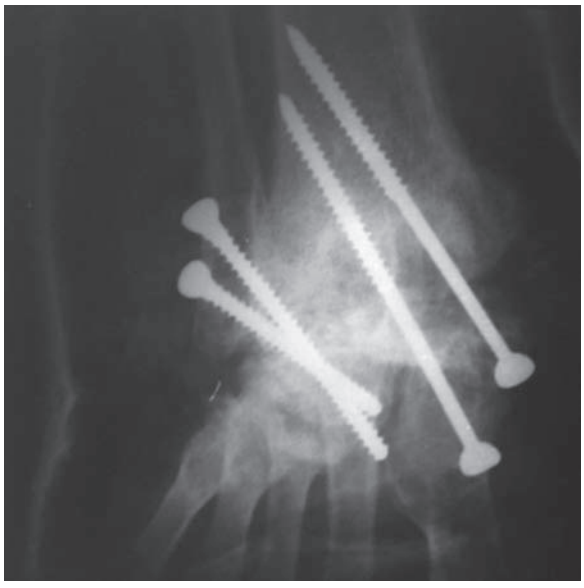

**Fig. 1C:** Postoperative in plaster-of-paris cast

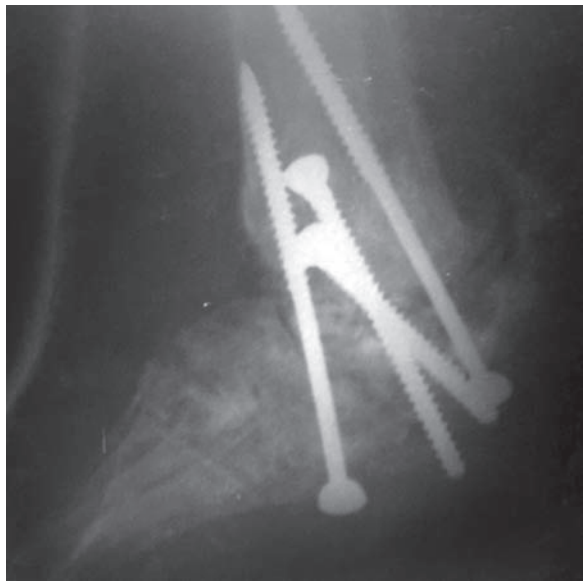

**Fig. 1D:** Postoperative in plaster-of-paris cast

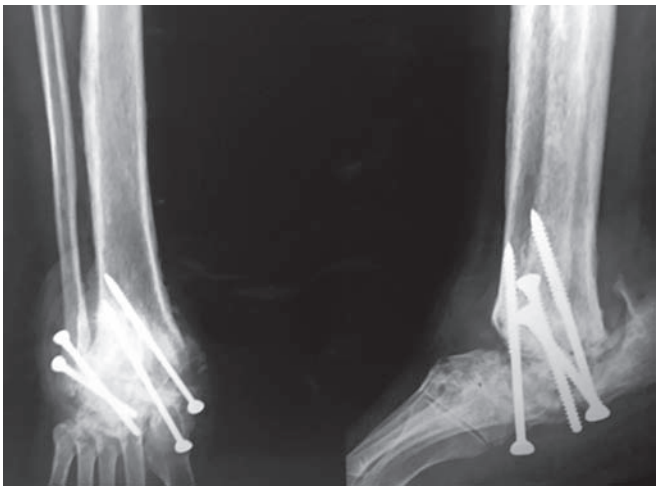

**Fig. 1E:** One year postoperative follow-up

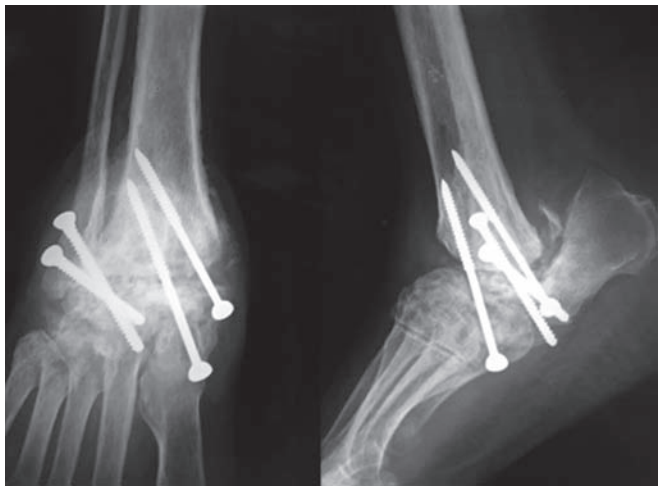

**Fig. 1F:** Two years' postoperative follow-up

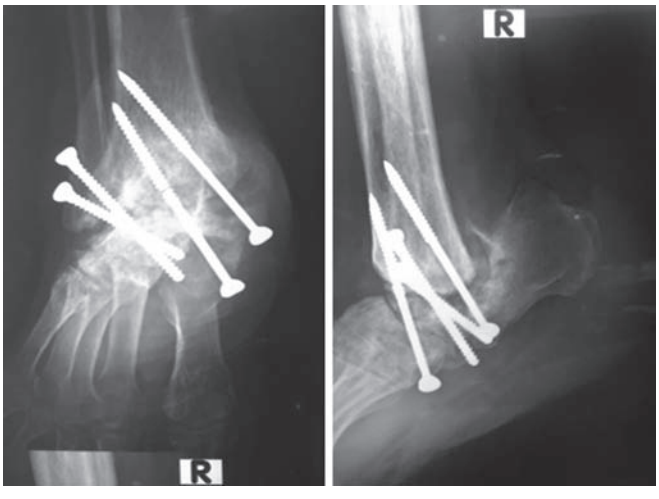


**Fig. 1G:** Three years' postoperative follow-up

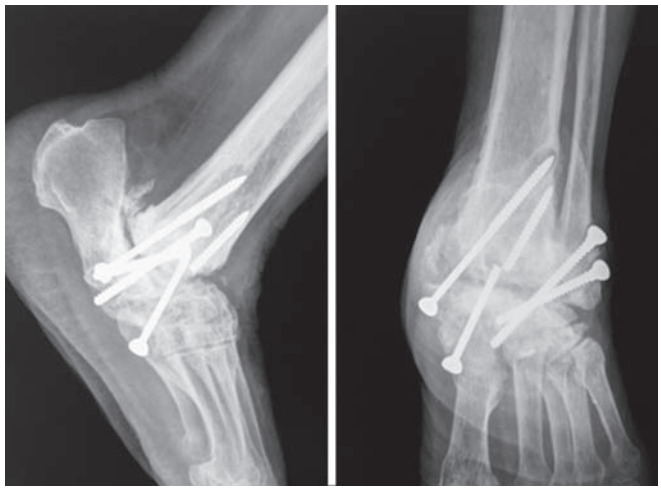

**Fig. 1H:** Seven years' follow-up

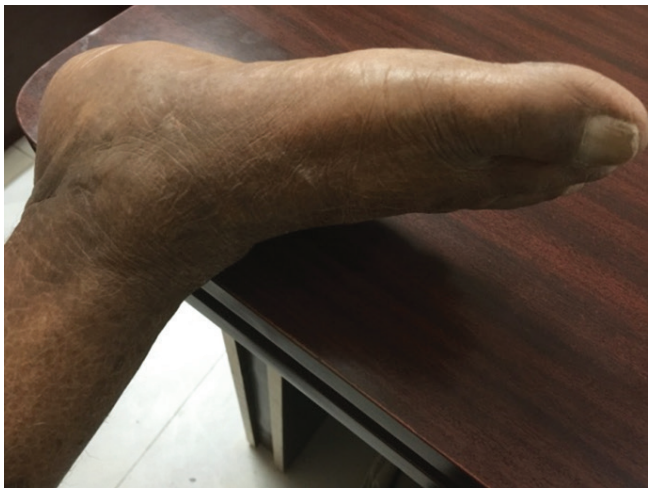

**Fig. 1I:** Seven years' follow-up clinical photograph with stable ankle

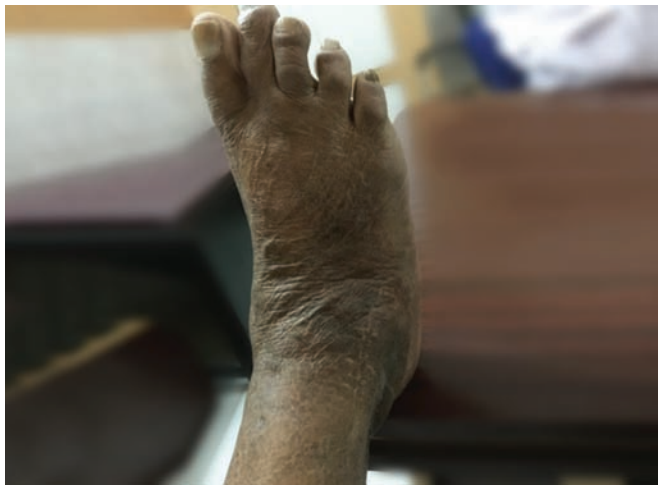

**Fig. 1J:** Seven years' follow-up clinical photograph with stable ankle

RANKL, NF-kβ, and osteoclasts, which in turn leads to continuing local osteolysis.[4] This has subsequently been shown by an increase in proinflammatory phenotypes of monocytes in those with active Charcot foot when compared with diabetic control subjects.[6]

Osteoclasts generated *in vitro* in the presence of macrophage colony-stimulating factor and RANKL from patients with active Charcot neuroarthropathy have been shown to be more aggressive and exhibit an increase in their resorptive activity compared with control subjects. However, these changes are only partially inhibited by OPG, indicating that other cytokines may also be important.[7]

## CONCLUSION

Many patients recall that the onset of the condition was precipitated by trauma that is often minor in nature.[8] Other cases may be triggered by different causes of local inflammation, including previous ulceration, infection, or recent foot surgery. In this respect, the occurrence of an acute Charcot foot as a complication of osteomyelitis is increasingly recognized in people with diabetes. Very occasionally, the onset of an acute Charcot foot may follow successful revascularization.

This particular patient is unique in its presentation where no relevant associated pathology could be attributed but for the initial injury. This gives credence to the theory that posttraumatic uncontrolled inflammation can initiate and continue the process till the final stage of destructive hypertrophic osteoarthropathy. Hence, this condition has aptly been labeled idiopathic. A treatment protocol with anti-TNF alpha or methotrexate is hypothesized and recommended.

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