Perineal groove: an old, little known entity

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ABSTRACT

Introduction. Perineal groove is an infrequent midline malformation. It is a humid, mucosal, non-keratinized groove located at the perineal midline, extending from the vulvar fourchette to the anterior anal border. It is rare and usually asymptomatic, and it heals spontaneously in most cases. It is frequently mistaken for other malformations, which means correctly identifying it is essential to avoid iatrogenesis.

Clinical case. We present the case of a female newborn with an asymptomatic lesion at the perineal midline consistent with anorectal malformation. Following assessment by the Pediatric Surgery Department, she was diagnosed with perineal groove.

Discussion. Perineal groove is a little known malformation among healthcare professionals as it is infrequent and there are not many publications in the literature about it. This case demonstrates how important it is to keep this abnormality in mind to avoid erroneous diagnoses, unnecessary treatments, and family stress.

KEY WORDS: Groove; Perineum; Pediatrics; Perineal groove; Congenital abnormality.

SURCO PERINEAL: UN VIEJO POCO CONOCIDO

RESUMEN

Introducción. El surco perineal es una malformación infrecuente de la línea media. Se trata de un surco húmedo, mucoso y no queratinizado localizado en la línea media del periné desde la horquilla vulvar hasta el borde anal anterior. Es una malformación infrecuente, usualmente asintomática y de resolución espontánea en la mayoría de los casos. Esta anomalía es frecuentemente confundida con otras malformaciones por lo que su reconocimiento es fundamental para evitar yatrogenia.

Caso clínico. Se presenta el caso de una recién nacida con una lesión asintomática en la línea media del periné sospechosa de malformación anorrectal. Tras valoración por el Servicio de Cirugía Pediátrica se diagnosticó de surco perineal.

Comentarios. Debido a la infrecuencia y escasa documentación bibliográfica del surco perineal, esta malformación es desconocida para muchos sanitarios. Este caso expone la importancia de tener presente esta anomalía para evitar diagnósticos erróneos, tratamientos innecesarios y estrés familiar.

PALABRAS CLAVE: Surco; Periné; Pediatría; Surco perineal; Anomalía congénita.

INTRODUCTION

Perineal groove (PG) is an infrequent congenital midline abnormality, mostly predominant in girls. It was first described in 1968 by Stephens and Smith as a humid groove between the vulvar fourchette and the anus, without urethral or vaginal alterations. It can be either full – from the fourchette to the anal sphincter – or partial – the groove starts at the vulva without reaching the anus (superior), or at the anus without reaching the vulvar region (inferior)[(1)]. In most cases, this malformation is asymptomatic, but it may also be associated with pain, secretion, bleeding, infection, and constipation. It is typically isolated, and even though it has occasionally been described to be associated with a small number of syndromes, no statistical association has been documented. Physical exploration is key in terms of differential diagnosis, since it may be mistaken for other pathologies requiring invasive tests or unnecessary therapeutic procedures. This case demonstrates how important it is to keep this malformation in mind to avoid iatrogenesis and family stress.

CLINICAL CASE

We present the case of a full-term female newborn, with pregnancy under control and no gynecological or obstetric history. She was referred from the primary care department as a result of an asymptomatic perineal lesion which had

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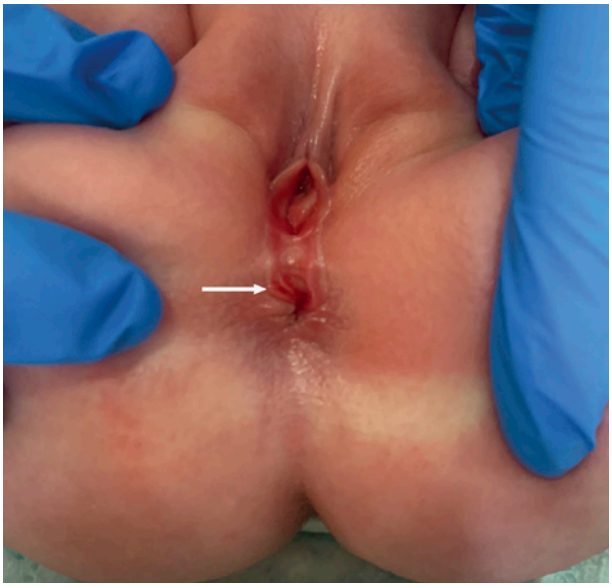

Figure 1. Full perineal groove (arrow) associated with anterior anus.

been treated as an anal fissure but had not responded to treatment. She also had constipation and suspected anorectal malformation. According to her patents, she had daily and spontaneous stools (Bristol 2-3), with no additional symptoms. The family was extremely worried and stressed facing diagnostic uncertainty and potential surgery. At exploration, the anus looked anterior, and a perineal lesion covered in mucosa from the anus to the vulvar fourchette was observed (Fig. 1). She was diagnosed with anterior anus and full PG, and a wait-and-see approach was decided upon. After 5 months of follow-up, she remains asymptomatic. Constipation has improved, and progressive epithelialization of the PG has been noted.

DISCUSSION

PG is an infrequent congenital abnormality, mostly predominant in girls, with a female:male ratio of 29.5:1[1-15]. Physiopathology remains unknown, but according to certain pathogenic hypotheses, it could be a result of defective uroanal septum or failed medial genital fold fusion[2,3,5]. The groove is asymptomatic in 80-90% of patients. When symptoms are present, the most frequent ones are constipation (16%) and defecation-related pain, which occurs in 1 out of 4 cases. Even though pathological association remains unclear, some cases associated with preeclampsia, gestational diabetes mellitus, placenta previa, and persistent ductus arteriosus (PDA), among others, have been described. However, these cases are rare, which prevents statistically significant associations from being established[4,7,8]. PG is typically isolated, but cases associated with hypospadias, bifid scrotum, PDA (5%), ectopic anus, imperforated anus, and urinary tract abnormalities have also been reported, especially in male patients[1,3,5]. This is consistent with our case, where anterior anus was present.

Diagnosis is primarily clinical, with no additional studies required in most cases. When additional tests are needed, they will be determined by lack of spontaneous closure and diagnostic suspicion. Thorough exploration of the perineal area allows the lesion to be identified and classified, and it is key in terms of differential diagnosis – anal fissure, trauma, abuse, infection, anorectal malformation (ARM), and genitourinary malformation –, since up to 5% of cases are initially classified as suspected sexual abuse[9,13,14]. In newborns, PG may be hard to distinguish from perineal fistula ARM. Therefore, if the patient remains asymptomatic, close surveillance is recommended, since the muscle complex is difficult to identify in neonates. If still in doubt, additional explorations will be required to establish an accurate diagnosis. Owing to the large differential diagnosis of PG and the confusion it generates, patients often receive all kinds of therapies. According to some reports, these unnecessary interventions range from pulsed laser as a result of suspected vascular abnormality, to surgery with removal and primary closure at birth owing to suspicion of peripartum neonatal tear[2,11]. Only 18% of PGs are diagnosed neonatally, and mean age at diagnosis is 14.6 months, which demonstrates how little known this malformation is[1-15].

Treatment is conservative, and surgical repair is only indicated out of cosmetic reasons or in patients without epithelialization at 2 years of age, with refractory symptoms, or scheduled for associated ARM repair[1,3]. Surgical treatment involves in bloc resection of the groove, layered closure, and application of topical cutaneous adhesives on the suture line to prevent contamination and dehiscence. PG prognosis is excellent, with spontaneous epithelialization occurring approximately at 1 year of age in 71% of patients – in some cases, epithelialization may not be observed until 4 years of age[1,4,6].

Even though PG was first described more than 50 years ago, it remains a little known pathology, especially as a result of the lack of bibliographic references. By correctly documenting this malformation in the literature, erroneous diagnoses, family stress, and unnecessary interventions will be avoided.

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