Perineal groove: a rare anogenital anomaly.

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Summary
The perineal groove is a rare and usually uncomplicated congenital malformation of the perineum characterized by a wet sulcus – because covered by non keratinizing epithelium – that extends from the posterior fourchette to the anus. It has been very rarely described in dermatology literature. Here we report a case of this uncommon entity in a 6-year-old female child.

Key words
Perineal groove.

The perineal groove is a rare and usually uncomplicated congenital malformation of the perineum characterized by a wet sulcus that extends from the posterior fourchette to the anus. The condition is more commonly seen in female patients (1-9). Of the previously described cases, very few cases have been reported in the dermatology literature hence the knowledge of this minor perineal anomaly will help avoid misdiagnosis and prevent extensive evaluations or unnecessary surgical procedures.

Case report
A 6-year-old female child born of non consanguineous marriage, adequately immunized for age with normal development of milestones, was brought by parents with complaints of itching and redness over vulva and perineal region from 4 years.

The patient was apparently alright 4 years ago when the mother noticed persistent erythema over the vulva and perineal region. The child used to touch that area repeatedly due to itching and discomfort especially after urination. But from the last 6 months she was complaining of severe persistent itching over that area associated with increased redness. Persistent itching and irritation used to divert child’s attention in the school activity.

There was no history of appearance of bullous lesions over the vulva. Her parents denied history suggestive of sexual abuse. There was no history of difficulty in micturition and defecation.

The patient received multiple treatments with topical steroids like fluticasone, mometasone and clobetasol dipropionate, mupirocin cream and clotrimazole cream for the last 6 months for variable duration with little reduction in the erythema but no improvement in the itching.

Cutaneous examination of the patient revealed a small wet groove extending from fourchette to anus with peripheral erythematous borders (Fig. 1). The rectal examination showed normal anal caliber and mucosa. The rest of the cutaneous
and systemic examination of the patient did not reveal any abnormality. The patient received a clinical diagnosis of perineal groove and was managed conservatively by counselling of parents, oral antihistamines and zinc oxide paste with significant relief.

Discussion

The perineal groove is a rare congenital anomaly that occurs mostly in female patients, with only one reported case in a male patient (3). It has been described as congenital wet sulcus lined with mucous membrane extending from the vaginal fourchette to the anus (5). Though there is a paucity of literature on this embryological anomaly, it is considered as a more common entity than officially reported. The incidence of perineal groove is unclear but it can vary between 6 to 8.5% in different studies (2).

The exact pathogenesis of perineal groove remains unclear. Various theories have been put forward to explain embryological origin of perineal groove.

Abdel Aleem et Al. suggested that perineal groove could be due to failure of fusion of median genital folds that contribute to the formation of perineum in the midline (1) or it may possibly be a relic of the open cloacal duct (9).
Mullassery et al. considered it as an embryological remnant of the urorectal septum, based on the histology of the intervening area between the two ends of perineal groove, lined by rectal type of mucosa (6). On the other hand, others suggested it may be a result of faulty development of the embryonic cloaca, a failure of fusion of perineal raphe or a defect in the development of the uroanal septum (6, 7).

Clinically it has been described by Stephens to have 3 features (8): a wet groove in the perineum extending from fourchette to anus, normal formation of vestibule including urethra and vagina and hypertrophy of minor tails that surround the wet sulcus. Children may present with mucous discharge when the area is not completely epithelialized. Long term complications include constipation, infection of external genitalia (50%) urinary tract infection (15%) and persistent mucosal discharge (4).

The perineal groove may initially be confused with an irritant dermatitis, infection, lichen sclerosus, perianal pyramidal protrusion, trauma, or sexual abuse or ulcerated hemangioma, often rendering the diagnosis difficult.

The diagnosis is made clinically, with biopsy rarely performed. While histologic findings vary, non keratinizing squamous epithelium with an intervening area of rectal type or transitional epithelium has been reported from excised specimens (3, 4, 6). Imaging for associated regional anomalies may be considered, although such anomalies are believed to be rare. Reported regional anomalies with perineal groove include hypospadias, bifid scrotum, ectopic anus and urinary tract anomaly (3, 4, 10).

The perineal groove is known to be a self resolving condition but it can take more than a year to become re-epithelized (6, 7). Surgical correction is considered for cosmetic reason or if there is incomplete re-epithelization leading to persistent mucosal discharge and recurrent infections.

Surgical correction consists of resection of groove mucosa and closure of tissue defect by interrupted sutures. However, dehiscence of suture secondary to urine and fecal contamination is well known. Hence covering the suture line with chemical glue can reduce the chances of wound dehiscence, risk of infection and improve the quality of cicatrization (4).

The perineal groove is unknown to many pediatricians and dermatologists as the majority of these cases are handled by the pediatric surgeons. Hence, the knowledge of this underreported entity is extremely important as it can be easily misdiagnosed as irritant dermatitis.
References