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Median raphe cyst: report of two cases

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Abstract

Median raphe cysts are rare congenital lesions of the male genitalia that occur as a result of altered embryologic development. We report two such cases of median raphe cysts in the pediatric age group. In addition, we review the literature.

Keywords: median raphe cyst; perimeatal cyst; penile cyst

Introduction

Median raphe cysts are uncommon congenital cysts, located anywhere from the tip of the penis to the anal orifice [1]. These are thought to arise from abnormal or incomplete development of the paired genital folds. Previously, this entity has been reported as "mucoïd cyst," "genitoperineal cyst," "perimeatal cyst," "hydrocystoma," and "apocrine cystadenoma" [2]. We hereby report two cases of median raphe cysts in the penile and perimeatal positions, respectively.

Case Synopsis

Case 1

A 9-month-old otherwise healthy infant was accompanied to the Dermatology clinic by his parents. He exhibited a solitary cord-like lesion in groin. It was a tiny fluctuant elevated lesion at birth, which showed a gradual increase in size to attain the present dimension. Cutaneous examination showed a linear, soft, painless cord-like cyst present along the midline extending from scrotum to anus (**Figure 1**). There were no features of inflammation. The rest of the regional, mucocutaneous, and systemic examination were unremarkable.



Figure 1. Canal-like soft cystic swelling in perineum extending from scrotum to anus in midline position.

Case 2

A 12-year-old boy presented with two fluid filled lesions near the urethral opening, present since birth. The lesion had not changed its morphology or dimension, since its development. Observation revealed two smooth, soft, cysts located near the



Figure 2. Two translucent cystic lesions in parameatal position.

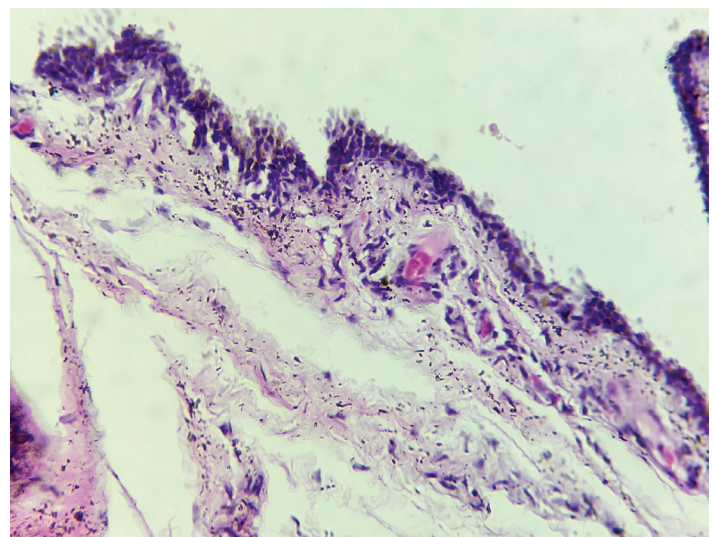
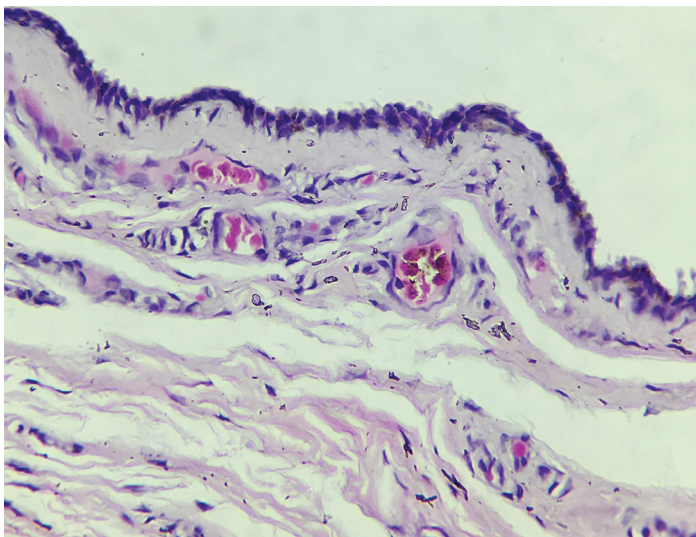
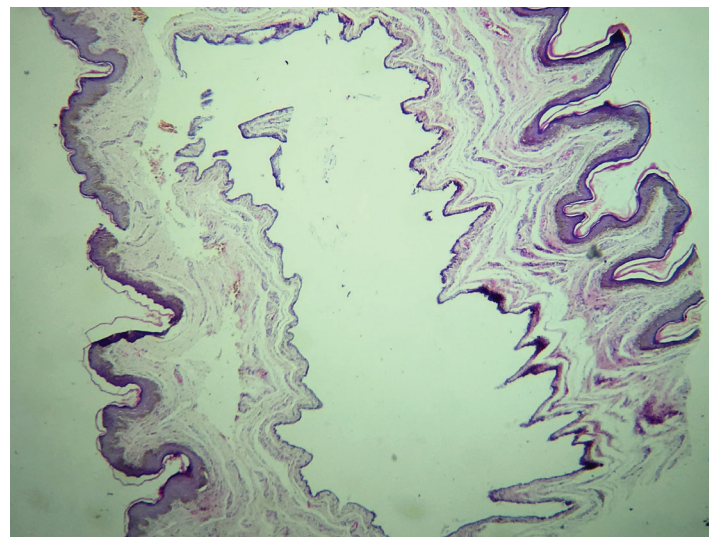
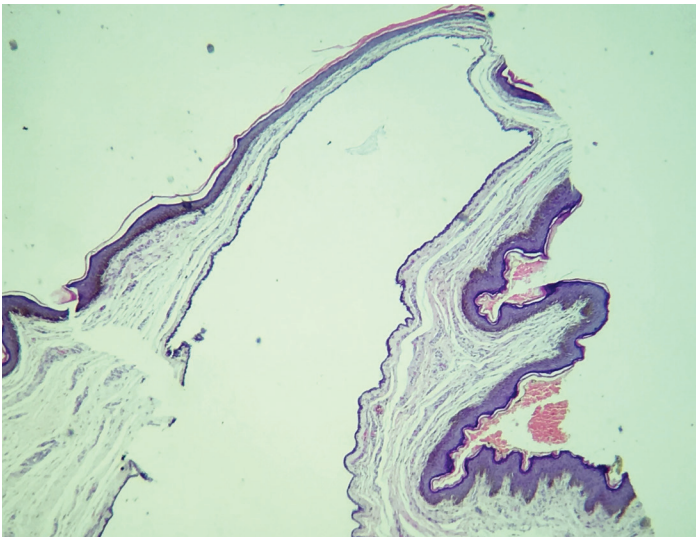


Figure 3. A) Sample from case 1 showing solitary cystic lesion in dermis, lined by pseudostratified columnar epithelium of varying thickness (H&E, 40x). B) Higher magnification (H&E, 400x).

Figure 4. A) Sample from case 2 showing single cystic lesion in dermis, lined by pseudostratified columnar epithelium of varying thickness (H&E, 40x). B) Higher magnification (H&E, 400x).

urethral meatus (**Figure 2**). Of note, the external urethral meatus was not obstructed. The rest of the regional, mucocutaneous, and systemic examination were non-contributory.

Histopathology from both patients showed similar findings and showed a solitary unilocular cystic cavity in the dermis, lined by a pseudostratified columnar epithelium of varying thickness and with occasional large, mucinous cells (Figures 3a and 3b for case 1; Figures 4a and 4b for case 2). The lumen appeared empty. Atypia and inflammatory infiltrate were absent. Based on histopathological findings both cases were diagnosed as median raphe cysts (MRC). Our case 1 had an unusual canal-like presentation. Our case 2 had two MRC lesions in a perimeatal position.

Case Discussion

Median raphe cysts (MRC) develop during the fusion of urethral folds as a result of tissue moving outside the urethral groove, leading to formation of abnormal outgrowths (cyst or canal-like lesions). The mean age at presentation is 26 years with a bimodal distribution [3]. Classically, MRC presents as asymptomatic solitary or multiple cysts, or canal-like lesions in the ventral midline part of the penis and perineum. They can be located anywhere from the external urethral meatus to the anus, in a midline position. MRC tend to grow as the child grows and therefore clinical symptoms are more likely to occur in adulthood. One interesting finding by Shao et al. was a direct correlation between the location of the cysts and the manifestation of the symptoms, with the more distally located cysts causing more interference in daily activities [3].

Complications include secondary infection and interference with sexual activity, if sufficiently large [1,4]. Atypical presentations include median raphe cysts of the penis with melanosis [5], cordlike or canaliform induration in the median raphe [6], similar to our case 1, and sudden presentation simulating sclerosing lymphangitis [7]. Clinical conditions in the differential diagnosis include glomus tumor, dermoid cyst, pilonidal sinus, epidermal inclusion cyst, urethral diverticulum, steatocystoma, condyloma, viral wart, hemorrhoid, hypertrophied papilla, and neoplasias [8].

The histology of MRC shows an irregular cystic cavity (not connecting with urethra), lined by an epithelium. The nature of lining epithelium depends on the type of tissue trapped. If the trapped tissue contains urothelium cells, the cyst would contain urethral epithelium. If non-keratinizing squamous epithelial cells in the distal urethra are trapped, the histology is epidermoid. If the trapped tissue includes the periurethral glands, the appearance on histology is glandular. When more than one type of tissue gets trapped, we find cysts with mixed epithelia [1,4]. Based on the histopathology, they can be classified into 4 types: urethral, epidermoid, glandular, and mixed. The urethral type is the most common type (55%), and it is characterized by a urothelium-like epithelium. The epidermoid type accounts for only 5% of cases. The glandular type (3%) consists of intraepithelial glandular structures. The mixed type (36%) consists of more than one type of epithelium [1], (urethral epithelium with partial squamous metaplasia, urethral epithelium with scattered or isolated mucinous cells). Histological variants include presence of both ectodermal and endodermal epithelium [9] and demonstration of ciliary cell metaplasia [10].

The natural history of the entity can be diverse. It may grow with age until adulthood, when it presents as a solitary asymptomatic nodule. Rarely, as a result of trauma or infection or even in the absence of any inciting factor, it may show a rapid increase in size [4,11]. Spontaneous regression has been reported rarely [8]. The best therapeutic option is excision followed by primary closure. However, observation can be considered for patients with small, asymptomatic lesions and those who are not

willing to undergo surgery [1,4,8].

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