Infantile perineal protrusion

Amor Khachemoune, MD, CWS,[a] Kjetil Kristoffer Guldbakke, MD,[b] and Eric Ehrsam, MD[c]
Brooklyn, New York; Boston, Massachusetts; and Le Cateau, France

Infantile perineal protrusion, a relatively newly recognized condition, is underreported in both the dermatologic and pediatric literature. The name "infantile perineal protrusion" has evolved based on the typical anatomic location, morphologic features, and prevalence in prepubertal children. It occurs in 3 settings: constitutional (sometimes genetic or familial); functional (after constipation, diarrhea, or other irritant exposure); or associated with lichen sclerosus et atrophicus. Recognition of infantile perineal protrusion by dermatologists and pediatricians has many implications regarding proper diagnosis and management. The condition may be mistaken for condyloma acuminata or as a sign of trauma, leading to an erroneous investigation of sexual abuse. In this article, we report two new cases and make a thorough review of the literature to elucidate the mechanisms, diagnosis, classification, and management to clarify this often misdiagnosed condition. (J Am Acad Dermatol 2006;54:1046-9.)

Infantile perineal protrusion (IPP) is a recently described entity. Although the number of cases reported in the literature is increasing, published data are still limited. The term "infantile perianal pyramidal protrusion" was initially introduced by Kayashima et al[1] in 1996. They reported 15 infants ranging from 1 to 30 months old with acrochordon-like protrusions occurring anterior to the anus. The average age of their patients was 14.1 months, and 14 (94%) were female. Perineal protrusion had previously been described as acrochordons or skinfolds. A subsequent report by Merigou et al[2] described an additional 4 patients, with a similar consensus on the terminology. Cruces et al[3] later argued that "perineal" is a more qualified term than "perianal" in describing lesions located in the perineal median raphe, and emphasized that "pyramidal" is sometimes inexact; the authors preferred "infantile perineal protrusion" as a simplified and more accurate term. To our knowledge, the literature to date is limited to 92 patients in 8 case reports and case series (Table I).

From the Department of Dermatology, State University of New York Downstate Medical Center[a]; International Training Program at Harvard Medical School, Boston[b]; and private practice, Le Cateau, France.[c]

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Reprint requests: Amor Khachemoune, MD, CWS, Department of Dermatology, State University of New York Downstate Medical Center, Box 46, 450 Clarkson Ave, Brooklyn, NY 11203. E-mail: amorkh@pol.net.

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CASE 1

A healthy, 15-week-old Caucasian girl was brought by her mother for the evaluation of a newly developed growth in her perineum. The mother reported that her daughter developed this lesion 2 weeks earlier, after an episode of constipation. On physical examination of the perineal region, there was a pink- to flesh-colored smooth-sided skin tag–like protrusion, measuring 8 × 6 mm, localized to the medial part of the perineum between the anus and vulva (Fig 1). The lesion partially resolved 2 months later without treatment.

CASE 2

A healthy, 9-month-old Caucasian girl developed a smooth, tongue-shaped, 18- × 10-mm red lesion, localized in the midline of the perineum between the anus and vulva; there were multiple erosions and crusted papules on the perineum (Fig 2). The diagnoses of infantile pyramidal protrusion and irritant diaper dermatitis were made, both of which developed after a prolonged episode of diarrhea. The dermatitis resolved after 1 week with topical antifungal therapy and local diaper area care, and the protrusion partially resolved 2 months later with no specific therapy.

EPIDEMIOLOGY

IPP occurs almost exclusively in girls (91/92 reported cases), affecting both infants and prepubertal children. Konta et al[4] evaluated 500 children (224 boys and 276 girls) who ranged in age from newborn to 11 years for the occurrence of IPP. In this series, it was noted that 13% of the girls (36 of 276) had nodules, but none of the boys. Among girls, the

Table I. Summary of published articles on infantile perineal protrusion to date

Author	Year of publication	No. of patients	Age range	Sex of patients	LS&A associated	Location from anus
McCann et al[5]	1989	18	Up to prepubertal	All female	None	Anterior: 17
Kayashima et al[1]	1996	15	1-30 mo	14 female, 1 male	None	Anterior: 15
Merigou et al[2]	1998	4	6-20 mo	All female	None	Anterior: 4
Cruces et al[3]	1998	4	2-7 y	All female	All 4	Anterior: 4
Konta et al[4]	2000	36	0-11 y	All female	None	Anterior: 36
Patrizi et al[6]	2002	13	7 mo-7 y	All female	3/13	Anterior: 8 Posterior: 3 Anterior and posterior: 2
Miyamoto et al[7]	2004	1	11 mo	Female	No	Anterior
Fleet et al[8]	2005	1	12 mo	Female	No	Anterior

LS&A, Lichen sclerosus et atrophicus.

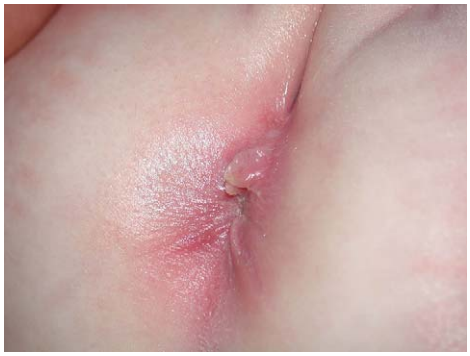

Fig 1. Case 1. Skin tag–like, 8- × 6-mm medial protrusion between anus and vulva of 15-week-old girl.

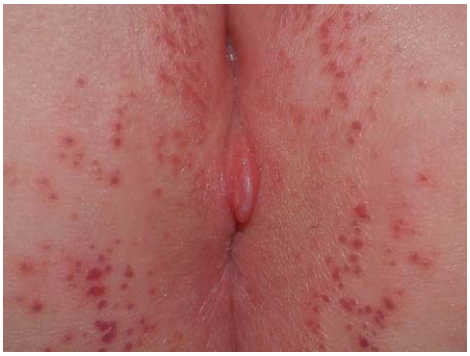

Fig 2. Case 2. Medial solitary protrusion (18 × 10 mm) anterior to anus in 9-month-old girl. Surrounding skin of protrusion has multiple erosive papules (extensive form of irritant diaper dermatitis).

incidence was highest in the newborn group (15.8%, 6 in 38) as compared with patients aged 2 months to 3 years (13.4%, 29 in 217) and 4 to 11 years (4.8%, 1 in 21). McCann et al[5] performed a study in 1989 delineating normal perineal pathology in 267 prepubertal children. In this study, anal skin tags and skinfolds, with similarities to the later-described entity of IPP, were noted in 11% (18 of 164). The authors of this review article agree with McCann et al[5] and estimate the incidence in girls at around 11% to 13%, making IPP a common condition in female infants and children.

CLINICAL FEATURES

A typical lesion of IPP appears as a pyramidal soft-tissue protrusion with a tonguelike lip. The surface is smooth or slightly velvety, covered with normal rose- or red-colored skin. They are classically located in the midline just anterior to the anus. There are also reports of IPP localized posteriorly, and two cases of double IPP anterior and posterior to the anus.[6] Besides the typical pyramidal shape, other morphologic descriptions include leaflike and hen's crest–, tongue tip–, peanut-, and cigar-shaped protrusions. Symptomatology is variable ranging from asymptomatic to pain on defecation, with or without fissuring, making it difficult to clean the perianal region.

PATHOGENESIS

The cause of IPP remains unclear. Patrizi et al[6] divided IPP into 3 groups: (1) familial and/or congenital; (2) functional; and (3) lichen sclerosis-associated.

A constitutional anatomic weakness in the perineum of female patients has been hypothesized as a potential cause.[2] The condition may also represent a constitutional weakness of the median raphe or perineum, which also would explain the rare occurrence in male patients. Supporting this hypothesis is the observation of congenital IPP and the presence of perianal protrusion in other family members.[3,4,6] Konta et al[4] proposed that the perineal nodule may be a remnant of a projected tip of the urogenital septum. This is based on the embryologic notion that the perineum is formed by elongation of the urogenital septum during fetal growth, and the