Infantile Pyramidal Protrusion as a Manifestation of Lichen Sclerosus et Atrophicus

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Background: A perineal infantile lesion previously described as “skin tag/fold” has recently been named infantile perianal pyramidal protrusion. It appears on the perineal median raphe of girls as a pyramidal soft tissue swelling, covered by smooth, red or rose-colored skin. Its pathogenesis is unknown. As in the case of other perianal lesions, knowledge about it is important, as concern about signs of child abuse grows.

Observations: Four girls, 2 of them sisters, with infantile perianal pyramidal protrusion were studied. Three of these girls showed subtle clinical evidence of classic lichen sclerosus et atrophicus on first examination. The other girl developed vulvar lesions of lichen sclerosus et atrophicus months after the diagnosis of infantile perianal pyramidal protrusion. All 4 protrusions disclosed histopathological findings diagnostic of lichen sclerosus et atrophicus.

Conclusions: Infantile perianal pyramidal protrusion is, at least in some patients, a peculiar form of lichen sclerosus et atrophicus that can precede other, more characteristic manifestations. We suggest changing the name to the more precise infantile perineal protrusion. Knowledge of this hitherto unrecognized clinical form of lichen sclerosus et atrophicus can help to explain ano-genital symptoms and avoid its misinterpretation as a sign of sexual abuse.

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REPORT OF CASES

CASE 1
A 7-year-old girl was brought to our hospital with a perineal lesion, present at birth, that had had several episodes of inflammation. She had previously consulted the Pediatrics Department for recurrent urinary tract infections, without finding the cause. She had a perineal, protruding, swollen lesion, $19 \times 10 \times 7$ mm, rose colored, with a smooth surface and whitish zones (Figure 1). On physical examination, flat warts were also found. The perineal lesion was excised with the patient under local anesthesia. Two months later, typical lesions of lichen sclerosus et atrophicus were found in the genitalia. The biopsy specimen showed, at low magnification, a sessile configuration (Figure 2). The epidermis, covered by an orthokeratotic cornified layer, showed focal atrophic areas with loss of rete ridges pattern and vacuolar alteration of basal cells, being hyperplastic at the margins. The papillary dermis was markedly thickened, with alternating areas of sclerosis and homogenization of the collagen, and areas of striking edema. Below this altered superficial dermis there was a moderately dense lymphocytic infiltrate on an apparently normal tissue (Figure 3).

CASE 2
A 4-year-old girl with syndactyly that had been surgically treated 2 years earlier was
examined because of a hypopigmented area in her vulva, present for the last 2 years. She reported no symptoms.

On examination, a 10 × 4 × 6-mm perineal protrusion was found, as well as signs of genital lichen sclerosus et atrophicus: porcelain white macules, with atrophic skin. The protrusion was covered by smooth, rose-colored skin. A biopsy specimen of this lesion showed typical changes of lichen sclerosus et atrophicus. The protrusion disappeared in 3 months, after topical corticosteroid therapy, but minimal lesions of lichen sclerosus et atrophicus remained. We learned later that a previous biopsy of vulvar skin, taken at another hospital, was diagnosed as lichen sclerosus et atrophicus.

CASE 3

A 2-year-old girl, sister of patient 2, was seen in our department because her parents were bothered by her genital hypopigmentation. This hypopigmented skin had been present for the last year, covering her genitalia and perineal area. She suffered from dysuria and constipation. On physical examination, signs of genital lichen sclerosus et atrophicus were found, as well as a perineal protrusion, 6 × 4 × 5 mm, in the midline anterior to the anus. A punch biopsy specimen of this lesion showed patchy lichenoid infiltrates, with vacuolar alteration and homogenization of the collagen in the papillary dermis, changes consistent with the diagnosis of lichen sclerosus et atrophicus. The lesion had nearly disappeared after 2 months, without therapy.

CASE 4

A 5-year-old girl was seen in our hospital for atopic dermatitis. On physical examination, a perineal protuberant lesion was found, as well as hypopigmented and atrophic vulvar skin (Figure 4). Her parents said that both signs had been present from early childhood, the patient remaining asymptomatic. A biopsy specimen of the lesion showed changes similar to those in previous cases.

Special stains for elastic fibers (orcein) and amyloid (thioflavin T) showed disappearance of elastic fibers in the papillary dermis and absence of amyloid in all biopsy specimens.

None of the girls had a history of sexual abuse or showed fear of family members or friends or behavioral alterations. Their parents were cooperative and con-
cerned about their lesions. We considered sexual abuse highly unlikely.

COMMENT

Infantile perianal pyramidal protrusion has recently been described as a new perineal lesion mainly affecting girls.1 McCann et al2 previously reported “skin tags/folds,” similar to the former lesion, in 18 girls (11%) among 164 children selected for non–sexual abuse. We consider this designation inappropriate: skin tags or acrochordons are acquired polypoid lesions present in the neck, trunk, or, infrequently, the thighs or inguinal or genital area of middle-aged or older persons. The lesions that McCann et al described are sessile and perianal and affect children, making the diagnosis of acrochordon (skin tag) inadequate. Kayashima et al1 suggested the name infantile perianal pyramidal protrusion “because it is more descriptive of the affected age group, and the characteristic location and shape of the protrusion.” Our experience of a few months was sufficient to convince us of the value of their findings on this previously neglected structure.

Regarding the proposed name, we think that perineal is a better qualifier than perianal to describe a lesion located in the perineal median raphe, and pyramidal is sometimes inexact. We propose infantile perineal protrusion as a simplified, more accurate name.

The main point in our observations is the relationship that we found between infantile perineal protrusion and lichen sclerosus et atrophicus in our 4 patients. Three of them showed subtle clinical lesions of lichen sclerosus et atrophicus when first seen. The other girl developed signs of lichen sclerosus et atrophicus in the following months. Biopsy specimens of infantile perineal protrusions showed, in all 4 cases, histopathological findings diagnostic of lichen sclerosus et atrophicus. It should also be pointed out that in the two patients described by Kayashima et al who underwent biopsy, the histological figures and description are highly suggestive of lichen sclerosus et atrophicus. We also found earlier reports of genital lichen sclerosus et atrophicus with unnoticed infantile perineal protrusion (see Figure 1 in the article by Meyrick Thomas and Kennedy3).

Our interpretation of these facts is that infantile perineal protrusion can frequently be a manifestation of lichen sclerosus et atrophicus in girls, appearing simultaneously or asynchronously with more characteristic signs of this disease. Sometimes the infantile perineal protrusion is the main manifestation of lichen sclerosus et atrophicus, as in case 1. This is somewhat similar to the appearance of lichen sclerosus et atrophicus in boys in the form of phimosis, concealing minor changes of lichen sclerosus et atrophicus.4,5 Oddly, infantile perineal protrusion is infrequent or nonexistent in boys.1,2

Although we have found signs of lichen sclerosus et atrophicus in all of our patients, the high incidence of infantile perineal protrusion reported by McCann et al2 raises some concern. Perhaps the lesions reported by McCann et al are not homogeneous, representing an admixture of various conditions. We consider that 3 important clinical signs to characterize infantile perineal protrusion are its perineal location, its swollen aspect (making it different from redundant skin folds), and the absence of preceding fissures or fistulas in the anal side (to exclude secondary inflammation from these lesions).

The high incidence reported by McCann et al can also mean that subtle lichen sclerosus et atrophicus is much more frequent than previously reported, or that some infantile perineal protrusions are not caused by lichen sclerosus et atrophicus, but other causes. Congential protrusions or protrusions affecting sisters are frequent findings for lichen sclerosus et atrophicus. Moreover, there are descriptions of somewhat similar, but multiple, lesions in girls with regional enteritis.6,7 Infantile perineal protrusion may be a peculiar form of reaction, influenced by anatomical factors, of the perineal region of girls.

So far, we think that infantile perineal protrusion can frequently be a manifestation of lichen sclerosus et atrophicus. The finding of an infantile perineal protrusion should induce a search for other signs of lichen sclerosus et atrophicus and can help to explain anogenital manifestations, such as recurrent urinary tract infections (case 1), dysuria, or painful defecation (case 3). Infantile perineal protrusion is also important as it can be misinterpreted as a sign of sexual abuse, like other manifestations of lichen sclerosus et atrophicus.

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