Scrotal pearl is not always a sign of anorectal malformation: median raphe cyst

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Pearls of meconium can be seen on the raphe of the scrotum and are considered as a sign of anorectal malformation (ARM). Scrotal pearls without ARM are rare in children and designated as median raphe cyst of the scrotum (MRC). A six-month-old boy with scrotal pearls without ARM is presented to discuss the clinical features and treatment modalities of MRC in infants.

Key words: scrotal pearl, anorectal malformation, median raphe cyst.

Pearls of meconium can be seen on the raphe of the scrotum and are considered to be a sign of low presentation of an anorectal malformation. Scrotal pearls without an anorectal malformation are very rare in infants and designated as median raphe cyst (MRC) of the perineum. MRC is an uncommon congenital lesion of the male genitalia and is described anywhere from the ventral aspect of the urethra to the anus in the midline position[1]. Diagnosis of MRC is rare in childhood[2]. Since most of the cases in children are asymptomatic, it is suggested to be more common than reported. Most of the cysts tend to grow as the child grows and become symptomatic in adolescence and adulthood.

A six-month-old boy with scrotal pearls but without an anorectal malformation is presented to discuss the clinical features and treatment modalities of MRC in infants.

Case Report

A six-month-old boy was referred by a pediatrician with a complaint of whitish pearls in the midline of the scrotum to differentiate an associating anorectal malformation. The boy had neither a remarkable history of constipation nor difficulty in stooling. On physical examination, he had canal-like multiple whitish cysts with a width of 2-5 mm on the entire line of the scrotal raphe (Fig. 1). There were no penile or perineal cysts. The anorectal and genitourinary examination revealed normal findings. Ultrasonographic evaluation of the testes and lumbosacral X-ray for spinal lesions revealed normal findings. Since the patient was asymptomatic and there was a possibility of spontaneous regression, follow-up without surgical intervention was planned. He has been followed for the last two months. The lesions showed neither complication nor regression.

Discussion

Median raphe cyst (MRC) of the perineum is a rare anomaly that usually presents with solitary, multiple or canal-like watery cysts[1]. They can develop at any site along the midline ventral aspect of the male genital tract, ranging from the meatus to anus[2]. Since most of the cases are asymptomatic in childhood, the true incidence of MRC in infants is not known. In

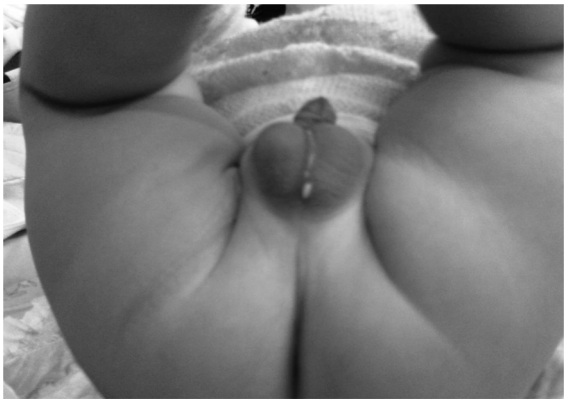

Fig.1. A 6-month-old boy presenting with MRC.

a large series of patients, 25% of cases were younger than 10 years of age, and only one infant was reported to have a scrotal MRC[3,4]. Median raphe cysts (MRCs) of the perineum are considered congenital defects of embryonic development of the male genitalia during fetal life. Two explanations have been proposed for the embryonic origin of the lesions. One is the development of embryonic outgrowths of the epithelium after primary closure of urethral or genital folds[2]. The other theory regards epithelial trapping after incomplete closure of genital folds[5]. We suggest that scrotal pearls in anorectal malformation probably have a similar embryologic origin. Instead of epithelial overgrowth, meconium may be trapped before closure of the genital folds via an anocutaneous fistula in anorectal malformation.

These theories were confirmed by histopathologic evaluation of the cysts, and three distinct histopathologic patterns have been described for MRC. These patterns include urethral type lined with pseudostratified columnar epithelium (uroepithelium), epidermoid type with squamous stratified epithelium, and mixed type with both epithelia[2]. The most common type is reported as urethral, followed by the epithelial and mixed forms[6].

Most MRCs are asymptomatic in childhood and become symptomatic as the child grows. Swelling, tenderness and purulent discharge can be seen when cysts become traumatized and secondarily infected[7,8]. The cysts localized in the scrotum and perineum can be confused with scrotal and perineal meconium pearls that are detected in anorectal malformations. As in our case, anorectal malformation should be excluded with careful physical examination, and the parents should be clearly informed about the benign course of these cysts. Since meconium pearls with anorectal malformations are detected in the early neonatal period, MRC should be kept in mind when scrotal pearls are encountered in the midline raphe during infancy. The differential diagnosis includes molluscum contagiosum, syringoma, urethral diverticulum, and pilonidal cysts[9]. These lesions can be differentiated easily from MRC with other systemic findings.

The treatment of choice is simple excision of the lesion followed by primary closure[4,6]. In asymptomatic children, surgical intervention can be postponed. Since spontaneous regression of the cysts was reported, small and asymptomatic cases in infants can be observed without surgical excision. Although marsupialization and unroofing were used in adults, surgical complications and poor cosmetic results have made these options less favorable. Although no malignant potential was observed during the long-term follow-up, close observation should be planned in small infants for possible complications, such as infection.

In conclusion, MRC is a rare congenital midline cyst of the male genitalia and can mimic scrotal pearls in infants with anorectal malformations. Careful physical examination of the anogenital region should be performed to differentiate an associating anorectal malformation. Close follow-up without surgical intervention can be performed in all asymptomatic cases. MRC should be kept in mind in infants with scrotal pearls in the absence of anorectal malformations.

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