

Congenital Midline Perineal Groove: A Rare Defect of Perineum

Ghosh D¹, Farman L², Dawud B² and Al-Sharqi A^{3*}

¹Department of Surgery, Consultant Pediatric Surgeon, Sultan Qaboos University Hospital, Muscat, Oman

²Department of Surgery, Specialist Pediatric Surgeon, Sultan Qaboos University Hospital, Muscat, Oman

³Department of Surgery, Intern Doctor, Sultan Qaboos University Hospital, Muscat, Oman

*Corresponding author:

Ali Al Sharqi,
Department of Surgery, Intern Doctor,
Sultan Qaboos University Hospital, Muscat,
Oman, E-mail: Ali.sharqi95@gmail.com

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1. Abstract

Midline perineal groove is a rare congenital anomaly which is seen mainly in newborns females as a non-keratinized wet mucous membrane sulcus that extends from posterior fourchette of vaginal wall toward the anal verge.

A small number of cases (25 cases) have been reported in English literatures so far. This anomaly may be misdiagnosed with dermatitis, fungal infection or even sexual abuse in older children. The natural course of this condition is spontaneous healing. However, In the event of localized irritation, inflammation, infection, fistula or urinary tract infection surgical correction may be needed.

This article describes a new case of midline perineal groove in term baby girl.

2. Case Report

A term baby girl, was born via spontaneous vaginal delivery. Immediately after birth, she had seizures desaturation and bradycardia. On examination the child had hypotonia and multiple dysmorphic features including; depressed nasal bridge, micrognathia, midfacial hypoplasia, protruding tongue, widely opened anterior fontanelle and overriding of toes.

Echocardiography showed a small atrial septal defect and a small patent ductus arteriosus.

After that, she was referred to pediatric surgery team with a suspicion of an ambiguous genitalia.

The clinical examination of the perineum showed normal female genitalia and normally located anal opening. A non-epithelialized and non-keratinized wet sulcus started from posterior fourchette of the vestibule till the anterior anal verge was identified (Figure 1). Due to the dysmorphic features, the child was investigated further for genetics disease. From the diagnoses had been made, the child is on ongoing follow up with pediatric surgery team.



Figure 1: A non-epithelialized and non-keratinized wet sulcus started from posterior fourchette of the vestibule till the anterior anal verge

3. Discussion

Congenital midline perineal groove is a rare anomaly, seen mostly in newborn female, with only 2 male cases reported in literature. A small number of cases (25 cases) have been reported in literature so far [1].

The etiology, incidence and the pathogenesis is not well understood and rarely described in English literature. However, there are some theories regarding the embryology of this congenital defect including; defect in urogenital septum, failure of fusion of the median genital folds that forms the formation of perineum or as a remnant of open cloacal duct which give rise to this defect [6]. The literature confirms that congenital perineal groove is found in both females and males [4], so there must be an embryological reason of this anomaly which need to be investigated more for better understanding.

There are multiple risk factors for congenital midline perineal groove as reported in the previous studies including; age of the mother, mothers with conditions like Gestational Diabetes, Thalassaemia, placenta previa, preeclampsia, or some infections like Group B streptococci [1, 2].

The characteristics of perineal groove were described by Stephens and Smith in 1968 as a wet groove in mid perineum between the fourchette and the anus. There are 3 characteristics of congenital perineal groove: (1) a normal vestibular structures, like the vagina or urethra; (2) wet groove in the perineum between the fourchette and the anus; and (3) hypertrophy of the minoral tails which course posteriorly around the perineum to converge at the anus or surround it [3]. While usually an isolated anomaly, it has been reported to associate with disorders like, ectopic anus, perineal hypospadias and bifid scrotum [4, 5].

This congenital anomaly is diagnosed by clinical examination, but there is no specific classification. Shen et al narrated that the congenital midline perineal groove could be described into complete or incomplete defect. Complete midline perineal groove can be described as a defect starting from the posterior fourchette till the anus similar to our case. Alternatively, an incomplete type would end at the middle of the perineum [7].

Congenital midline perineal groove in most of cases is asymptomatic and may resolve spontaneously within 2 years so conservative management is preferred. However, surgical intervention is advised in case of failure of epithelization, recurrent localized infection, UTI or even cosmesis [6, 8]. The surgical intervention consists of excision of this wet non-keratinized mucosa and suturing the defect. These patient may need protective colostomy to avoid wound dehiscence due to urine or fecal contamination. One study, suggested the use of chemical glue prevents wound infection, believing it results in perfect cicatrization and can be used by parents at home or best is to apply as an in-patient [9].

The follow up is required in case of non-operative treatment because it may heal by epithelization within 1st two years of life and to avoid complications (infections of external genitalia & Urinary tract infection) and even the prognosis is excellent.

4. Conclusion

The diagnosis of this congenital midline perineal defect is clinical and can be misdiagnosed with other congenital anorectal or uro-rectal defects. Though the incidence of this anomaly is very low, but if misdiagnosed can have problems like, localized infection, cellulitis, or even may present with UTI. The prevalence is lower in males, only two cases are reported in English literature.

Generally, this condition may not need any surgical intervention and resolve by conservative treatment, but due to some reason surgical correction is uttermost. The understanding of this midline line congenital defect and proper counselling may avoid the untoward effects of the disease, like sever infections, or the worst mistaken as sexual assault. In case of conservative treatment careful follow up is necessary.

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