A Report of Amniotic Band Syndrome and Keratoderma with a Review of Constricting Band Syndromes

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Abstract
Amniotic bands are intrauterine fibrous cords that adhere to the growing fetus and cause mechanical trauma or vascular compromise. Associated defects range from hand creases to amputation, or even death in severe cases. We present a patient with a history of amniotic band syndrome with right leg amputation, constriction of the right fourth digit, and new-onset keratoderma involving the left plantar foot and bilateral palmar bands. This patient’s medical presentation emphasizes the importance of understanding the relationships between constricting band syndromes and palmoplantar keratoderma. We review previous literature documenting the association between amniotic band syndrome, pseudoainhum, and palmoplantar keratoderma.

Introduction
Although some authors have asserted that amniotic band lesions and pseudoainhum are distinct disease processes, pseudoainhum has been seen in patients with amniotic band syndrome. While both diseases involve constricting cutaneous lesions, their onset, causality, and relationships are dramatically different. We present a patient with a history of amniotic band syndrome with resulting right lower extremity amputation who presented to our clinic with new-onset keratoderma of the bilateral palms and left plantar foot. We also provide a literature review of palmoplantar keratodermas associated with amniotic band syndrome and pseudoainhum.

Case Presentation
A 26-year-old Caucasian female presented with roughening over the plantar portion of her left foot that had been progressing over the past several years. The patient had attempted to soften the area using foot soaks, a pumice stone, topical antifungals, and topical clobetasol over this same period, without noticeable relief. The patient denied any previous similar episodes. She also denied a family history of palmoplantar keratoderma or other cutaneous pathologies. Her medical history was positive for childhood eczema that had long since resolved, right lower extremity amputation, and a congenital constriction band deformity of the right fourth finger. She took no prescription medications but did take multivitamins. Physical exam showed focal hyperkeratotic plaques on the weight-bearing surface of the left foot with secondary fissuring (Figure 1). The hands were involved to a lesser degree, with thin scales over the left palm and digits bilaterally. The right hand showed a bulbous appearance of the fourth digit secondary to a focal constriction band distal to the interphalangeal joint (Figure 2). Differential diagnosis included acquired palmoplantar keratoderma with pseudoainhum, palmoplantar psoriasis, punctate palmoplantar keratoderma, irritant contact dermatitis, and atopic dermatitis. The patient was sent home with a working diagnosis of acquired palmoplantar keratoderma and was prescribed topical urea (40%) cream to apply to the foot BID at night with occlusion, and 10-minute bleach bath soaks two to three times per week. The patient failed to return for her next appointment, and despite repeated attempts to contact the patient, she was subsequently lost to follow-up.

Discussion
Constriction bands, which result in significant morbidity and mortality, are seen in amniotic band syndrome (ABS),1 ainhum, and pseudoainhum.2 ABS is thought to result from tears in the amnion that lead to fibrous adhesions to, and constrictions of, the developing fetus. The incidence is thought to be 1:10,000. Maternal risk factors for ABS include epidermolysis bullosa, connective tissue disorders, and prenatal abdominal trauma, such as amniocentesis. Ainhum, which is most prevalent in continental sub-Saharan Africa, is defined as idiopathic auto-amputation of a digit due to a constricting band. Pseudoainhum presents as a circumferential constriction band, usually around a digit, that has the potential to progress to auto-amputation. Unlike ainhum, pseudoainhum has no racial predilection.2 It has been described as secondary, sporadic, and a component of hereditary palmoplantar keratodermas, as well as in association with psoriasis.3,4 While many authors exclude cases of ABS auto-amputation from the pseudoainhum category, others use “pseudoainhum” and “ABS” interchangeably, with the rationale that the constriction bands are secondary to amniotic band syndrome.6

Theories on the pathogenesis of amniotic band syndrome are speculative and point to both intrinsic and extrinsic processes. The intrinsic theory states that deformities result from defective germ cells within the embryo. The extrinsic theory states that an early rupture of amnion results in decreased intra-amniotic fluid, which allows the mesoderm to contact the fetus, resulting in constricting bands.7

Theories on the pathogenesis of pseudoainhum are speculative and include infectious processes, fibrogenetic tendencies related to race, vascular abnormalities, and mechanical effects secondary to hyperkeratosis.2 Hyperkeratosis of the skin leads to formation of a circumferential groove around a digit. As the groove continues to deepen, the underlying vasculature becomes compromised, leading to a cessation of blood flow. The bone distal to the groove is eventually separated from the body, and auto-amputation occurs. Although the presentation of pseudoainhum does not always follow this pattern, hyperkeratosis is consistently present on histological exam.

Palmoplantar keratodermas (PPKs) appear clinically as abnormal thickening of the skin of the palms and soles.8 Hyperkeratosis is present on histological examination.5 PPKs can be inherited

Figure 1. Focal hyperkeratotic plaques of the left foot with secondary fissuring.

Figure 2. Bulbous deformation of the distal fourth digit secondary to a focal constricting band. Also note subtle scale over the bilateral digits and left palm.

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or occur sporadically and, in contrast to ABS, have often been seen in association with pseudoainhum and various inherited syndromes such as mal de Meleda and Vohwinkel syndrome. Mal de Meleda is an autosomal-recessive disease that appears early in life and is characterized by a triad of acral hyperkeratosis, malodorous hyperhidrosis, and nail abnormalities. Vohwinkel syndrome is an autosomal-dominant disease characterized by diffuse honeycombed palmoplantar keratoderma with associated sensorineural deafness, mental retardation, and alopecia. Vitamin A analogs, such as etretinate, have been shown to be effective in treating hereditary PPKs, as have urea-based topical formulations. Our patient's case does not fit into any of the known hereditary PPKs.

The diagnosis of ABS is primarily clinical and does not routinely require workup beyond prenatal ultrasonography showing amniotic bands and restricted motion of the fetus. Similarly, in most instances, pseudoainhum is a clinical diagnosis. Plain radiograph of the affected digit should be performed to assess the integrity of the bone distal to the constricting band, as complete resorption of the digit has been observed in some cases. A thorough workup must be initiated to determine if the pseudoainhum is an isolated occurrence, part of a larger hereditary syndrome, or secondary to another disease process. A family history is necessary to rule out hereditary PPKs. In equivocal cases, a biopsy can be performed.

When pseudoainhum arises from psoriasis, treating the underlying psoriasis, with oral retinoids and topical corticosteroids, has led to resolution in some cases. A combination of a topical calcineurin-inhibitor and UVB phototherapy was successful in a single case. Other reports show success with surgical release of the fibrotic band under local anesthesia. Amputation can be considered when all other treatment modalities are exhausted, as auto-amputation is inevitable and can be painful in some cases.

**Conclusion**

In summary, our patient, with a previous history of ABS, presented with new-onset PPK. While the co-occurrence of these distinct pathologies is likely coincidental, this presentation provided an opportunity to review the existing literature on ABS and constriction band syndromes. Additionally, it prompted the investigation of a seldom-explored relationship between ABS and PPK. Unfortunately, our patient, with no personal or family history of constriction band lesions or PPK, was lost to follow-up and further in-depth workup that may have been beneficial, including histology and genetic testing.

**References**