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Case Report

Canaliform Median Raphe cyst: A rare variant of a rare diagnosis

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Abstract Median raphe cyst are rare lesions, with fewer than 15 case reports from the Indian subcontinent. They are usually present over the midline and ventral aspect of the penis, usually near the glans. They can be present over the glans, penile shaft, scrotum or perineum. Although present since childhood, they are usually reported by young adults when they increase in size, get infected or cause sexual discomfort. The typical location of the cysts helps in pin pointing the diagnosis. Although asymptomatic, in case of complications like infection, trauma or discomfort they should be referred to an Urologist. **Case Report:** A 4 year old male patient, presented with multiple cystic, fluctuant, opaque swellings present over the midline of the scrotum, ranging in size from 0.2cmX0.3cm to 0.5cmX0.5cm. They were present since birth and asymptomatic. No other pathology was detected in the genital or perianal area. As the lesion was not any source of discomfort, the mother was offered only reassurance and asked to follow up in case of any complications. **Discussion:** Median Raphe cysts are rare benign, congenital lesions over the male external genitalia. Histology demonstrates cysts lined by stratified columnar epithelium with no connection to the overlying epithelium. Differentials include epidermal cysts, pilonidal cysts, dermoid cysts etc. The pathogenesis is not fully understood, theories include defective urethral fold closure and ectopic periurethral glands among others. Surgical excision with primary closure leads to definitive cure.

Key words
Median raphe cyst, perineum, parameatal cyst, canaliform.

Introduction

Median raphe cysts are benign, infrequently reported cystic lesions. Fewer than 15 cases have been reported from the South Asian subcontinent.[1] They are present along the midline from urethral meatus to the anus. Rarely para-urethral cases have been documented.[2] Most cases report solitary cysts. The condition was first described by Mermet in 1895.[3] They are divided clinically into classic, nodular, linear, canalicular, multi-cystic and pigmented types. Histologically the variants include urethral, glandular, epidermoid and mixed types[4]. The exact pathogenesis of the formation of the cyst is unknown. Although, defect during embryonic development is believed to the most likely cause. Typical location and clinical appearance help in clinching the diagnosis, which can be confirmed with histopathology. Here, we report a case of Median raphe cyst in a 4 year old boy.

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Case report

A 4 year old boy presented to our OPD along with his mother. She complained of multiple small, skin colour to whitish cystic swellings along the midline of the genitalia of the child **(Figure 1)**. They started along the dorsal aspect of the scrotum and continued along the perineum posteriorly. She had noticed them shortly after birth. The swellings had gradually been

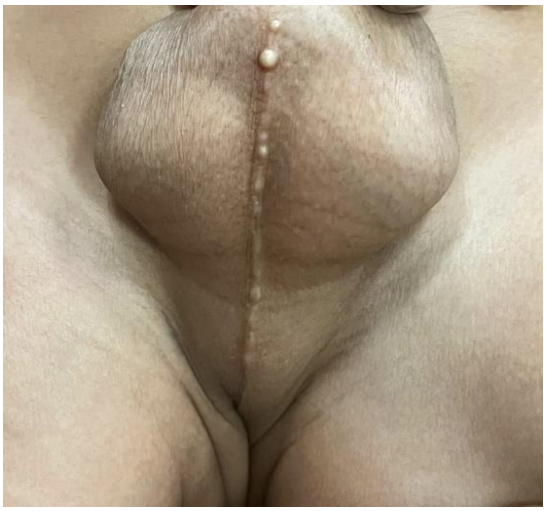

Figure Multiple small skin colored to whitish cystic swellings along the midline.

increasing in size as the child was growing. There were no complaints of any discharge, pain, itching, burning, tenderness or oozing. The swelling did not interfere with urination. There was no history of trauma or application of any topical medication. There were no other diagnosed congenital anomalies or medical illness in the child.

On examination, there were multiple, soft, opaque, cystic swellings; ranging in size from 0.2cmX0.3cm to 0.5cmX0.5cm (as seen in the image). The overlying surface was white to skin colored and shiny. The surrounding skin was normal. The cysts were soft, fluctuant and non-tender on palpation. The genitalia, perineum and groin area were otherwise unremarkable. The inguinal lymph nodes were non palpable.

Discussion

Median raphe cysts although present since birth or early childhood are usually not reported at this age due to their asymptomatic nature. They are reported by young adults in case of trauma or infection which leads to pain or sexual discomfort, or in case of cosmetic disfiguration. The parameatal cysts may cause urinary obstruction.[4-6] The more distal the cyst and earlier the age at presentation, the greater the chances of it being symptomatic.[5] In our case the child was 4 years old and asymptomatic.

Majority of the cases are solitary. In our case there were multiple cord like swellings along the median raphe which probably point towards the rare variant- canaliform.[7] This can be confirmed through histopatholgy. Under rare circumstances solitary and canaliform lesions may coexist.[8]

Multiple pathologies have been suggested which include defective closure of the median raphe,[9] para-urethral duct occlusion[9] and sequestered ectopic peri-urethral glands of Littre.[10]

The histopathology of the cyst depends on the site of the origin of the cyst. The lining is pseudostratified columnar if the cyst is arising from the proximal urethra, glandular in case of paraurethral glands and squamous in case of distal urethra.[12,13] Mixed lining has been described in case of multiple cysts arising from different locations. Shao *et al.* have also described the lesions as urethral, epidermoid, glandular and mixed.[4] Ciliated and melanised variants have been added by Syed *et al*.[14] IHC staining in most cases is positive for Cytokeratin 7,[15] which demonstartes urethral origin.

The diagnosis is mostly clinical and is confirmed histologically. USG and MRI have been performed in certain cases, they help determine the size, anatomic extent and communication with underlying structures. The main differential diagnosis include steatocystomas, dermoid cysts, eccrine cystadenomas, pilonidal cyst, urethral diverticulum, and steatocystomas.[5]

Upon diagnosis the case should be referred to an Urologist as complete excision along with the cyst wall and primary closure is the treatment of choice. Other treatment options like aspiration,

deroofing, marsupilization may lead to recurrence or unsatisfactory results.

Conclusion

Median Raphe cyst is a rare condition. The typical site and morphology help with the diagnosis. This case is rare as there are multiple lesions, present over the scrotum and most likely it belongs to the canaliform type.

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