## Case Report

# Pigmented median raphe cyst of the penis that developed after middle age without infection or trauma history

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**Abbreviations & Acronyms**
MRC = median raphe cyst
NA = not available

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**Introduction:** MRCs are rare benign lesions of the male genitalia that can develop anywhere along the midline from meatus to anus. They are believed to be caused by a defect in closure of median raphe during embryonic development. These cysts commonly appear in childhood or adolescence, although some are diagnosed after middle age, typically triggered by infection or trauma. Pigmented median raphe cysts, or those containing melanin pigment and/or melanocytes, are extremely rare.

**Case presentation:** A 78-year-old man visited our hospital with a complaint of a penile mass that he first noticed in his 50s which slowly grew, eventually causing voiding difficulty. He had no history of infection or trauma. The lesion was excised, and the pathological diagnosis was pigmented median raphe cyst.

**Conclusion:** We successfully treated a rare case of pigmented median raphe cyst of the penis that developed after middle age without infection or trauma history.

**Key words:** foreskin, median raphe cyst, melanin, penis, pigmented.

## Keynote message

MRCs are rare benign lesions of the male genitalia that can develop at any site along the midline from meatus to anus. These cysts generally appear in childhood or adolescence although some are diagnosed after middle age, typically triggered by infection or trauma. Furthermore, pigmented MRCs, or those containing melanin pigment and/or melanocytes, are extremely rare. We report a case of pigmented MRC of the penis that developed after middle age, without infection or trauma history. Clinicians should be aware of such a late-onset MRC without any trigger.

## Introduction

MRCs are rare benign lesions of the male genitalia that can develop at any site along the midline from meatus to anus (i.e. parameatus, glans penis, penile shaft, scrotum, perineum, or perianal).[1–19] The condition was first described by Mermet in 1895,[1] and MRC is currently believed to be caused by a defect in closure of the median raphe during embryonic development.[19] MRCs commonly appear in childhood or adolescence,[13,17,18] although some patients are diagnosed after middle age.[13] In such older age cases, cysts may progress rapidly or become symptomatic with advancing age as a result of infection or trauma.[3,12,13] By contrast, some cysts may grow rapidly for unknown reasons even in the absence of infection or trauma.[11,13] Pigmented MRCs, or those containing melanin pigment and/or melanocytes, are extremely rare, with only six cases reported in the literature.[4,5,10,14,15] We herein report a rare case of pigmented MRC of the penis, which developed after middle age without a history of infection or trauma.


**Fig. 1** Pictures of the external genitalia (a) before excision of the penile lesion and (b) 14 days after surgery. After removing the lesion together with the surrounding foreskin, the remaining foreskin was repaired in a reverse T-shaped manner.

## Case presentation

A 78-year-old man visited our hospital with the complaint of a penile lesion that caused voiding difficulty. He had a current medical history of type 2 diabetes mellitus (hemoglobin $A_{1c}$ = 7.9%), essential hypertension, hyperuricemia, benign prostatic hyperplasia, and asymptomatic abdominal aortic aneurysm with routine follow-up. He also had a history of orthopedic surgery for a shoulder joint. At his first visit, a painless, bilocular, skin-colored, elastic-soft, and movable mass with a diameter of approximately 6.0 cm was located at the ventral side of the penile shaft (Fig. 1a). He first noticed the lesion in his 50s, at which time he had no history of infection or trauma; it then slowly grew in size and eventually caused difficulty with urination (note: although the mass did not infiltrate the urethra directly, existence of the "dangling" mass interfered with standing urination). Under the diagnosis of penile cyst with ultrasonography, he underwent ablative surgery following improvement in glycemic control. The mass was excised together with the surrounding foreskin (Fig. 2a), and the remaining foreskin was repaired in a reverse T-shaped manner (Fig. 1b). Histological examination revealed the lesion to be a bilocular cyst (Fig. 2b) lined mostly by non-keratinizing stratified squamous epithelium and partially by stratified columnar epithelium (Fig. 2c). The cyst was filled with light tan liquid substance. There were several melanocytes at the basal layer of squamous epithelium (Fig. 2d). Accordingly, the definitive diagnosis was pigmented MRC of the penis. The patient's voiding difficulty

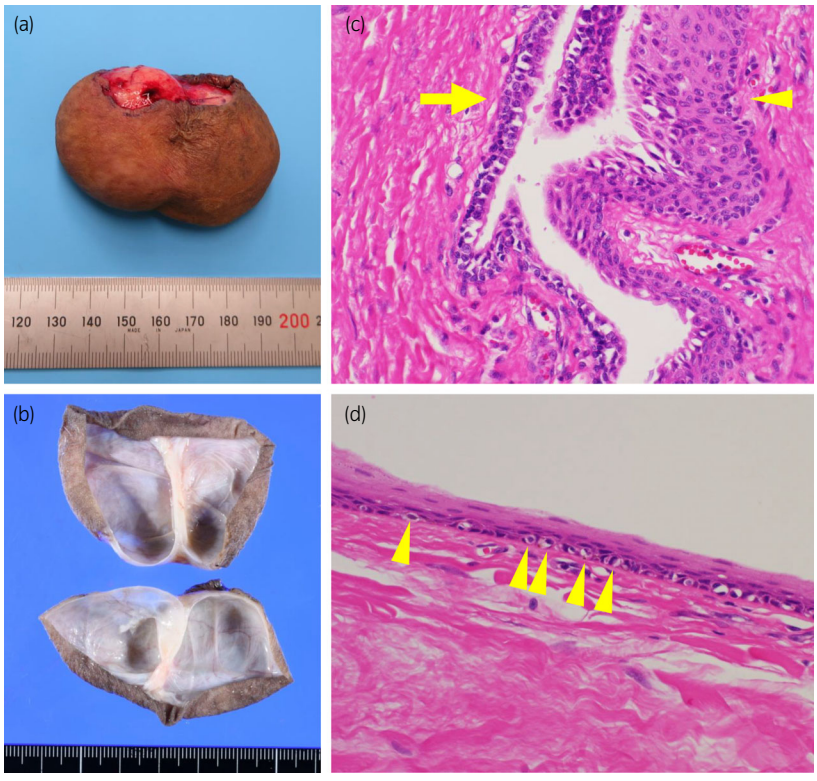


**Fig. 2** (a) Macroscopic image of the fresh specimen, which was excised together with the surrounding foreskin. (b) Macroscopic image of the fixed and cut specimen, showing the lesion to be a bilocular cyst. (c) Microscopic image of the lesion (hematoxylin and eosin stain) showing the cyst to be lined mostly by non-keratinizing stratified squamous epithelium (arrowhead) and partially by stratified columnar epithelium (arrow). (d) There were several melanocytes (arrowheads) at the basal layer of squamous epithelium.

**Table 1** Clinicopathological features of seven cases of pigmented MRC

| Case | First author | Age, years | Site of lesion | Size, mm | Macroscopic color | Epithelial lining of MRC |
|---|---|---|---|---|---|---|
| 1 | Fetissof[4] | NA | Penile shaft | NA | NA | Squamous epithelium |
| 2 | Hitti[5] | 29 | Penile shaft | 60 | NA | Pseudostratified columnar epithelium |
| 3 | Urahashi[10] | 4 | Perineum | 2–4 | Brown–black | Pseudostratified columnar epithelium |
| 4 | Urahashi[10] | 5 | Glans penis | 1, 5 | Brown–black | Pseudostratified columnar epithelium |
| 5 | Nishida[14] | 6 | Penile shaft | 8 | Brown | Cuboidal epithelium |
| 6 | Ishida[15] | 48 | Penile shaft | 10 | Skin-colored | Cuboidal epithelium |
| 7 | Present case | 78 | Penile shaft | 60 | Skin-colored | Squamous epithelium and stratified columnar epithelium |

was cured by surgery. There were no severe complications or evidence of recurrence after surgery.

## Discussion

MRCs are rare benign lesions of the male genitalia that are believed to be caused by a defect in closure of the median raphe during embryonic development of the male urethra.[19] The male urethra develops by several steps including fusion of the urethral folds and envelopment of the urethral groove. During the process, "tissue trapping" can occur due to incomplete fusion of the urethral folds and/or anomalous epithelium outgrowth following primary closure.[12,13] Trapped tissues outside the urethral groove can result in formation of cystic lesions along the median raphe (i.e. MRCs). Epithelial linings of MRCs are histopathologically classified into four types (urethral, epidermoid, glandular, and mixed), which may reflect the cell type in the trapped tissue.[13] The "urethral type," the most common form, is composed of a pseudostratified columnar (or urothelium-like) epithelium. The "epidermoid type" comprises a squamous epithelium, whereas the "glandular type" consists of a urethral epithelium with interspersed glandular structure. The "mixed type," the second most common form, comprises more than one type of epithelium, such as urethral epithelium with squamous metaplasia, or mucinous cells, or all three coexisting.[13] Although extremely rare, MRCs containing melanin pigment and/or melanocytes (i.e. pigmented MRCs) have been reported, with six cases to date having been published.[4,5,10,14,15] Given that the cyst of our patient was lined mostly by non-keratinizing stratified squamous epithelium and partially by stratified columnar epithelium (Fig. 2c), it was considered the "epidermoid type"-based "mixed type." Furthermore, given that several melanocytes were detected at the basal layer of squamous epithelium (Fig. 2d), we considered this the seventh case of "pigmented MRC." For reference, we have summarized the clinicopathological features of these seven reported cases (Table 1).

MRCs can theoretically develop anywhere along the midline from meatus to anus, including parameatus,[9] glans penis,[10,19] penile shaft,[3–5,11,14,15] scrotum,[12] perineum,[7,10] and perianal,[6,16] but they present most commonly in the penile shaft.[13] MRCs commonly appear in childhood or adolescence, with a bimodal distribution at approximately 1–10 and 21–40 years old.[13,17,18] MRCs; generally remain asymptomatic without interference with urinary or sexual function; therefore, affected patients are either presented to a physician by their parents during childhood or they visit a physician themselves later in their 20s and 30s due to development of symptoms or cosmetic reasons.[13] Some MRCs may progress rapidly or become symptomatic with advancing age as a result of infection or trauma.[3,12,13] Treatment of MRCs consists of surgical removal, although simple aspiration alone is not recommended because of a high risk of recurrence.[13] The present case was atypical in two respects: (i) the lesion was melanin pigmented (i.e. "pigmented MRC") as described above; and (ii) the lesion was first noticed while the patient was in his 50s, without any infection or trauma history, and grew to eventually cause voiding difficulty. Although it is unclear why the MRC of this patient became evident after middle age, clinicians should be aware of such a late-onset type of MRC without any trigger.

In summary, we successfully treated a rare case of pigmented MRC of the penis that developed after middle age without any trigger such as infection or trauma.

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## Conflict of interest

The authors declare no conflict of interest.

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