Case Report

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Mixed Type of Median Raphe Cyst Presenting as Canaliform Variant: A Rare Case Report

Pathology Section

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ABSTRACT

Median raphe cysts are benign congenital developmental anomalies that can occur at any site along the midline of the ventral aspect of external genital region. Their pathogenesis is unclear. They are thought to be arising from defective closure or epithelial trapping during embryological development of urethral folds. They are rare with less than 15 cases being reported in the Indian subcontinent. Here is a report of one such case in a three-year-old child presenting with difficulty in passing urine and also difficulty in passing stools. On examination, there were multiple cystic lesions arranged in linear fashion along the median raphe from perineum to penile shaft since birth. Ultrasonography showed multiple isoechoic cystic lesions from the middle of shaft of the penis to perineum, very well separate from urethra, lying entirely within the mucosa without any evidence of solid components, septations or vascularity. Surgical excision was performed. Histopathological examination showed multiple, canaliform median raphe cysts with mixed type. Differential diagnosis on histopathology includes apocrine cystadenoma, mucinous cysts, epidermal cysts and dermoid cysts. These cysts are best treated by complete surgical excision to prevent recurrence. Canaliform variant of median raphe cysts are uncommon and rarely reported. They have bimodal age distribution. Histopathological confirmation is essential for accurate diagnosis. Complete excision remains the treatment of choice. Timely diagnosis and surgical excision prevent further complications.

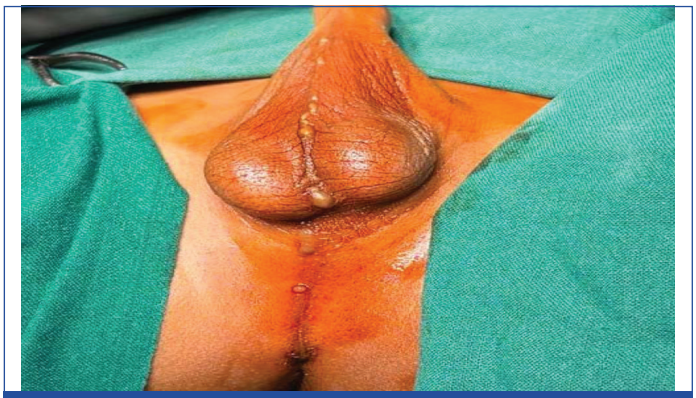
Keywords: Canaliform, Penile shaft, Perineal raphe

CASE REPORT

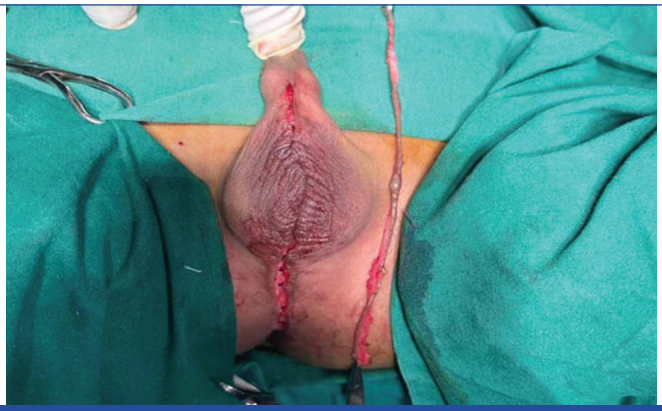
A three-year-old male child presented with a one-week history of difficulty in micturition, along with a six-month history of constipation.

The perineal examination showed multiple cystic lesions arranged in linear fashion along the median raphe from perineum anterior to anal verge to terminal part of penile shaft. These were well defined, multiple, small, tan-white, canal like cystic lesions measuring 0.5-1 cm [Table/Fig-1]. The cysts were soft and non-tender on palpation. The scrotal and testicular examination was normal with bilateral normal size for age, and bilaterally descended testes in scrotum. The rectal and per rectal examination was normal with no mass/growth, no fissure or any other lesion. The urethral meatus was at normal site and of good size. Rest of the perineum, sensations and bulbocavernosus reflex were normal. The lesions were present since birth which progressively increased in size and number. There was no history of trauma. No signs of infection or discharge were noted. There was no medical history nor any history of illness or admission for any ailment earlier or now, except of the present symptoms and the disease. The ultrasonography showed multiple isoechoic cystic lesions from the middle of shaft of the penis to perineum very well separate from urethra, and lying entirely within the mucosa without any evidence of solid components, septations or vascularity. A diagnosis of median raphe cyst was made based on clinical and ultrasonographic findings.

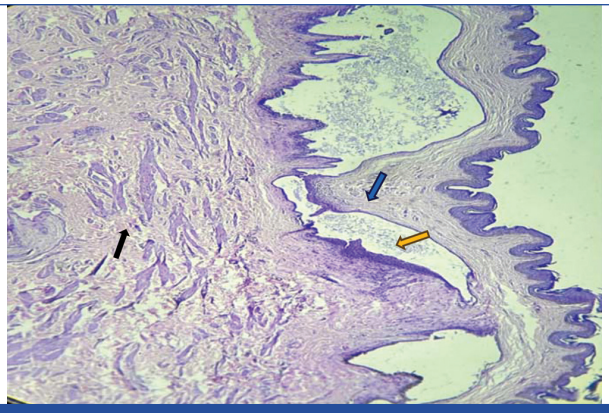
The surgically excised specimen was submitted for histopathological examination, which revealed a skin-covered, linearly arranged cluster of multiple thin-walled cysts, measuring 7 cm in length [Table/Fig-2]. On histopathological examination, the differential diagnosis was median raphe cyst and epidermal cyst. Microscopy showed skin covered cystic lesions with subcutaneous tissue showing fibromuscular connective tissue and moderate chronic inflammatory infiltrate comprised of lymphocytes, few macrophages and occasional plasma cells. Adnexal structures were not evident [Table/Fig-3]. The cysts showed variable lining of squamous epithelium with keratin debris to stratified cuboidal to flattened epithelium with foci showing apocrine snouts at the cuboidal lining. Based on these findings, a diagnosis of mixed-type median raphe cyst was established [Table/Fig-4,5]. At 3-months postoperative follow-up, the child was doing well with a well-healed surgical wound and had no surgery-related symptoms, urinary complaints, or defecation-related issues.



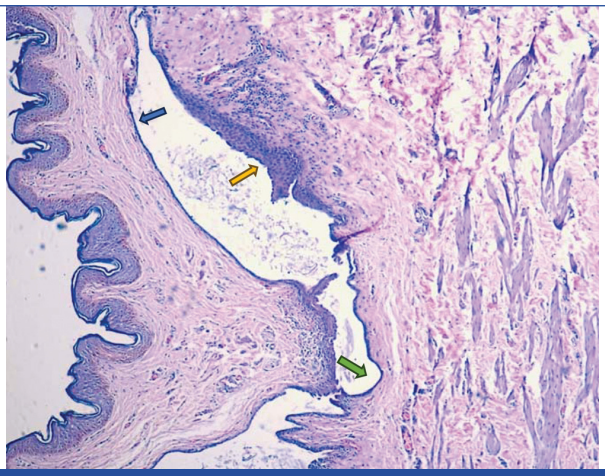
[Table/Fig-1]: Multiple, small, tan-white, canal like linear cystic lesions.



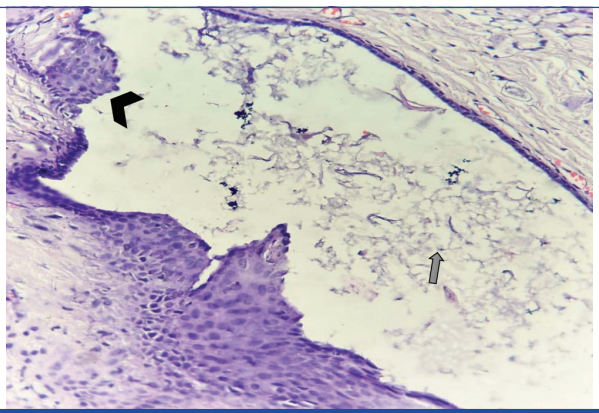
[Table/Fig-2]: Surgically excised skin covered linearly arranged, multiple cysts.



[Table/Fig-3]: Photomicrograph revealed skin covered multiple cystic lesions showing variable lining (coloured arrows) with fibromuscular connective tissue in the subcutis (Black arrow). (H&E, x100).



[Table/Fig-4]: Cyst showing variable lining of squamous epithelium (Green arrow) to stratified cuboidal (Yellow arrow) to flattened epithelium (Blue arrow) (H&E, 200x).



[Table/Fig-5]: Cyst showing mixed type of lining epithelium, with foci showing apocrine snouts (Arrow head) at cuboidal lining and keratin debris within the cyst lumen (Grey arrow) (H&E, 400x).

DISCUSSION

Median raphe cysts are rare, benign congenital developmental anomaly that can occur at any site along the midline of the ventral aspect of external genital region [1]. They are genital cysts previously designated as genitoperineal raphe cysts, hydrocystoma or parameatal raphe cysts. They were first described by Mermet in 1985 and are thought to be congenital defects as a result of altered embryonic development [2,3]. These lesions can extend from urethral meatus to the anus and the perineum along the perineal raphe. They have bimodal age distribution, presenting between 1-10 years and 21-40 years [4]. Less than 200 cases have been reported worldwide. Among these, less than 15 cases have been reported in the Indian subcontinent [5].

The presentation of median raphe cysts can vary depending on the location, size, number of cysts, histological type and patient age [6]. They classically present as a solitary asymptomatic lesion in the ventral midline part of penis and perineum. Atypical presentation includes multiple, canaliform or cord like cysts [7]. A largest retrospective study in current relevant literature by Shao IH et al., reported 56 median raphe cysts in 55 patients with a mean age of 26.7 years. The lesions were predominantly located on the penile shaft (42.9%) and parameatal region (33.9%), with less frequent involvement of glans penis (7.1%), prepuce (12.7%), and scrotum (3.6%). None of these cases showed involvement of the perineal raphe, highlighting the rarity of the present case [6]. Lee JN et al., described a rare case of a 26-year-old male presenting with canaliform median raphe cysts involving the scrotal and perineal regions with recurrent infection and on histology lined by stratified squamous epithelium, which is different from the present case in terms of age and lining epithelium [8]. Shin S et al., reported a case of 7-month infant with canaliform median raphe cysts having mixed type of epithelium involving scrotal and perineal raphe, which is similar to the present case [1].

Diagnosis of MRCs is rare in childhood, since majority of the cases in children are asymptomatic or unrecognised. Most of the cysts tend to develop rapidly as the child grows and become symptomatic in adolescence and adulthood in the setting of infection and trauma. Some children might present with painful swelling, dysuria, frequency, haematuria. Cysts might cause problems such as discomfort during sexual intercourse, difficulty in micturition, and cosmetic discomfort [4,7]. In the present case the symptoms were mild and were due to the proximity of the cyst to the urethra and their recent onset acute enlargement and were treated and recovered with oral medication. Constipation was related to abnormal diet and treated with diet regimen and oral laxatives.

The pathogenesis of median raphe cysts is still unclear. Three hypotheses have been proposed. First, the tissue trapping theory states that they are caused by either defective/incomplete fusion of the urethral folds or an anomalous epithelium outgrowth following primary closure during the process of development of urethra. Second theory states that they may result from the anomalous development of periurethral glands of Littre. The third hypotheses suggest it may be due to blockade of paraurethral ducts causing cystic dilatation and retention of secretions [1]. The clinical differential diagnosis for median raphe cysts includes steatocystoma, pilonidal cyst, dermoid cyst, epidermal inclusion cyst and urethral diverticulum [5]. The first three lesions usually present as skin covered dermal masses with pilonidal cyst showing frequent purulent discharge. Epidermal inclusion cyst usually occurs following trauma and urethral diverticulum presents as pseudocyst secondary to trauma, infection and stricture [9].

Imaging studies have a very limited role in diagnosis. Ultrasonography (USG) and Magnetic Resonance Imaging (MRI) can help in determining the anatomical extent of the cyst. Urethrogram shows absence of communication between the cyst and urethra [2].

Histopathologically, the epithelial linings of median raphe cysts are classified into four types which may reflect the cell type in the trapped tissue. “Urethral type” the most common form (55%) is comprised of pseudostratified columnar or urothelium - like epithelium. “Mixed type”, the second most common form (36%) comprises of more than one type of epithelium such as urethral epithelium with squamous metaplasia or mucinous cells. The “Epidermoid type” (5%) comprised of stratified squamous epithelium and “Glandular type” (3%) comprised of urethral epithelium with intraepithelial glandular structure [1,6,7]. The differential diagnosis on histopathology are apocrine cystadenoma, mucinous cyst, epidermal inclusion cyst and dermoid cyst [5,10]. Apocrine cystadenoma shows lining consisting of inner cuboidal epithelial cells with apical snouts and an outer layer of flattened myoepithelial cells. Epidermal inclusion cyst and dermoid cyst are lined by keratinised squamous epithelium having granular layer and arefilled with keratin. In addition, dermoid cyst shows presence of adnexal structures [9].

The definitive treatment for symptomatic lesions is complete local excision with primary closure to prevent possible complications such as infection and abscess formation, rupture of the cysts, fistula formation, voiding dysfunction, sexual interference and recurrence. In asymptomatic cases, cysts may resolve spontaneously without any surgical intervention [1,10,11].

CONCLUSION(S)

Median raphe cysts represent rare congenital developmental anomaly. Canaliform variant is uncommon and is rarely reported. Due to its rarity, diagnosis can be challenging and thus requires histopathological confirmation. The present case had cysts which were multiple, canaliform with scrotal and perineal localisation and on histology showed mixed type of epithelium. Removal of cysts is safer to prevent future complications.

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