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Median Raphe Cyst: A Rare Lesion with unusual localisation

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Abstract
Median raphe cyst is an uncommon developmental anomaly of male genitalia. Only a few hundred cases have been published in the English literature and the lack of awareness of this entity can lead to confusion and misdiagnosis. Raphe cysts are generally solitary and translucent, and can develop at any location over the midline of the external genitalia from the parameatal region to the perineum. The cysts can occur at parameatus, glans penis, penile shaft, scrotum or perineum. The penile shaft is the more common location whereas perineum is least common site. We report here a case of median raphe cyst located in the midline over perineal region of 6 month old infant. Early recognition of this condition may avoid the confusion in diagnosis at later age, when patient present with complications.

Introduction

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]. It appears commonly in childhood or adolescents. In most patients it is usually asymptomatic or unrecognized during childhood. The cysts become symptomatic with advancing age due to infection or trauma, which make diagnosis difficult. The cysts can occur at any site including parameatus, glans penis, penile shaft, scrotum, or perineum. Presentation of median raphe cyst in the perineal region is exceptional[2].

Case Report

A 6-month-old otherwise healthy infant was brought by his parents. He exhibited a solitary translucent cord-like lesion in perineal region. Cutaneous examination showed a linear, soft, painless cord-like cyst present along the midline in perineal region (Figure 1). There were no features of inflammation. The anorectal and genitourinary examination revealed normal findings. Ultrasonographic evaluation of genitourinary system 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 next six months. The lesions showed neither complication nor regression.

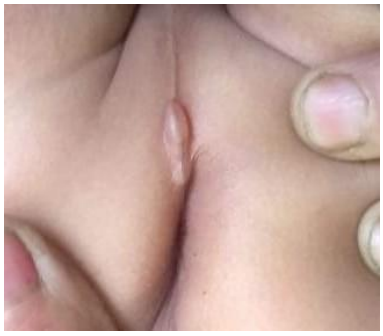

Figure 1 Median Raphe Cyst

Discussion

Median raphe cyst is a congenital lesion occurring anywhere in the midline between the external urethral meatus and anus[1]. The other terms used for the median raphe cyst include the mucoid cyst of the penile skin, the genitoperineal cyst of the median raphe, and the parameatal cyst[3]. They are usually asymptomatic lesions detected in the first decade of life with symptomatic clinical presentations typically occurring in the second and third decades of life. In children, symptoms might include dysuria, frequency, redirected urinary stream, infection and hematuria[4]. In some cases, the cyst may grow rapidly following a period of relative indolence and can even become inflamed or traumatized, complicating diagnosis[4][5].

The penile shaft is the more common location at presentation, regardless of patient age[6]. This is likely due to the lesion being readily identified by direct inspection and the reported increase in symptoms associated with parameatal and penile lesions[4][6]. Median raphe cyst is an embryological developmental defect of the male genital apparatus. Male genital differentiation begins after the tenth week of the embryological life due to androgenic stimulation. Subsequently, the urethral groove of the endodermal origin elongates parallel to the growth of the penis. The urethra forms at around the third month, largely as a result of the fusion of the urethral folds enveloping the urethral groove. Its most distal portion develops during the fourth month from a group of cells of the ectodermal origin in the glans tip[7]. The diagnosis of median raphe cyst is difficult, but needs to be differentiated from other conditions such as epidermal cyst, steatocystoma, glomustumor, dermoid cyst, urethral diverticulum, and pilonidal cyst when it presents in the penile (most common site) and scrotal region[3]. Most commonly, the diagnosis of median raphe cyst is established postoperatively on histogolical and immunohistochemical studies. The epithelial lining of median raphe cyst includes columnar stratified, pseudo stratified, or squamous cells, correlating with histology in different portions of male urethra[8]. Management of median raphe cyst depends on clinical presentation. Observation in asymptomatic patients is acceptable and spontaneous resolution of the lesion has been reported[6]. For symptomatic patients or those seeking a cosmetic resolution, surgical excision with primary closure is the definitive treatment. In our case since child was asymptomatic, so we decide to keep the child under follow up.

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