Ankle Reconstruction in Charcot’s Neuroarthropathy: Challenges and Solutions

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ABSTRACT

Ankle injuries in the presence of neuropathy have potential to cause deformity in the ankle, which can in turn cause ulcerations and infections leading to significant morbidity including loss of limb or even life. These injuries although devastating can start off innocuously and may not be immediately apparent to the patient because of lack of pain. This can lead to a late presentation. Often, the main concern is swelling, for which the patient seeks attention. And even when medical attention is sought, unless the physician is vigilant, the condition can potentially be missed or misdiagnosed as deep vein thrombosis or cellulitis. A sprain or even a fracture in the ankle is not suspected as the patient retains the ability to walk. This condition and its true nature further escapes detection if, as often is the case, a nonweight-bearing radiograph is obtained. Radiographs obtained without the weight-bearing or other stress modality may not reveal the instability at the ankle joint due to ligament damage. A thorough clinical evaluation including assessment for neuropathy and weight-bearing radiographs are necessary for diagnosis. Stable fractures and ankle sprains can be managed conservatively in a cast or boot. Unstable ankle following ligamentous disruption and/or fractures in the vicinity of the ankle needs internal or external stabilization. The stabilization in the presence of neuropathy should be stronger and the protection from weight bearing last longer and twice that used for patients with similar condition but without neuropathy.

Keywords: Ankle, Charcot, Diabetes, Hindfoot nailing, Peripheral neuropathy.

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INTRODUCTION

Charcot neuroarthropathy (CN) is a destructive process that can occur in the ankle joint in patients with peripheral neuropathy. Trauma to the ligaments and bones either acute or repetitive can set this process off in the ankle. There is potential for complications, such as severe deformity in the ankle and hind foot followed by ulceration and limb amputation. Therefore, early recognition and appropriate management is critical. Stabilization of the ankle and maintaining or restoring anatomical alignment is the goal of management of this condition as the CN process burns itself out. This article reviews challenges and offers solutions in ankle reconstruction in patients with CN at the ankle joint.

ETIOLOGY

The pathogenesis of CN is not yet well understood, and there is no single theory for the abnormal processes that cause CN. It is likely that multiple abnormal mechanisms are responsible. The neurotraumatic theory suggests that CN is an overuse injury in which insensate joints are subjected to either repetitive microtrauma or a single traumatic event that leads to typical Charcot changes. Abnormal sensation prevents the affected individual from adopting normal protective mechanisms, specifically offloading and activity modification, and from seeking medical attention. This theory has been supported by several studies using experimental animals with insensate limbs. Investigators denervated joints through division of the spinal cord or peripheral nerves and then subjected the animals to trauma or repetitive overuse. This experimental model often resulted in the changes seen in CN. However, reports of Charcot changes in nonweight-bearing joints, such as the shoulder, as well as in the hips of bedridden patients, have cast doubt on this theory as an explanation for the primary cause of CN. The neurovascular theory proposes that autonomic dysfunction leads to increased blood flow via arteriovenous shunting, resulting in bone resorption and weakening. Bone turnover markers have been found to be elevated in acute CN compared with controls, whereas bone formation markers have been found to be unchanged, indicating increased osteoclastic activity. Several studies also have shown an increase in bone resorption markers. Bone density analysis confirms the presence of osteopenia and indicates an increased risk for neuropathic fracture.

More recent theories implicate the role of inflammatory cytokines, such as tumor necrosis factor-α and...
interleukin-1 in the pathogenesis of CN. On the molecular level, these factors lead to increased expression of nuclear transcription factor-κB, which in turn stimulates osteoclast formation.11,12

In 2006, Baumhauer et al13 confirmed the increased presence of osteoclasts, tumor necrosis factor-α, and interleukin-1 through examination of pathologic specimens and immunohistologic staining of surgical specimens from patients with CN.

CLINICAL PRESENTATION

Charcot neuroarthopathy can occur in patients with peripheral neuropathy due to a variety of disease processes, but it most frequently has been associated with diabetes mellitus. In persons with type 1 diabetes, CN most frequently presents in the 5th decade, after an average duration of diabetes of 20 to 24 years; in those with type 2 diabetes, CN typically presents in the 6th decade, after an average duration of diabetes of 5 to 9 years.14

Acute CN in the ankle manifests as a hot and swollen leg. The swelling and redness is prominent at the ankle joint itself, but can extend up the leg and distally into the foot. The distal pulses are bounding. A careful history may reveal an unrecognized traumatic event. Often the patient is able to bear weight on the involved extremity with minimal or no discomfort even in the presence of a fracture. Patients may present late. High index of suspicion must be maintained, especially in a younger patient with an ankle fracture if the patient does not have the discomfort or pain that can be expected from the injury sustained or the patient has been walking or presents late. As diabetes is not a commonly expected comorbidity at a younger age, there is a potential for patients in this age group to be mismanaged, if a history of diabetes is not actively sought and presence of peripheral neuropathy not checked. Fixation of ankle fracture undertaken in a routine way in such a situation may not provide adequate stabilization.

Skin temperature of the affected leg has been found to be an average of 3.3°C higher than that of the unaffected extremity.15 The progression of CN most often follows a predictable clinical and radiographic pattern, and the widely recognized Eichenholtz classification continues to help guide the practitioner through the treatment process.16

The clinician may often suspect either deep infection or cellulitis, given the marked swelling and erythema. A simple method of distinguishing the dependent rubor of CN from infection involves elevating the leg and watching for a decrease in erythema.14

Patients presenting with late deformity may have ulcerations due to soft tissue breakdown on bony prominences, such as over the medial malleolus or lateral malleolus and shortening of the limb, depending on the direction of instability and fracture or collapse and resorption of the bone.

INVESTIGATIONS

Radiographs are essential to check for fractures and evaluate bone destruction and deformity. They should be done with patient bearing weight on the extremity when possible. Radiographs taken with the patient not bearing weight may not reveal underlying instability. Often, three views of both ankles, feet, and alignment views are sufficient. There is no single test that can differentiate this condition from cellulitis in the absence of a fracture or deformity. The diagnosis is based on collaborative information from clinical evaluations, presence or absence of systemic signs of infection, such as fever, leukocytosis, inflammatory markers, and blood glucose levels or insulin requirement. Other investigations, such as bone scans and magnetic resonance imaging (MRI) are not essential for diagnosis or management and should be used judiciously.

MANAGEMENT

Management is based on a variety of factors, including presence and location of fractures in the bones that make the ankle joint, displacement of fractures, phase of the disease process, bone destruction, presence of ulcers, infection, deformity, and comorbidities. Treatment should be guided by specific and realistic goals, depending on the severity of the disease and the patient’s functional capacity. This can vary from initial treatment in an offloading device to internal fixation or reconstruction in stages or amputation. Most treatments have been guided by level IV studies.17 There are no prospective randomized trials that have evaluated surgical interventions. The goal of management is to ensure that the ankle joint area remains aligned anatomically and biomechanically, and at the end of the Charcot process, the result is a stable ankle without deformity or, if this is not achievable, a braceable deformity.

A nondisplaced fracture of the ankle, such as in the lateral malleolus can be managed in a total contact cast, but needs careful follow-up with weekly radiographs and cast changes as there is risk of displacement and reduction of swelling, causing cast sores. Prolonged immobilization may be necessary as healing times in diabetics have been shown to be longer than in nondiabetic population.
Unstable fractures and deformities at the ankle require surgical stabilization. Fractures in the lateral or medial or posterior malleolus or ligamentous disruption in the deltoid ligament and/or the syndesmosis area can render the ankle joint unstable. Surgical stabilization of an unstable ankle joint should be robust, to minimize risk of displacement and deformity. This is often achieved by either inserting multiple screws across the syndesmosis to gain additional purchase in the tibial metaphysis. This strategy is recommended even when syndesmosis ligaments are intact. Routine fixation of lateral malleolus fracture may not be sufficient. There is a risk of loss of fixation and deformity (Figs 1 to 3). Patients are often unable to comply with nonweight-bearing instructions; therefore, implant selection and surgical strategy should be tailored to realistic expectations that the patients can comply with. Lack of compliance is possibly due to cerebral involvement similar to neurological involvement in the leg. The author believes that “cerebral neuropathy” leads to cognitive deficits, which make patients with neuropathy noncompliant with nonweight-bearing instructions. In the absence of robust fixation, therefore, there is risk of displacement (Figs 4 and 5) and need for subsequent surgical intervention if the displacement continues and results in deformity. If patients are not able to comply or can not be expected to comply due to other comorbidities, such as obesity or poor coordination or balance, more robust fixation that extends into adjacent joints is recommended. This is achieved by intramedullary stabilization of the hind foot. Schneekloth et al\textsuperscript{17} provided a systematic review of studies published from 2009 to 2014 and to review the indications for surgery. A Medline search was performed, and a systematic review of studies discussing the surgical management of CN was undertaken. They found that arthrodesis, specifically tibiotalocalcaneal, seems to be gaining popularity as a surgical treatment option for CN. However, no randomized, prospective, multicenter trials have yet been published regarding this topic, and the proper timing of surgery remains undefined. The goal of treatment, whether nonoperative or operative, is to achieve a plantigrade, stable foot that remains ulcer free. If the CN deformity involves more proximal anatomic regions (ankle and hind foot), the need for surgical intervention becomes more likely.

In the presence of an open ulcer or significant shortening, such as in a long-standing dislocation at the ankle joint, external fixation can be used to get the ulcer healed. Additionally, gradual distraction in an Ilizarov-type circular frame can restore limb length. The fixator can be
Fig. 4: An ankle fracture fixed using routine fixation, showing lateral displacement of talus in the ankle mortise with widening of the medial gutter 2 weeks after fixation.

Fig. 5: The fixation was revised with more robust “super construct” using multiple “syndesmotic” screws across from fibula into tibia showing no displacement at 12 weeks in the ankle joint.

Fig. 6: Late presentation of ankle fracture and dislocation of patient with diabetes and CN, showing an ulcer over the medial malleolus.

Fig. 7 to 9: Anteroposterior, lateral, and oblique weight-bearing radiographic views showing the lateral dislocation of the hind foot and resultant shortening.

used as definitive method of stabilization. Alternatively, to minimize risk of pin site infection, the frame can be used only in the initial stage to restore length and heal ulcers followed by internal stabilization, such as with an intramedullary tibiotalocalcaneal arthrodesis nail or as a definitive method of stabilization (Figs 6 to 13).

Siebachmeyer et al\textsuperscript{18} reported the outcomes of 20 patients with Charcot neuroarthropathy who underwent correction of deformities of the ankle and hind foot using retrograde intramedullary nail arthrodesis. At a mean follow-up of 26 months, limb salvage was achieved in all patients, and 12 patients (80\%) with ulceration achieved healing, and all but one patient regained independent mobilization.

DeVries et al\textsuperscript{19} compared the use of internal vs external fixation of CN in the ankle. They surgically stabilized 52 deformities with an intramedullary arthrodesis nail, 7 of which had the addition of a circular external fixator. The endpoint of their study was either major amputation or...
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CONCLUSION

Ankle injuries in the presence of neuropathy if missed or not managed appropriately can lead to deformities. Patients with long-standing diabetes should be educated to seek attention in the presence of swelling and/or erythema in the leg and foot. Physicians must check for presence of neuropathy and obtain weight-bearing radiographs whenever possible. An injured or unstable ankle should be stabilized and limb protected from any deforming weight-bearing forces, throughout the time the ankle is at risk and until the Charcot neuroarthropathy process resolves.

It is important to be vigilant and check for neuropathy in patients presenting with swelling in the leg and foot with or without a history of injury to the ankle, even when they are able to ambulate without pain and consider Charcot neuroarthropathy in the differential diagnosis.

A major amputation was performed in 10 of 45 patients (22.2%) in the intramedullary nail cohort and 2 of 7 patients (28.6%) in the combined intramedullary nail and external fixator group. The addition of the circular external fixation device did not affect the overall limb salvation rate or complication rate.
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