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Infantile Perianal Protrusion

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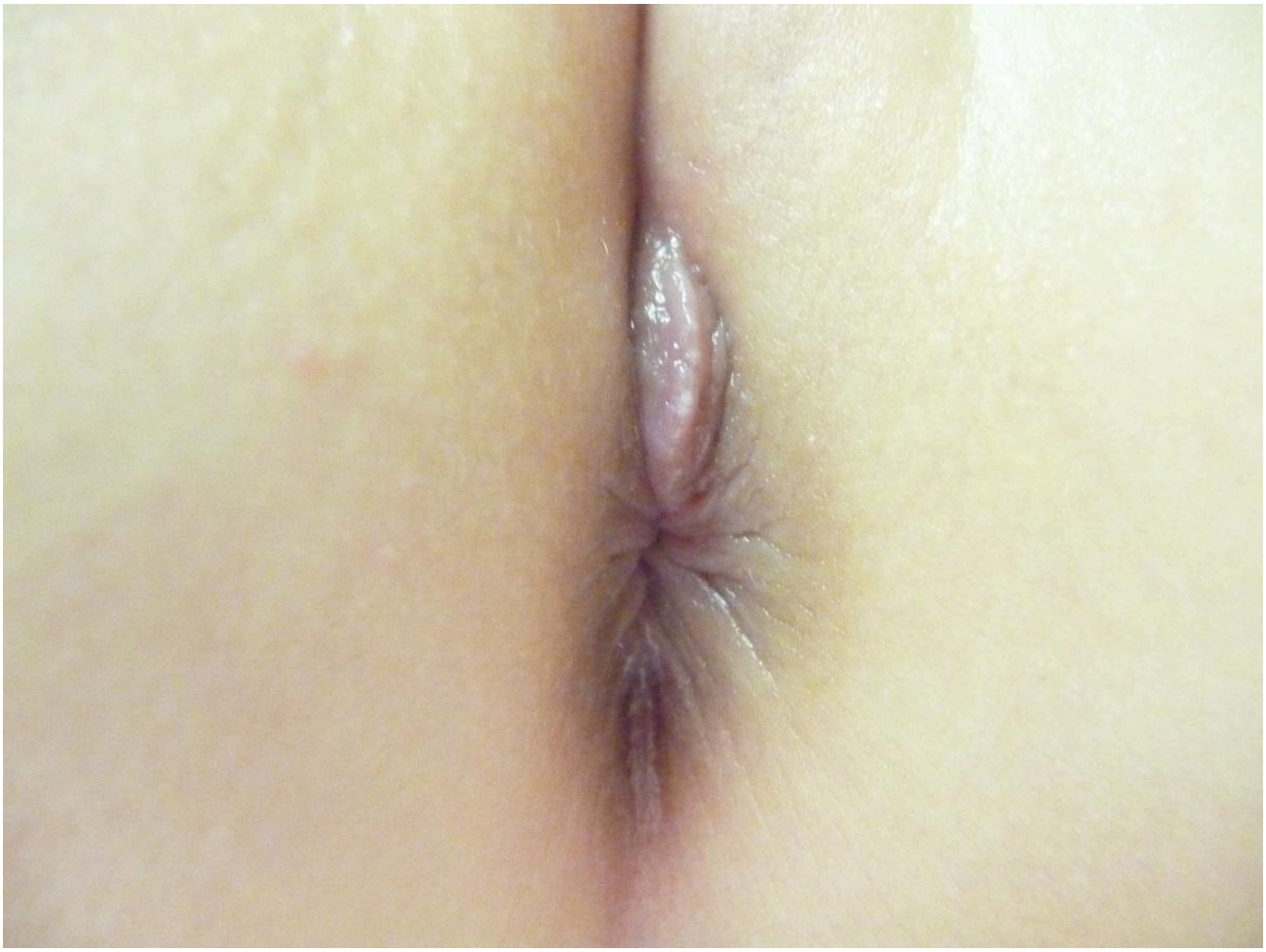
An 11-month-old girl was noted to have a protrusion in the perianal area during a routine physical examination. She had been born at term via normal vaginal delivery to a gravida 3, para 2, 26-year-old mother following an uncomplicated pregnancy. The neonatal course had been uneventful. The patient had been exclusively breastfed for the first 6 months of life. Solid food had been introduced at 6 months, and she had experienced constipation since then. She had bowel movements once every 3 or 4 days. Stools were hard and pellet-like, and the patient seemed to be in pain on defecation. There was no blood on the surface of the stool. The parents denied vigorous wiping of the girl's perianal area. There was no suspicion of sexual abuse. Her past health and family history were unremarkable.

On examination, there was a pyramidal protrusion in the perianal area in the 12 o'clock position. The protrusion was purplish red in color, smooth surfaced, nontender, and soft to firm in

consistency. There was no anal fissure noted or lichen sclerosus et atrophicus visible. The rest of the physical examination findings were unremarkable.



Based on the clinical appearance, a diagnosis of infantile perianal protrusion was made. The patient was treated with lactulose, liberal intake of fluid, and increased intake of fruit and vegetables. The constipation subsided in 1 month. The perianal pyramidal protrusion gradually decreased in size with time and was no longer visible at the 18-month routine checkup.

Discussion. Infantile perianal protrusion is a relatively newly recognized clinical entity. The term *infantile perianal pyramidal protrusion* was coined by Kayashima and colleagues in 1996 to define a condition characterized by a solitary pyramidal soft-tissue swelling with a red or rose-colored surface at the midline of the perianal area, usually anterior to the anus.[1] Some authors have proposed *infantile perineal protrusion* as a simplified or more appropriate name, given that the shape of the protrusion might not always be pyramidal.[2-5] Also, protrusions situated posterior to the anus have also been described.[1,6] As such, the term *perianal* seems to be more appropriate than *perineal*.[4,5]

Epidemiology. Infantile perianal protrusion is a common condition in children. Konta and colleagues examined 500 children (224 boys and 276 girls) at the Hirosaki Public Health Center and the Hirosaki University Hospital in Hirosaki, Japan, for the occurrence of infantile perianal protrusion.[3] The children ranged from newborn to 11 years of age. In this study, the condition

protrusion.[3] The children ranged from newborn to 11 years of age. In this study, the condition was found in 13% (36 of 276) of the girls and none of the boys. Infantile perianal protrusion is most common in children younger than 1 year, although the condition can also affect prepubertal children.[3,7,8] The majority of affected children (>95%) are girls.[3,5,8-10]

Etiopathogenesis. Three types of infantile perianal protrusion have been recognized—constitutional, functional, and lichen sclerosis et atrophicus-associated.[3-6,11-13] The constitutional type is the most common type and is often present at birth.[9,14] Presumably, constitutional infantile perianal protrusion is due to an inherent weakness in the perianal region in girls.[5,8,10,15] Constitutional predisposition to infantile perianal protrusion is supported by the fact that the condition can be congenital or familial.[2,5,6,8,11] Infantile perianal protrusion has been reported in monochorionic twins, suggesting genetic predisposition to this condition at least in some cases.[11] When it is congenital, infantile perianal pyramidal protrusion may be a remnant of a projected tip of the urogenital septum.[3-6] The functional type may be secondary to constipation or to mechanical irritation from vigorous wiping after defecation.[4-6,8,13,15] Infantile perianal pyramidal protrusion can be a manifestation of vulvar lichen sclerosis et atrophicus.[2,5,6,8] This is the least common type.[5]

Histopathology. Histopathologic findings are nonspecific and include epidermal acanthosis, slight thickening and/or elongation of rete ridges, upper dermal edema, dilation or engorgement of capillaries, and dilation of lymphatic vessels.[7,8] The histology of the lesion associated with lichen sclerosis is distinctive. Typically, the epidermis is atrophic with orthokeratotic hyperkeratosis. Other features include vacuolar degeneration of basal keratinocytes, edematous and sclerotic papillary dermis, homogenization areas of the collagen in the papillary dermis, and lymphohistiocytic infiltrates in the dermis.[8,16]

Clinical manifestations. Infantile perianal protrusion is characterized by asymptomatic protrusion with a rose red or purplish red surface, along the midline, usually anterior to the anus, with the major axis consistent with the median raphe.[4,6,8,16,17] At times, the protrusion may occur posterior to the anus.[5] The protrusion usually ranges from 0.5 to 1.5 cm. The lesion is usually solitary.[1,10,13,18] Rarely, concomitant anterior and posterior perianal protrusions have been reported.[6,17] Also rarely, lateral perianal protrusions have been reported.[9] The protrusion is usually pyramidal in shape.[4,5] In congenital cases, the protrusion usually has a leaf-like appearance.[4,5,9] Occasionally, the protrusion may be hen's crest-, tongue-tip-, peanut-, or cigar-shaped.[5,7]

Most children with infantile perianal protrusion are asymptomatic. The condition is usually discovered incidentally, either by a physician during a routine physical examination or by a parent while changing the diaper of the child or bathing the child.[5] Some children with perianal protrusion have a history of constipation or painful defecation.[5]

Infantile perianal protrusion can at times be a manifestation of anogenital lichen sclerosis et atrophicus. Therefore, the finding of infantile perianal protrusion should warrant a search for other clinical features of anogenital lichen sclerosis et atrophicus, such as dysuria, recurrent urinary tract infection, painful defecation, fecal soiling, itchiness or soreness in the anogenital area, anal fissure, and shiny, ivory-white, atrophic plaque in the anogenital region.[5]

Diagnosis. The diagnosis is mainly clinical, based on the history and physical findings. The clinical features of infantile perianal protrusion are so distinct that there should not be any difficulty in making the diagnosis. No laboratory test is necessary. In doubtful cases, dermoscopy and ultrasonography should be considered, which will increase the diagnostic accuracy. Dermoscopy typically shows patchy, structureless, white areas and a vascular pattern composed of red globular and dotted vessels.[16] On the other hand, ultrasonography usually reveals a thick hypoechoic area of skin consistent with high blood flow, probably due to vasodilation and inflammation.[16]

Differential diagnoses. Infantile perianal protrusion may be mistaken for perianal skin tag, genital wart, traumatic lesion resulting from sexual abuse, hemorrhoid, rectal prolapse, granulomatous perianal lesion of inflammatory bowel disease, and capillary hemangioma.[1,4,8,12,13,16]

Complications. Infantile perianal protrusion might be of medicolegal significance if the lesion is mistaken for condyloma acuminatum or a sign of trauma resulting from sexual abuse.[5]

Prognosis. Most constitutional lesions eventually resolve spontaneously with time, usually within several months.[4,5] Functional and lichen sclerosis et atrophicus-associated lesions usually heal within 2 to 6 weeks with proper treatment of the underlying cause.[4,5]

Management. Underlying conditions, if present, should be treated.[4,5] For children with constipation, remedies to loosen the stool may help expedite resolution of the lesion. Affected children can be treated with liberal intake of fluid, increased intake of food with a high fiber content, and, if necessary, a stool softener such as lactulose and polyethylene glycol.[5]

Anogenital lichen sclerosis et atrophicus, if present, should also be treated with either ultrapotent topical steroids such as clobetasol, or topical immunomodulators such as tacrolimus or pimecrolimus.[5,8]

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