Osteomyelitis in a Patient with Split-Hand/Foot Malformation

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Statement of Purpose

To increase awareness of types of ectrodactyly and associated pathology. Few case reports in the literature cite long term sequelae of a congenital pedal deformity and their ability to be effectively accommodated in shoegear. A review of the literature and treatment options will be discussed in concomitant with a case report.

Introduction

Electrodactyly has been synonymous with many terms in literature such as split-hand/split-foot deformity (SHSFD) or lobster claw (1). The earliest description of electrodactyly dates back to the 1770s in South Africa (2). This rare congenital abnormality occurs when the central portion of the apical ectodermal ridge fails to develop during limb bud formation. The ultimate result is an absence of one or more of the central rays of the foot (3). With lack of a central ray, the primary characteristic of electrodactyly is produced: a conical deficit extending proximally towards the tarsus (2). In addition, patients presenting with electrodactyly may develop a multitude of other deformities, such as genitourinary malformations, cleft lip and palate, fibular hypoplasia, syndactyly, polydactyly, triphalangeal thumb and deafness (2,3).

There are three forms of inheritance for split-hand/split-foot deformity (4). In its most common form, SHSFD is acquired through a pattern of autosomal dominant inheritance at one of six loci, producing a bilateral cleft foot (3,4). Electrodactyly has been known to occur due to mutations of TP63 gene via translocations, inversions, deletions and duplications (1,6). In addition, literature has shown that there is a predictable course of inheritance, known as anticipation. With anticipation, genetic disorders present themselves in a progressive manner through successive generations. Lastly, inheritance can occur through single gene autosomal recessive or X-linked recessive patterns (4,6).

Blauth and Borisch developed a classification scheme for electrodactyly of the foot. Type I displays five normal metatarsals with partial aplasia of phalanges 2-5. In Type II, five metatarsals are present but they are atypical, displaying hypoplasia and/or synostosis. With Type III electrodactyly, the second or third metatarsal is absent. In Type IV, two metatarsals are missing; this may be any combination of the second, third or fourth metatarsals. Type V electrodactyly, also know as a “lobster claw” deformity, displays the absence of the second, third and fourth rays entirely. Type VI presents with only the fifth metatarsal and the fifth digit, known as a monodactylous foot (5). Any of these deformities will lead to altered biomechanics with predisposition to pain and ulceration. Treatment encompasses conservative and surgical care prevent the pain and breakdown (4,8).

There are many case studies published on treatment of electrodactyly with long-term follow up. Each patient must be treated uniquely due to the heterogeneous nature of the deformity (3,6). Ultimately, the goals are to: 1) achieve function in an accommodative shoe and 2) have a socially acceptable appearance. In familial SHSFD, there is value to understanding how the disease was treated in previous family members (6). Currently, there is limited high-level evidence on the treatment of pedal electrodactyly. The following is multi-year follow-up of a patient with an atypical case of electrodactyly and conocominant rheumatoid arthritis. Electrodactyly is seen in approximately 1 in 25,000 births (7). Blauth and Borisch have published a classification of this disorder, as described in the introduction (Figure 3). Ultimately, this should allow for peer reviewed standard of care for the deformity. Unfortunately, the classification falls short due electrodactyly’s heterogenous and complex presentation. Due to its uncommon nature and variable presentation, a standard treatment regimen has not been established. It is important to develop treatment protocol because successful management gives patients a cosmetically pleasing appearance of the foot, the ability to fit into accommodative footwear, and prevents painful ambulation and ulceration (2,8).

Case Report

A 80-year-old male presented with bilateral pedal deformities (Figure 1. A,B,C) and a right foot ulceration present for weeks in duration. His foot became red, hot, swollen, and painful before he sought medical treatment from a local podiatrist. Patient denied any constitutional symptoms. Previous treatment had included wet to dry dressings with no improvement. The patient’s medical history included Rheumatoid arthritis, atrial fibrillation, cataracts, corneal dystrophy, chronic kidney disease, diabetes mellitus, and history of stroke. Patient admitted to being a former smoker of a 19 pack year history, quit in 1961.

On physical examination, the patient’s pulses were palpable but protective touch-pressure sensation was diminished to the plantar pedal skin bilateral. An ulceration was noted plantar to the second metatarsal head with surrounding erythema and edema. Radiographs were suspicious for osteomyelitis of the second metatarsal and deep wound swab cultures confirmed soft tissue infection for methicillin sensitive staphylococcus aureus. Conservative treatment was exhausted with a six week course of culture specific intravenous antibiotics. The patient’s ulceration failed to improve and he was then treated with a partial second and third ray amputation (Figure 2. A). The patient’s immediate post-operative course was uncomplicated and he subsequently ambulated in custom diabetic shoes without incident.

However, two years later the patient returned with an ulceration plantar to this first metatarsal head. The patient’s first metatarsal head and rudimentary digit had quadrupled in size (Figure 2 B,C). The surrounding area was fluctuant with erythema, calor, and pain on palpation noted. An magnetic resonance image revealed an abscess and suspected osteomyelitis of the first metatarsal. The patient then underwent a partial first ray amputation and tolerated the procedure well without complication. The patient then ambulated in a new pair of custom diabetic shoes after healing post-operatively.He did not experience any pedal complications until two years later.

In 2014, the now 84 year old male presented with an ulceration plantar to his fifth metatarsal head. Again, examination of the lesion was suspicious for infection, through the presence of erythema, edema, and calor (Figure 2. E). Radiographs were also suspicious osteomyelitis of the fifth metatarsal. The patient then underwent a partial fourth and fifth ray amputation to complete a transmetatarsal amputation. The metatarsal parabola was re-established. A two month follow-up revealed that the patient once again healed uneventfully without immediate complication.

Figure 1.

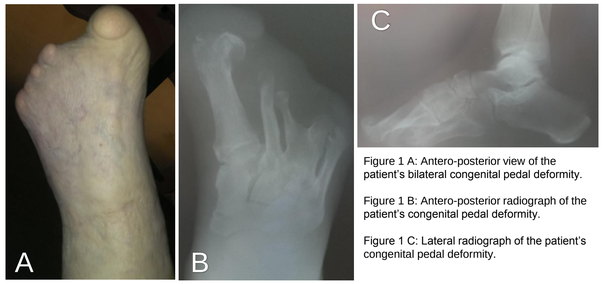

Figure 1 A: Antero-posterior view of the patient’s bilateral congenital pedal deformity.

Figure 1 B: Antero-posterior radiograph of the patient’s congenital pedal deformity.

Figure 1 C: Lateral radiograph of the patient’s congenital pedal deformity.

Case Report Continued

Figure 2.

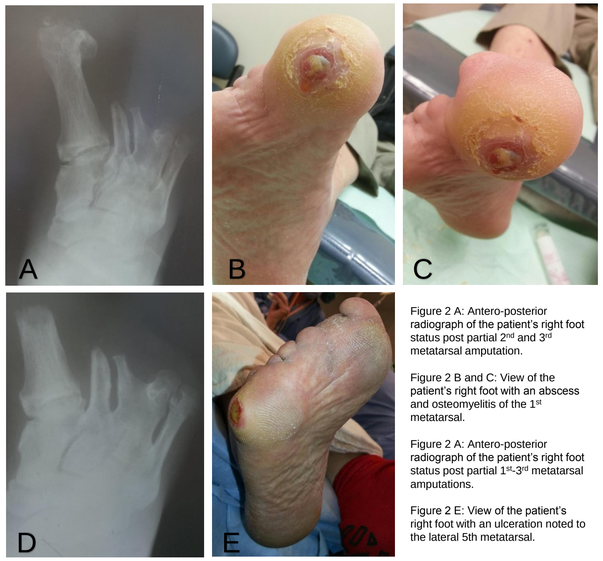

Figure 2 A: Antero-posterior radiograph of the patient’s right foot status post partial 2nd and 3rd metatarsal amputation.

Figure 2 B and C: View of the patient’s right foot with an abscess and osteomyelitis of the 1st metatarsal.

Figure 2 A: Antero-posterior radiograph of the patient’s right foot status post partial 1st-3rd metatarsal amputations.

Figure 2 E: View of the patient’s right foot with an ulceration noted to the lateral 5th metatarsal.

Figure 3.

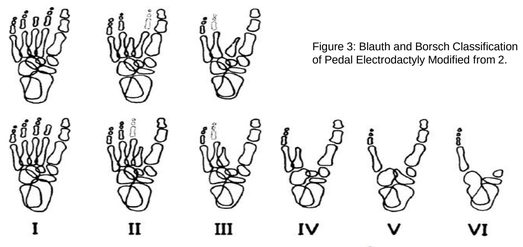

Figure 3: Blauth and Borsch Classification of Pedal Electrodactyly Modified from 2.

Discussion

The present case most closely resembles a combination of Blauth and Borisch classifications I, II and III as the following can be appreciated: 1. absence of phalanges, 2. a synostosis of rays three and four, and 3. under-development of rays two through five (Figure 3). This combination of deformities, combined with the patient’s rheumatoid arthritis, presented a complicated and difficult case. Publications suggest various osteotomies to decrease dorsiflexion of the metatarsals and decrease the width of the foot (2,4).

The patient originally presented with an ulcer beneath the second metatarsal along with signs of local infection. Radiographic analysis revealed an absence of a hallux and significant deficits in rays two through five. In addition a congenital union of rays three and four and large increase in metatarsal declination angle was appreciated (Figures 1. B,C). The patient had a history of plantar forefoot fat pad atrophy, a result of rheumatoid arthritis. Fat pad atrophy, along with the length and higher declination angle of the second metatarsal, predisposed the area to plantar ulceration. A partial second and third ray amputation were performed in order to decrease pressures at the distal portion of this digit. Following surgical resection, a functional plantigrade foot was achieved and the patient was satisfied.

Two years later, the patient presented with a new ulcer and chronic osteomyelitis at the distal first metatarsal (Figures 1. A,B,C). In order to heal the wound, surgical removal of infected bone and partial first ray amputation were performed (Figure 2. D). The procedure again normalized pressures across the plantar foot. Once more, the patient was satisfied and able to ambulate in accommodative shoe gear. Two years later, the patient presented with yet a third ulceration to the fifth digit (Figure 2. E). At this time, the plan is to perform partial 4th and 5th ray resection to decrease the width of the foot.

In review, the goal of surgery is to give the patient a functional plantigrade foot with the ability to fit into accommodative shoe gear. With the patient’s history rheumatoid arthritis and Orencia therapy, it is understood that they are immunocompromised. In such a patient population, there is a significant increase in risk of below knee amputations due to ulceration and infection. The procedures performed decreased risk of ulcer and ultimately salvaged a limb. However, a prophylactic TMA, TAL and dorslifexory osteotomy of the midfoot may have decreased the number of operations required to achieve the final result. The goal of the procedures could have created a proper metatarsal parabola, established a proper declination angle and allow for the tripod distribution of pressure across the plantar foot at an earlier point. It can also be noted that the classification scheme created by Blauth and Borisch does not have much relevance for planning of surgical treatment of electrodactyly.

In conclusion, excessive resection of one or more metatarsals will result in lack of purchase and increased load to adjacent metatarsals. Excessive load to adjacent metatarsals can result in transfer lesions to these areas. It is thus important to maintain a metatarsal parabola with metatarsal resection to avoid such a complication.

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