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A novel association of pseudoainhum and epidermolytic ichthyosis, successfully treated with full thickness skin graft after failed z-plasty repair

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Abstract

Pseudoainhum is a rare constriction band variant that may progress to spontaneous digital strangulation and auto-amputation. Although its association with palmoplantar keratodermas is well established, it has not been reported in conjunction with classic epidermolytic ichthyosis. We describe the first such case in a 25-year-old woman who presented with a painful constricting band of the fifth toe. We also describe her treatment course, which consisted of a failed z-plasty, the traditional therapeutic option for acute pseudoainhum, and report the success of subsequent full thickness skin graft, suggesting the benefit of this procedure as a therapeutic alternative for patients with pseudoainhum.

Keywords: epidermolytic ichthyosis, congenital icthyosiform erythroderma, epidermolytic hyperkeratosis, pseudoainhum

Introduction

Pseudoainhum (PA) is an annular constriction-band variant that most commonly affects the fifth digit, potentially leading to digital strangulation and auto-amputation. In contrast with ainhum, a spontaneous constriction-band syndrome affecting the fifth toe seen almost exclusively in black African males, PA has no known racial or sex predilection and may be a congenital or acquired phenomenon [1]. PA has been reported in association with a variety of diseases (**Table 1**), including inherited and acquired palmoplantar keratodermas (PPKs), [1-5]. Notably, a search of the PubMed database did not reveal any publications describing PA in a patient with epidermolytic ichthyosis (EI); we herein describe such a case. Furthermore, we outline the patient's course of management with failure of initial z-plasty repair (the traditional intervention for PA) and subsequent success of a full thickness skin graft, suggesting this procedure as an alternative therapeutic option for this condition.

Case Synopsis

A 25-year-old woman presented to the dermatology clinic with a one-day history of severe, progressive pain of the left fifth toe. She denied recent trauma or infection to the area and had no previous similar episodes. Her medical history was significant for a history of EI, with associated PPK consistent with a KRT1 defect. In keeping with the autosomal dominant mode of transmission typical of the diagnosis, her mother and several family members were also affected. Prior management included emollients and tazarotene (0.1%) gel.

In addition to classic flexural EI-type ridged hyperkeratosis and a PPK, physical examination revealed an exquisitely tender left fifth toe with a deeply grooved constriction band consistent with PA (**Figure 1**A). There was diminished light-touch sensation and sluggish capillary refill distal to the constriction band. Moderate edema and hyperemia were noted proximally, but there was no evidence

Table 1: *Diseases associated with type 2 pseudoainhum*

PA with associated keratoderma
Pachyonychia congenital
Vohwinkel syndrome
Kindler syndrome
Papillon-Lefevre syndrome
Mal de meleda
Psoriasis
Yaws (frambesia)
PA without associated keratoderma
Scar formation secondary to burns, frostbite, trauma
Diabetes mellitus
Porokeratosis of Mibelli and congenital linear porokeratosis
Systemic sclerosis
Raynaud disease
Frambesia

of infection. Digital range of motion was decreased secondary to pain with toe flexion and extension. A similar (but asymptomatic) band was noted on the ipsilateral third toe. The rapidly progressive pain and neuromuscular changes prompted immediate referral to the orthopedic surgery department for surgical intervention.

Excision of the left foot small toe PA and proximal interphalangeal joint exostectomy with z-plasty closure was accomplished. The constriction band encompassed approximately 50% of the total circumference of the digit and the band was excised in toto with a transverse elliptical incision. A sizable exostosis at the medial aspect of the proximal phalangeal condyle at the level of the constriction was excised as well. A two-flap z-plasty was then fashioned and closed with interrupted chromic gut suture (**Figure 1**B, C). Excellent capillary refill was noted distally at the completion of the procedure and there were no complications. Histopathologic examination of the excised tissue confirmed a PA with EHK-type change in the adjacent epidermis (**Figure 2**A-C).

The patient was seen in follow up six days postoperatively with significant improvement in her pain level and excellent distal perfusion. Further follow-up at six weeks revealed a fully healed digit and the patient had returned to her activities of daily living. At her eight-month follow up, however, she reported toe swelling and intermittent and partial recurrence of the constriction band (**Figure 3**A). At ten months after the initial procedure, progression of symptoms prompted a second surgical procedure.

A dorsal elliptical transverse skin excision incorporating the recurrent PA as well as a small portion of healthy appearing skin was completed and a 2x4cm full thickness skin graft was inset into the defect (**Figure 3**B, C). Histologic examination of the tissue confirmed recurrence of the PA and demonstrated more extensive EHK change in the adjacent epidermis (**Figure 2**E-F). Excellent capillary refill was noted and the procedure was well tolerated.

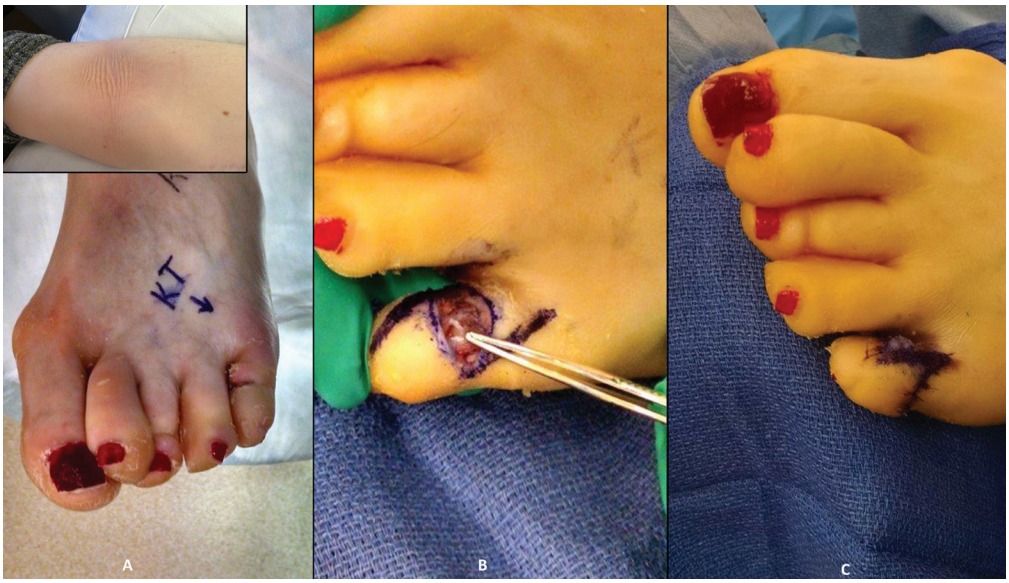

Figure 1. *A constriction band consistent with pseudoainhum encompassing approximately 50% of total circumference of the left small toe (A). A firm nodule representing an exostosis is also present at the dorso-medial aspect of the digit base. Note surrounding changes of a keratoderma and flexural courrugated hyperkeratosis characteristic of epidermolytic hyperkeratosis (A, inset). The pseudoainhum band was excised in toto with a transverse elliptical incision (B) and repaired with a z- plasty closure (C).*

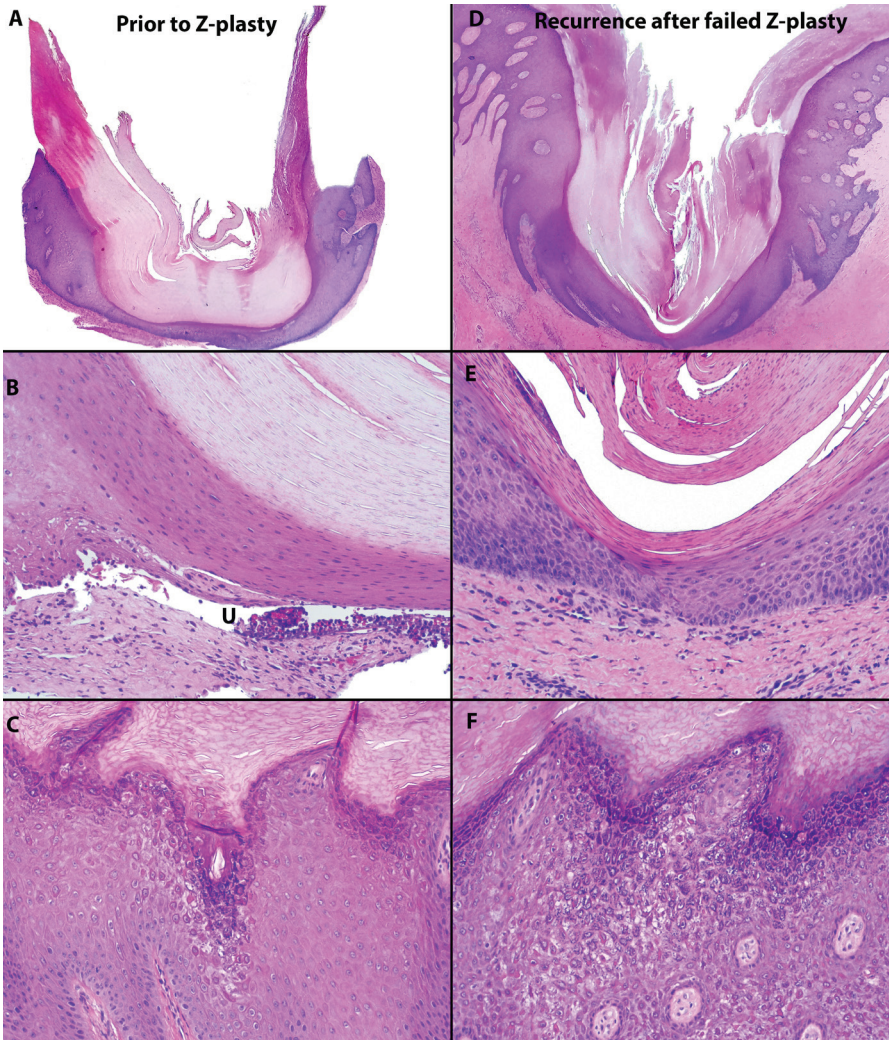

Figure 2. *Histopathologic examination of the excised skin prior to z-plasty (A-C) revealed an acanthotic epidermis, marked hyperkeratosis overlying a deep epidermal invagination (A) and focal ulceration at the base of the invagination (B) consistent with pseudoainhum. Focal hypergranular epidermoylytic change consistent with EHK was present in the epidermis adjacent to the groove (C). After recurrence of the constriction (D-F), histologic examination demonstrated a similarly hyperplastic epidermis with a crateriform invagination (D, E). The adjacent epidermis showed changes of epidermolytic hyperkeratosis that were more extensive compared to the previous specimen (F). Original magnification A, D, 20%; B, C, E, F, 100%.*

Three weeks post-operatively there was 100% take of the graft and both the medial forearm donor site and recipient site wounds were healing well and pain free. The surgical scar continued to remodel and the patient returned to her normal activities. At two-year follow up the recipient site had remodeled with no evidence of recurrence, pain, or neuromuscular compromise and the patient was satisfied with the functional and cosmetic outcome.

Case Discussion

This is a rare documented case of PA in a patient with EI. Based upon a search of the PubMed database using the terms "full thickness skin graft," "skin graft," "pseudoainhum," and "ainhum," it is also, to our knowledge, a novel case of PA treated with full thickness skin graft.

EI is an autosomal dominant condition caused by mutations in the KRT1 or KRT10 genes, affecting 1 in 200,000-300,000 people [6]. It presents at birth, often as erythema and bullae, progressing with age to a characteristic cobblestone and corrugated hyperkeratosis with predilection for the limbs. In adulthood, the genodermatosis is characterized by one of several heterogeneous clinical phenotypes consisting of varying degrees and distributions of hyperkeratosis, erosions, and bulla formation [6]. PPK, as seen in our patient, is associated with mutations in KRT 1 and is a likely contributor to the development of PA given the increased incidence of this entity with other keratodermas. It is unclear how hyperkeratosis contributes to the development of PA. Interestingly, while EI-type change was noted in both surgical specimens, it was more pronounced in recurrent PA; although not definitive, this finding raises the possibility that epidermal disease contributed to failure of the surgical procedure.

Given our patient's history, her constriction band can be classified as a type 2 PA, associated with infection or underlying disease process. In contrast, type 1 PA is secondary to congenital foreign body constriction (e.g., umbilical cord) and type 3 to acquired foreign body constriction (e.g., hair), [2]. In types 1 and 3 the patient's epidermis is presumed to be otherwise normal, a presumption that is not made in type 2. This distinction may be of particularly relevance to therapy.

Conclusion

Surgical excision of the ring and subcutaneous tissue with z-plasty closure has long been considered the

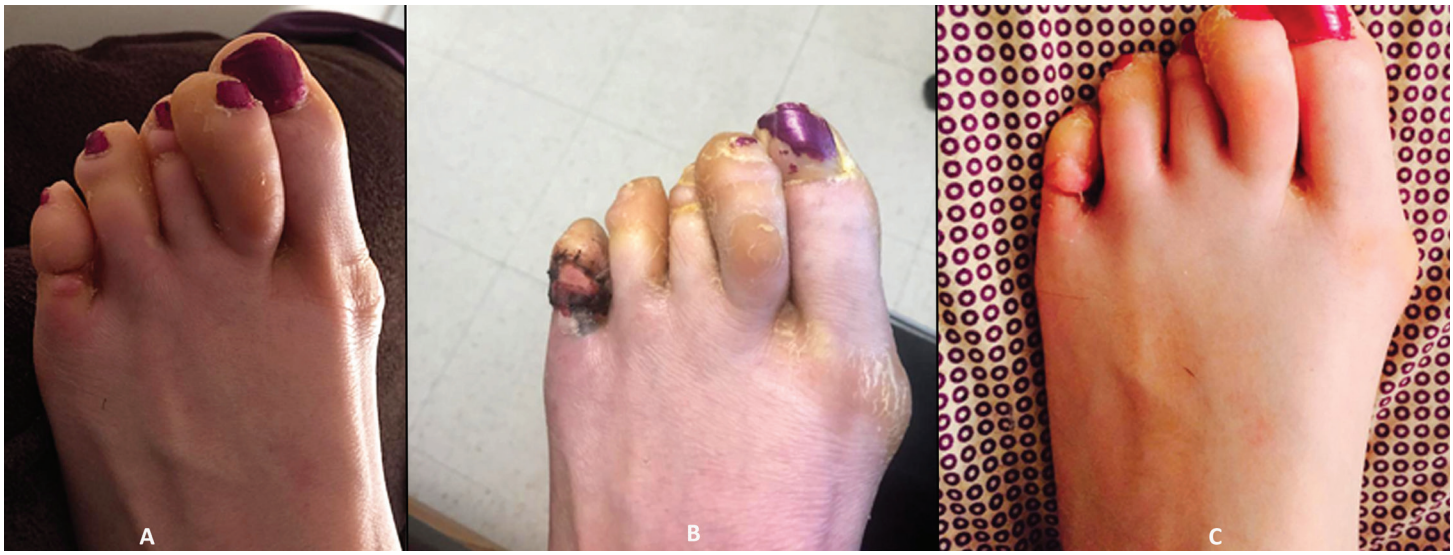

Figure 3. *Recurrent pseudoainhum at 10 months after initial z-plasty (A). Note successful repair with a full thickness skin graft, viable at 3 weeks (B) with no recurrence of pseudoainhum at 1 year.*

preferred treatment for acute PA [2, 7], although additional surgical procedures have also been described (**Table 2**), [4, 8]. In light of this case, we propose that full thickness skin graft be considered as a therapeutic option in cases of PA associated with a hyperkeratotic epidermis or as a salvage modality in the setting of a failed z-plasty.

Table 2: *Reported therapeutic options for acute pseudoainhum*

Single or multi-stage z-plasty closure
Medium thickness skin graft with advancement flap
Linear circular closure
Amputation

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