



## PENILE MEDIAN RAPHE CYSTS: SERIAL RETROSPECTIVE ANALYSIS.

## Urology

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## ABSTRACT

**INTRODUCTION:** Median raphe cysts are very rare midline cysts resulting from anomalies in the development of the urethral groove and may occur anywhere in midline from the urinary meatus to anus. It was first described in detail by Mermet in 1895. In general the lesions are asymptomatic and does not interfere with urinary or sexual function. We present 7 cases of penile median raphe cyst including 1 case of exceptionally large prepuccial cyst (largest size ever reported) on account of their rarity and interesting histological findings.

**METHOD:** The medical records of patients who were diagnosed with median raphe cysts between January 2001 and December 2015 were reviewed retrospectively to include 7 cases. Age of patients varied from 3 to 50 years. All patients presented with asymptomatic, slow growing, non tender, soft cystic swelling over glans penis measuring 1.5 x 1 to 2.5 x 2 cm size except in a case of 50 year male who presented with large prepuccial swelling of 6x3cm, smooth, cystic, non tender which was present since childhood and had gradually enlarged leading to difficulty in voiding and performing intercourse. All patients of glanular swelling were managed with excisional biopsy while circumcision combined with cyst excision done in case of prepuccial swelling.

**RESULT:** All patients had uneventful post operative period and were discharged from hospital on same day. HPE revealed cystic lesion lined partly by stratified squamous epithelium, partly by transitional epithelium as well as pseudostratified columnar epithelium suggestive of median raphe cyst. Follow up period ranged from 6 to 36 months with no recurrence or any other unexpected outcome in any case.

**CONCLUSION:** Median raphe cysts are rare, benign and usually asymptomatic except in case where they attain very large size. Local examination is sufficient to make a diagnosis in majority of cases, and complete surgical excision is necessary to obtain good cosmetic results without recurrence.

## KEYWORDS

## INTRODUCTION

The median raphe of the genitoperineum is a permanent surface marking formed during the embryonal development of the urethra and cystic lesions developing in relation to this raphe have been classified as median raphe cysts.<sup>1,2</sup> It was first described in detail by Mermet in 1895.<sup>1,2</sup> An extensive literature search has revealed only less than 200 reported cases and among these, only less than 10 have been reported from the Indian subcontinent. Median raphe cysts can develop anywhere in the midline on the ventral side of the male genital area, from the prepuccial orifice to the anus. The most common location of such cysts is the penile shaft and the paramental position. Very rarely, prepuccial median raphe cysts have been reported.<sup>2,3</sup> These cysts does not communicate with the urethra and is lined by pseudostratified columnar or stratified squamous cell epithelium, mimicking the epithelial lining of the male urethra.<sup>4,5,6</sup> They are generally solitary lesion and measures only a few millimeters in diameter.<sup>7</sup> However, they may extend over several centimeters in a linear fashion. In general the lesions are asymptomatic and does not interfere with urinary or sexual function but can be complicated by infection or trauma.<sup>4,5</sup> Only a few cases of penile median raphe cysts have been described that to as case reports, mostly in dermatology journals rather than urology publications. However, serial reports of such cases are rare.

We present retrospective analysis of 7 cases of penile median raphe cyst including 1 case of exceptionally large prepuccial cyst (largest size ever reported, figure 1) on account of their rarity and interesting histological findings.

## PATIENTS &amp; METHOD

The medical records of patients who were diagnosed with penile median raphe cysts between January 2001 and December 2015 were reviewed retrospectively to include 7 cases. Patients with skin tags or cysts located anywhere other than the median raphe were excluded. Relevant data recorded and analysed in view of patients age, clinical presentation, characteristics of the cysts, treatment employed, histopathological findings, and patients outcome. Age of patients varied from 3 to 50 years with mean age 14.5 years and median age 10 years. All patients presented with asymptomatic, slow growing, non

tender, soft cystic swelling near meatus measuring 1.5 x 1 to 2.5 x 2 cm in size (figure 2) except in a case of 50 year male who presented with large prepuccial swelling of 6x3cm, smooth, cystic, non tender which was present since childhood and had gradually enlarged leading to difficulty in voiding and performing intercourse (figure 1). All patients of glanular swelling were managed by excisional biopsy with primary closure (figure 3) while circumcision combined with cyst excision was done in case of prepuccial swelling in view of narrow prepuccial opening (figure 4).

FIGURE 1



Prepuccial cyst (50 yr): 6x3cm, located on ventral aspect Demonstration of meatus Catheter in situ

FIGURE 2



Penile median raphe cyst Paramental cyst Paramental cyst with catheter

FIGURE 3



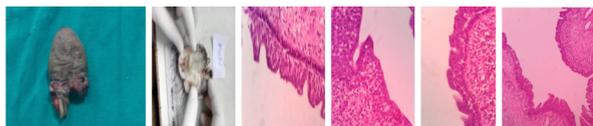
FIGURE 4



## RESULT

All patients had uneventful post operative period and were discharged from hospital on same day. HPE in all the cases of glanular cyst revealed cystic lesion lined by stratified squamous epithelium as well as pseudostratified columnar epithelium. While histopathological examination of prepuccial cyst showed gross specimen of 6 x 3 x 2 cm, which was soft to spongy in consistency and was covered with skin. The lesion was confined to dermis and in addition to lined by stratified squamous epithelium, partly by pseudostratified columnar epithelium was also partly lined by transitional epithelium. The surrounding stromal tissue is hyperemic with dense diffuse infiltrate of LMN cells (figure5). Further immuno-histochemical examination done in this case showed epithelial cells positivity for Cytokeratin-7 (CK7), epithelial membrane antigen (EMA) and negativity for anti-S100 protein (S100), Cytokeratin-20 (CK20), smooth muscle actin (SMA) and carcinoembryonic antigen (CEA). HPE in all the cases was suggestive of median raphe cyst. Follow up period ranged from 6 to 36 months with no recurrence or any other unexpected outcome in any case.

## FIGURE 5



**HPE:** Cystic cavity lined by pseudostratified columnar, stratified squamous and transitional epithelium (HE x45)

## DISCUSSION

Median raphe cysts of the penis are very rare benign lesions that occur only in males.<sup>12</sup> The cysts are usually asymptomatic; hence, many cases are probably not reported. Numerous terms including mucus cyst of the penis, genitoperineal cyst of the median raphe, paramental cyst, hydrocystoma of the penile shaft have been proposed to describe the lesion and should be regarded as synonymous.<sup>4</sup>

The pathogenesis of this disease has not been very well understood as reflected by various theories which have been put forth for the development of median raphe cysts. The most accepted theory suggested that these cysts are thought to arise from an abnormality in the formation of the urethra whereby embryonic nests persist during invagination and closure of the urogenital folds.<sup>2,4,8</sup> Another theory holds that they may form from ectopic Littre's (periurethral) glands.<sup>9</sup> Some authors also believed that it may develop from the blockage of the paraurethral ducts.<sup>10</sup>

The cysts can develop at any site on the ventral side of the male genital area, including the paramentus, glans penis, penile shaft, scrotum, perineum or prepuce. Median raphe cysts mostly present at birth and may remain asymptomatic or unrecognized during childhood.<sup>2</sup> As the patient grows older, the cyst may also progress slowly. In some cases, the cyst may progress rapidly or become symptomatic owing to infections or trauma, which could make diagnosis difficult.<sup>5,8</sup> However, in most cases, the lesions remain asymptomatic and do not interfere with urinary or sexual function.<sup>2</sup>

In our study majority of cysts were located in the paramental site in the younger patients aged less than 15 years, whereas only 1 cyst was prepuccial in origin, in the patient aged 50 years. A possible reason for this is that the cysts on the paramental site could be easily noticed, or were more likely to induce symptoms such as voiding difficulty, prompting parents to seek treatment earlier. Although all the patients of paramental cysts in our study group were asymptomatic but underwent surgical excision in view of gradual enlargement in size of cyst, apprehension of parents about the progress of cyst and for cosmetic reasons. The more distal the location of the cysts, the greater was the manifestation of symptoms, such as difficulty voiding and performing sexual activity as seen in one patient of large prepuccial cyst in our study.

Histologically, median raphe cysts reveal a single cystic space located in the dermis with no urethral communication lined commonly by one or both of the following types of epithelium: stratified squamous epithelium or pseudostratified columnar epithelium, the latter being the most common. The majority of cysts have only one type of epithelium, while some may have areas of both. Rarely the cyst has

transitional epithelial lining in addition to stratified squamous epithelium and pseudostratified columnar epithelium constituting very rare and interesting triple histological lining.<sup>2,3,11</sup>

In our cases all the 6 paramental cysts were lined partly by stratified squamous epithelium and partly by pseudostratified columnar epithelium. While in case of prepuccial cyst histological sections showed transitional epithelial lining in addition to lined by stratified squamous and pseudostratified columnar epithelium constituting it a very rare entity of symptomatic large prepuccial cyst with triple histological lining. Immunohistochemical study in case of prepuccial cyst showed, the epithelium staining with cytokeratin 7 (Ck7), carcinoembryonic antigen (CEA) and cytokeratin 13 (CK13), while non staining for S-100, cytokeratin 20 (CK20), or human milk fat globulin 1 (HMFG1).

The clinical differential diagnosis includes steatocystoma, molluscum contagiosum, pilonidal cyst, dermoid cyst, epidermoid cyst, and urethral diverticulum. These entities can be easily distinguished on clinical and histologic grounds.<sup>2</sup>

When treatment is required in view of symptomatology or cosmetic concern, surgical excision with primary closure is the treatment of choice.<sup>4,12,13</sup> If clinical infection is present, excision should be postponed until after successful treatment of the infection. The recurrence rate following surgical excision is extremely low. In our study all patients of paramental cysts were managed by excisional biopsy with primary closure while circumcision combined with cyst excision was done in case of prepuccial swelling. Follow up period ranged from 6 to 36 months with no recurrence or any other unexpected outcome in any case.

Although expectant management may be chosen in cases of small, non enlarging and asymptomatic cysts. But if expectant management is chosen, the patient and/or family should be informed the cyst may enlarge slowly and, rarely, quickly over time, it may become infected or symptomatic and surgical excision may then be needed and done in the future.

## CONCLUSION

Median raphe cyst of the penis is an uncommon entity that usually presents as a cystic lesion on the ventral aspect of the penis, and which has a broad spectrum of histopathological appearances. They are benign and usually asymptomatic except in case where they attain very large size that interferes with urinary and sexual function. Local examination is sufficient to make a diagnosis in majority of cases, and complete surgical excision with primary is necessary to obtain good cosmetic results without recurrence.

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