A Case Report: Chronic Foot Deformities Secondary to Neurological Disorders

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Abstract

Congenital neuromuscular disorders pose a far more detrimental problem to patients not diagnosed at an early stage in their development. Therefore, a pre-emptive systematic assessment of the child will allow the diagnosing clinician to assemble a medical team specifically designed to better fit the child’s condition and better their prognosis in life. Some congenital presentations may be subtle at first but may have a significant impact on the patient’s long term goals such as ambulation. Deformities of the lower extremity are at the front line of these complications. Neurological evaluations should be sought out in addition to biomechanical evaluations in order to identify these issues before the physical manifestations arise. The increase in uneven pressure distribution on the foot as a result of such deformities puts patients at higher risk for ulcers. Having an open wound is one of the major risk factors towards developing osteomyelitis or bone infection. In this review, the study patient suffered from Spina bifida and has, since childhood, presented with deformational complications of the foot that developed into a chain of events progressing from multiple lacerations to ulcer and inevitably to osteomyelitis. The prognosis of bone or even limb salvage with osteomyelitis is very poor unless detected early on. In retrospect, to identify the association between neurological deficit and deformity of the foot early on may provide a better prognosis for the patient during their lifespan.

Introduction

Spina bifida arises as a congenital disorder resulting in the incomplete closure of the embryonic neural tube (known as a neural tube defect) [1,2]. Beginning in embryogenesis and continuing throughout adulthood, neural tube defects comprise the majority of chronic disabilities of an estimated 70,000 to 100,000 individuals in the United States [3]. Literature has shown that Spina bifida may be categorized into various types: Occult Spinal Dysraphism (OSD), Spina Bifida Occulta, Meningocele and Spina Bifida Cystica also known as myelomeningocele [2,4]. These may differ in the absence or presence of herniation of the spinal cord components, respectively [4]. Being one of the most common neural tube defects, Spina bifida results in myelodysplasia of the spinal cord sensory components as well as acquired deformities in the lower extremity (clubfoot, equinus, vertical talus, calcaneovalgus, hindfoot/ankle valgus and cavovarus foot-type) [1].

When considering a cavus deformity, sensory impairments such as peripheral neuropathy commonly pose a problem of recurrent ulcerations [1]. Often, individuals are able to carry out routine activity; however, in others the complications may introduce a major impediment to their everyday lifestyle depending on the severity and level of the spinal cord lesion at hand. The etiology of ulceration may be explained both neurologically and biomechanically via the absence of sensory function (due to peripheral neuropathy) in the foot as well as an imbalance between muscle actions of the lower extremity. The absence of sensation along the lateral plantar aspect of the foot deems the individual unaware of integumentary compromise and thus prone to infection arising from pressure sores. Biomechanically, spina bifida causes problems such as knee flexion/extension contractures as well as knee valgus deformities [1]. In a low-level lumbar involvement, constant contraction of the tibial is posterior muscle and/or absence of the peroneus muscles may lead to an imbalance in plantar flexion/inversion and dorsiflexion/eversion, thus resulting in clubfoot [5]. Consequently, upon ambulation constant pressure and shearing forces to the lateral column (oblique axis) of the foot disrupt the normal makeup of the plantar skin leading to potential breakdown [5]. In the most severe cases, buildup of infection in the wound may progress to osteomyelitis and thus amputation of the lower extremity may be required [6].
Case Presentation

A 44-year old Caucasian female presented to the Barry University Foot & Ankle Institute with an extensive history of non-healing ulcers. This patient was born with Spina bifida. As mentioned, the patient was suffering from the consequences of Spina bifida such as peripheral neuropathy and equinovarus deformity of the left foot which lead to complicated ulcerations. She went through extensive visits to various physicians as a child as well as into her late teens. Her first injury took place in childhood when she suffered a laceration injury to her left foot while taking swimming lessons. Following this injury, later that same year, she went through her first foot surgery involving a bilateral tendon transfer to correct the foot deformity. One year later, she underwent a second foot surgery only at this time consisting of a tendon transfer along with a triple arthrodesis to the same foot. The postoperative results of this second surgery were dissatisfying since the left foot still remained in the varus position; hence the physician took her back to the surgery to perform a different procedure involving a calcaneal osteotomy with internal fixation to the left foot. At this point in life, the patient was stable and did not experience any other problems from age eight to fifteen (Figure 1 and 2).

At age fifteen, she suffered another ulcerating injury to her left foot while at a local pool. This wound was treated extensively by her physician with the suspicion of osteomyelitis to the foot. After the treatment, the ulcer remained closed for about nine years without any complications. At age twenty-four, the ulcer reopened again, and her physician performed another surgery consisting of a partial calcanectomy. This kept the wound closed for about four years. Unfortunately, at age twenty-nine, it reopened again. At this time, her physician proposed a total calcanectomy; however, she was not willing to compromise her ambulation. Hence in 2011, she decided to get a second opinion and came to the Barry University Foot & Ankle Institute (Figure 3-6).

NOTE: Please refer to figure 6 for a better understanding of her extensive medical history before her visit to the Barry University clinic.

Figure 1: Lateral X-ray of Left Foot Status-Post calcaneal osteotomy with internal fixation.

Figure 2: Lateral X-ray of Left Foot Status-Post calcaneal osteotomy with internal fixation. Note: osseous changes to plantar surface of the osteotomy site.

Figure 3: Clinical image of patient’s Left foot during her visit to our office. Note: multiple ulcerations with periwound erythema and 50%:50% necrotic/granular wound base.

Figure 4: MRI of patient’s Left foot status-post partial calcanectomy.

Figure 5: Lateral X-ray of patient’s Left foot upon first clinical visit. Note: destructive osseous changes to the calcaneus.

Figure 6: Please refer to Figure 6 for a better understanding of her extensive medical history before her visit to the Barry University clinic.
Medical management

The patient presented to the Barry University Foot and Ankle Institute with an open ulcer that displayed clinical evidence and suspicion of osteomyelitis. A plain radiograph was taken of her left foot which revealed chronic osseous changes. This urged her podiatric physician to admit her and request for an Infectious Disease consult. While being admitted, the patient underwent surgical debridement of soft tissue and a bone biopsy of the calcaneus which tested positive for osteomyelitis; as a result, IV antibiotic therapy was started for 8 weeks in conjunction with Negative Pressure Wound Therapy (NPWT). The patient was also prompted to be non-weight bearing on her left lower extremity.

The initial attempt to close the wound failed; however, for the next three years her podiatric physician continued with his plan to treat the infection with multiple soft tissue debridements, two regiments of NPWT, and another 8-week course of IV antibiotic therapy. A different approach was used to keep the patient non-weight bearing throughout her healing process. A number of offloading techniques including post-op shoes, a CAM walker with plastazote insoles, a Toad Anti-gravity brace, and crutches were tested; the CAM walker was found as the best fit for the patient’s needs (Figure 7 and 8).

Amputation of the left lower extremity and reconstructive surgery with external fixation were two treatment courses discussed with the patient; however, the patient stressed that neither of these options were a consideration for her. The last surgery consisted of resection of the remaining non-viable bone from the calcaneus while keeping the Achilles tendon intact which ultimately closed and healed the ulcer wound (Figure 9). In her six month follow-up visit, the patient’s wound remained closed, her radiographs were stable, and the podiatric physician discussed with the patient about planning an Achilles tendon transfer procedure in the future (Figure 10 and 11).

Discussion

Neuropathy-associated ulcerations, specifically those associated
with Spina bifida, Cerebral Palsy, spinal cord lesions, peripheral nerve injuries, and leprosy are usually slow healing and unresponsive to treatment [7,8]. Matters are further complicated by Osteomyelitis, which may develop secondary to local spreading of the infection into the underlying bone resulting in inflammation and bone destruction [7,9,10]. An insensitive foot is at higher risk of developing ulceration, which in itself is the most significant risk factor leading to osteomyelitis [11]. A severely dislocated and unstable foot or ankle, as observed in cases such as Spina bifida and Cerebral Palsy, may also be a predisposing factor [11].

There have been numerous individualized studies published on the incidences of congenital disorders, foot deformity, neuropathy, ulceration and resultant osteomyelitis as separate entities; however, none have attempted to establish the connection between patients born with these congenital disorders, foot deformities, ulceration and thus osteomyelitis as a whole chain of events (one factor inevitably leading to the other). Furthermore, the idea of diagnosing structural foot deformities at a very early age rather than upon the first symptomatic visit should be mentioned here. More often than not, it is the podiatrist or other primary care physician who gets the chance to diagnose these future problems in patients before they arise. Woods et al. mentions in their article that Podiatric physicians should be part of the team that treats patients with neurological disorders since more often than not neurological disorders have subclinical podiatric manifestations (2004). No matter who diagnoses the patients, if the Podiatric physician is consulted early enough, the progression of the deformity can be subsided before it worsens. Contrary to this understanding, there is always still the notion that the physician is never fully able to diagnose the problem until it manifests itself because the patient’s first visit is usually a symptomatic one. However, keeping in mind the progressive sequence of these events may help a Podiatrist diagnose the problem earlier in infancy through routine examination and help to take pre-emptive measures so the patient may live a normal adult life.

Peggy Lin “et al.” [4] presented in their article a similar patient-base of a 57-year old Caucasian female with lateral foot ulceration due to a congenital disorder. The risk of ulceration was proportional to the combination of risk factors present ranging from peripheral neuropathy alone (increased by a factor of 1.7) to peripheral neuropathy and foot deformity (increased by a factor of 12). In addition, it was mentioned that the structural deformity of the foot causes increased pressures on bony prominences on the plantar surface, thus rendering the patient more prone to ulceration. Given these abnormal pressures, it was concluded that cutaneous lesions may be initial markers of spinal abnormalities [4]. However, to go one step further beyond that idea and say that the initial marker of concern is a child born with a structural deformity of the foot, even before the age of ambulation and thus cutaneous buildup of lesions, is ideal.

James A Birk “et al.” [12] mentions the relationship between limited motion due to factors such as motor neuropathy of the peroneal and tibial nerve distribution. Respectively, weakness in these distributions may lead to an equinus foot or calcaneus foot so often seen in congenital diseases such as Spina bifida and Cerebral palsy. Cerebral Palsy is a non-progressive motor disability encompassing a variety of symptoms that hinder a child’s normal lower extremity functioning, particularly their posture and movements. This condition from a CNS lesion of the immature brain believed to be of myelin-deficient origin occurs most often around the perinatal age; as a result, the child experiences hypotonia and spasticity. Spasticity and dystonia among other conditions in a child with cerebral palsy are permanent; however, they are not unchanging. The resulting equinus from the acquired spasticity and hypotonia of affected muscle groups are salvageable. With a dynamic team consisting of therapists, orthopedists, podiatrists, pediatricians, etc. these conditions are manageable. Early recognition followed by orthotic bracing, physical therapy, and exercises that incorporate full range of motion for lower extremity joints in particular are cardinal steps for the cerebral palsy patient.

In addition, structural deformities such as a varus foot-type show a certain degree of increased plantar pressures based on the degree of compensation: a compensated varus deformity may abnormally pronate to its end range of motion and thus medial ulcerations are more prominent. On the other hand, an uncompensated varus deformity may show ulcerative tendencies on the lateral column [12]. Calcaneovalgus deformity usually results from the unopposed action of dorsiflexors and evertors of the ankle joint; most cases result from spasticity of the muscles involved secondary to neurological impairment at the L4 – L5 level [5]. Biomechanical analysis of these muscle groups and their respective actions during the gait cycle leads us to understand how such deformities put a patient with Spina bifida or Cerebral palsy at higher risk of progressing to pressure ulcers. The effect of an anterior tibial is tendon transfer serves to facilitate transitioning action of the anterior group muscles to the forefoot thus improving ambulation as well as decreasing shearing forces at the heel [13]. These foot-types may very well be seen in an infant with Spina bifida, Cerebral palsy, or any other neuromuscular congenital disorder after routine biomechanical examination in the office. If observed during the early stages of infancy, further steps may be taken to accommodate for that specific foot-type such as orthotics or surgical correction. With this goal in mind, the Podiatrist or any other physician may significantly decrease the chances of ulceration and thus osteomyelitis as the child grows to adulthood.

It should be noted that although this progressive thought process should be considered ideal in the eyes of a Podiatrist or practicing physician, it must be understood that not all patients are ideal in their own respect. Thus osteomyelitis may inevitably present; at this point, early detection and treatment should be indicated. Early detection of osteomyelitis can be treated empirically with broad-spectrum antibiotics followed by a sensitivity-based course tailored to the patient and orchestrated by a multidisciplinary team [4,10]. Conversely, however, such therapy generally fails when in presence of chronic osteomyelitis. At this point surgical intervention is deemed necessary in order to attain a viable vascularized environment and eliminate dead bone [7,9,10].

In patients with adequate blood supply and oxygen tension at the infected site, debridement of bone followed by four to six weeks course of antibiotics should be adequate treatment for osteomyelitis. However, in those patients with poor vascularity and oxygen tension, the wound fails to heal and amputation of the infected foot is eventually necessary [9].

**Conclusion**

Peripheral neuropathy in individuals with spina bifida can be perilous. These individuals are at high-risk for pedal complications. Spina bifida and many other neurological conditions need be accessed...
by a group of multispecialty physicians including a Podiatrist for a thorough examination as a preventative measure. Many of these pedal problems can be managed successfully without reaching to a limb threatening circumstances.

References