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Case presentation

Linear acral pseudolymphomatous angiokeratoma of children with associated nail dystrophy

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Abstract

Acral pseudolymphomatous angiokeratoma of children (APACHE) is a rare entity that typically occurs on the extremities of young females. Although linear arrangement of cutaneous lesions has been rarely reported, accompanying nail dystrophy has not been linked with this condition to our knowledge. We describe a case of linearly-oriented infiltrative papules and nodules on the index finger of a young female with associated onychodystrophy. Histology demonstrated a heavy lymphocytic infiltrate with plasma cells and proliferation of blood vessels consistent with APACHE. Our case is unique given the linear array of cutaneous lesions and associated nail dystrophy.

Key Words: acral pseudolymphomatous angiokeratoma of children, pseudolymphoma, nail dystrophy

Introduction

As the name suggests, acral pseudolymphomatous angiokeratoma of children (APACHE) is a benign pseudolymphomatous condition most often occurring on acral sites in children. Previous cases have demonstrated a female preponderance with 81% of cases occurring in young girls between the ages of 2 and 13. Clinically, lesions can be variable in appearance but often have an angiomatous quality with erythematous to violaceous color [1]. We report a 10-year-old girl with linearly-arrayed pseudolymphomatous papules on the index finger with accompanying dystrophy of the index nail plate.

Case synopsis

A 10-year-old girl with past medical history of asthma and atopic dermatitis presented to the dermatology clinic with a 4-month history of mildly pruritic erythematous infiltrative papules and nodules on her left dorsal index finger (Figure 1). Lesions were pseudovesicular in appearance and initially herpetic whitlow was

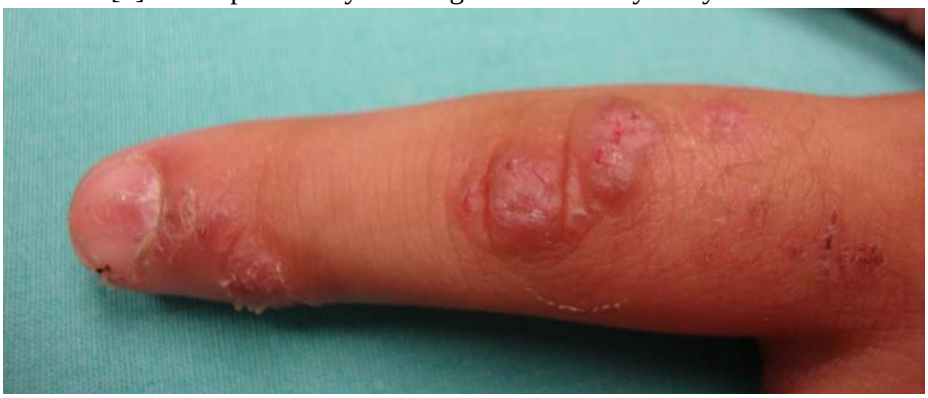


Figure 1. Pseudovesicular papules and nodules on the left dorsal index finger with prominent dystrophy of the central nail plate.

considered. Herpes PCR, however, was negative. Dyshidrosis was also considered given her history of atopic dermatitis. She was initially treated with triamcinolone 0.1% cream with some improvement, but without complete resolution.

At the follow-up visit, biopsies were performed for hematoxylin and eosin examination and direct immunofluorescence. Histology was suggestive of a pseudolymphoma with accompanying vascular proliferation. Given the age of the patient, clinical appearance of lesions, and histological findings a diagnosis of APACHE was made. Treatment was initiated with clobetasol ointment topically and intralesional triamcinolone. Lesions were much improved and barely visible at a six week follow-up. With time and the steroid treatments, the nail plate dystrophy was anticipated to normalize.

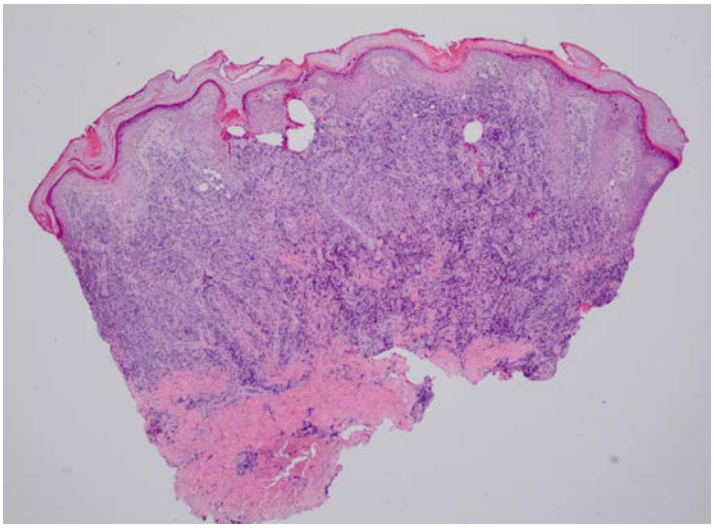
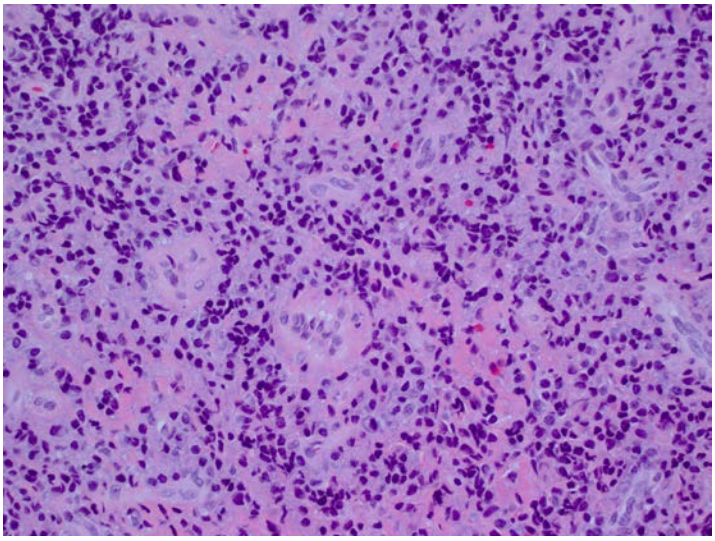


Figure 2. A dense and nodular lymphocytic infiltrate is visible extending to the deep reticular dermis (HE, 40x).

Pathological findings

Histologic examination revealed a diffuse band-like infiltrate composed mostly of lymphocytes; primary or secondary lymphoid follicles were not observed. The infiltrate extended from the papillary dermis to the deep reticular dermis. Within the lymphocytic infiltrate, several admixed plasma cells were identified (Figure 2). Eosinophils were notably absent. As seen in Figure 3, increased blood vessels lined by plump endothelial cells were a prominent feature. Dilated blood vessels were also visible in the papillary dermis being clutched by rete ridges of acanthotic epidermis (Figure 4). No atypia was noted of the infiltrating cells or vessels.



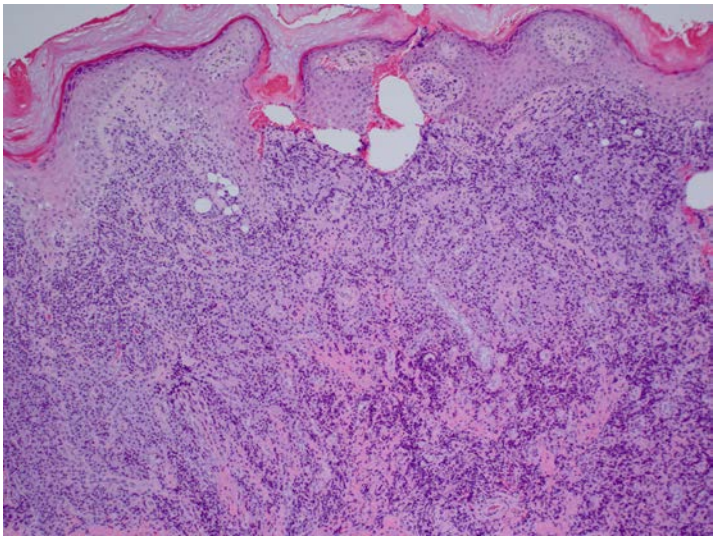


Figure 3. Multiple blood vessels lined by plump endothelial cells are noted (HE, 400x). **Figure 4.** Dilated vascular spaces in the papillary dermis being clutched by acanthotic epidermal rete ridges (HE, 100x).

Discussion

Acral pseudolymphomatous angiokeratoma of children is a rare, benign condition that was first described in 1988 by Ramsay *et al*[2]. The underlying cause of APACHE remains unknown. It was initially regarded as a benign vascular condition, but the most recent consensus is that it represents a pseudolymphoma [3,4]. This is still under debate, however, as a recent study by Tokuda *et al*[5], noted increased staining of vessels with lymphatic endothelium-specific marker podoplanin and blood vessel endothelium-specific marker CD34, suggesting that APACHE may represent a type of vascular malformation. Twenty-six cases have been reported in the literature to date. As the name suggests, most people diagnosed with this entity are children, but this diagnosis has been made in adults as well. The majority of patients are females (81%) and most, but not all, lesions are located on the upper or lower extremities [1-5]. In our case, the heavy infiltrate of benign lymphocytes, proliferation of blood vessels, and acral location in a young female were consistent with the diagnosis of acral pseudolymphomatous angiokeratoma of children (APACHE).

Clinically, lesions are often unilateral and can be variable in presentation. Lesions can be grouped and range from skin-colored papules and nodules to more vesicular and bullous-appearing erythematous and violaceous lesions. Linear cases have been reported previously, but to our knowledge, this is the first case of APACHE involving nail plate dystrophy. Cutaneous inflammatory entities with a linear presentation associated with nail dystrophy in children include, but are not limited to, lichen striatus [6], linear porokeratoses [7], linear lichen planus, linear psoriasis, and inflammatory linear verrucous epidermal nevus. Our case suggests that a diagnosis of APACHE should also be considered in the clinical differential diagnosis of an eruption in children with a linear distribution and accompanying nail dystrophy.

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