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Authors

Margulies, Shae
Welborn, Macartney
Vincek, Vladimir

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Diagnosis of perianal pyramidal protrusion in infants

Shae Margulies[1] MD, Macartney Welborn[2] MD, Vladimir Vincek[2] MD PhD

Affiliations: [1]Department of Medicine, University of Florida College of Medicine, Gainesville, Florida, USA, [2]Department of Dermatology, University of Florida College of Medicine, Gainesville, Florida, USA

Corresponding Author: Macartney Welborn MD, Department of Dermatology, University of Florida College of Medicine, 4037 NW 86 Terrace, 4th Floor, Gainesville, FL 32606, Tel: 352-594-1930, Fax: 352-594-4336, Email: mwelborn@sagisdx.com

Abstract

Infantile perianal pyramidal protrusion is characterized by a light pink to skin-colored soft tissue protrusion that is often midline and anterior to the anus. It most commonly occurs in young females and is relatively asymptomatic. Although biopsies are not routinely done, histopathology is relatively nonspecific and can appear similar to an acrochordon. The differential diagnosis is broad and clinical misdiagnosis as condyloma can lead to unnecessary accusations of child abuse. We report a case of perianal pyramidal protrusion that was originally biopsied owing to concern of condyloma acuminatum or molluscum. This case raises awareness of this diagnosis to help avoid unnecessary procedures and prevent emotional distress that could come for families with an inaccurate diagnosis of condyloma in young children.

Keywords: infantile, PPP, perianal, pyramidal protrusion

Introduction

Infantile perianal pyramidal protrusion is a benign lesion found in young children, most commonly females. It is characterized by a skin-colored or pink protrusion near the anus that is usually self-resolving. Importantly, infantile perianal pyramidal protrusion can present similarly to condyloma and can lead to a misdiagnosis of child abuse. Herein, we present a case of infantile perianal protrusion to raise awareness of the diagnosis and reduce unnecessary emotional distress to patients and families during evaluation.

Case Synopsis

A one-year-old girl presented to clinic with a several-month history of a non-pruritic skin lesion on the buttocks. Physical examination was significant for a skin-colored papule on the gluteal cleft that was originally clinically favored to be molluscum contagiosum (**Figure 1**). As the lesion on the gluteal cleft did not resolve, a shave biopsy was performed to rule out condyloma acuminatum. Histopathology showed an orthokeratotic stratum corneum overlying an epidermis with slightly irregular acanthosis (**Figure 2**). Within the dermis, there was a perivascular lymphocytic infiltrate. No molluscum or koilocytes were identified. When correlated with the clinical presentation, pathology was most compatible with infantile perianal pyramidal protrusion (PPP).

Case Discussion

Infantile perianal pyramidal protrusion was first described in 1996 by Kayashima who reported

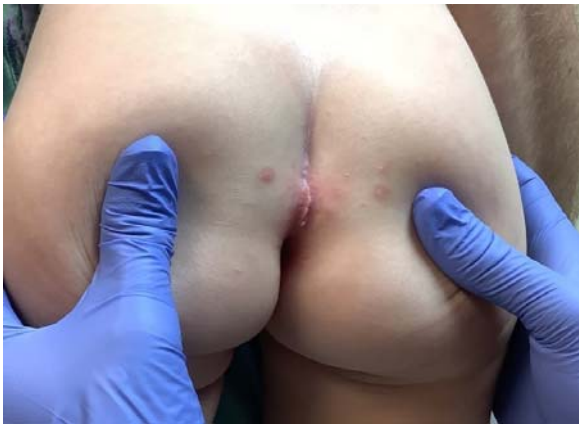

Figure 1. *Skin colored papule on the gluteal cleft.*

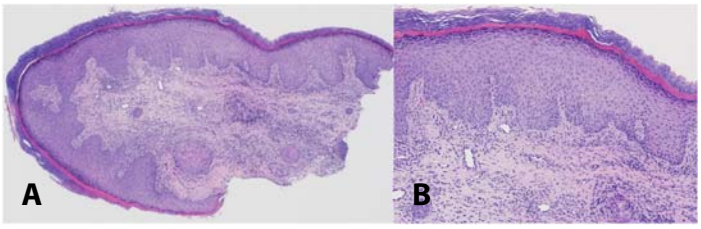

Figure 2. *H&E histopathology.* ***A)*** *Low power view of histopathology showing orthokeratotic stratum corneum overlying an epidermis with slight irregular acanthosis, 40×.* ***B)*** *Higher power view of histopathology showing orthokeratotic stratum corneum overlying an epidermis with slight irregular acanthosis, 200×.*

fifteen cases of perianal eruptions located in the midline anterior to the anus [1]. In our case presentation, the infantile PPP was found superior or posterior to the anus which may have made the clinical diagnosis more challenging. In the diagnosis of PPP, the location is always midline but can be located anywhere along the median raphe from the perineum to the intergluteal crease. Perianal pyramidal protrusion most commonly presents in girls; a review of the literature from 1989 to 2005 showed that 91 of the 92 reported cases were female [2]. The entity occurs in children aged newborn to 11 years, with the highest incidence in newborns [3]. Clinical characteristics of PPP include a pyramidal soft tissue protrusion with a smooth surface and red/pink colored skin [2]. Dermoscopy may aid in distinguishing PPP from other similar appearing and more serious pathologies, such as condyloma. Bartolomeo et al. proposed standardized dermoscopy findings which include a waxy surface with globular, linear, and dotted vessels in a linear arrangement [4,5]. Additionally, the dermoscopy has been compared to a "prickly pear pad," as there are notable prominences on a waxy surface [4]. The pathogenesis is unclear, but there may be a component of weakness of the female perineum or persistent constipation that could lead to the development of PPP [6]. Perianal pyramidal protrusion has also been hypothesized to be a potential sign for early lichen sclerosus. In a study of four patients with PPP, three of the patients showed subtle clinical findings of lichen sclerosus during their first visit [7]. The fourth patient in the study by García-Doval et al. developed signs of lichen sclerosus a few months after first being seen and biopsy results of all patients showed histopathologic features of lichen sclerosus [7].

Histopathology is relatively nonspecific, and there has not been extensive data collected from patients with PPP. Performing a biopsy on an infant or young child can be uncomfortable for both the patient and parents/caregiver. Most providers would opt against a biopsy in such a young patient unless necessary for the diagnosis, especially in such a sensitive region. For those patients who did receive a biopsy, common histologic findings include epidermal acanthosis, dermal edema, and overall acute inflammatory changes [3]. If there is clinical concern for child abuse, PPP can be distinguished from condyloma through histologic findings of koilocytes and a positive human papillomavirus (HPV)test on the tissue [8].

As the clinical presentation of PPP can be similar to other dermatologic conditions, the differential diagnosis is broad. Other pathologies to consider include hemorrhoids, granulomatous lesion of inflammatory bowel disease, perianal midline malformation, rectal prolapse, infantile hemangioma, and condyloma acuminata, which could suggest sexual abuse. [2]. Possibly the most serious of these diagnoses to rule out is that of sexual abuse/condyloma acuminata. Perianal pyramidal protrusion can be easily mistaken for child abuse which can lead to significant emotional distress for the family that can be avoided. It is important to be aware of PPP as a benign entity unrelated to child abuse to help prevent misdiagnosis. However, not all HPV-associated warts are related to child abuse. Studies have shown that in children with anal warts, the range of associated sexual abuse was very wide, 0-80% [9]. The association of abuse has been shown to increase with age, particularly over the age of 8 [9]. More awareness of PPP can also prevent unnecessary procedures for a benign condition in a young child.

Multiple treatment options have been proposed for PPP, including topical corticosteroids (hydrocortisone and betamethasone), tacrolimus, and zinc oxide [10]. Most commonly, lesions resolve on their own without treatment. Because of the association with constipation in children, ensuring that the patient is having regular stools may help

resolve the PPP and should always be considered as first line treatment.

Conclusion

Infantile PPP is a benign lesion that can clinically mimic condyloma and molluscum, but has nonspecific histopathology. It is important that dermatologists and dermatopathologists are aware of this diagnosis to avoid unnecessary biopsies and emotional distress for families.

Potential conflicts of interest

The authors declare no conflicts of interest.

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