

Case Reports

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MEDIAN RAPHE CYST. REPORT OF TWO CASES AND LITERATURE REVIEW

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Summary.- OBJECTIVES: To report 2 cases of median raphe cysts, 1 in the penis and the other in the perineum

METHOD: Two cases of median raphe cyst are described; the first was treated by surgery and the second required no treatment.

RESULTS: The surgical patient experienced no complications and was asymptomatic with no recurrence at 2 years.

CONCLUSIONS: Median raphe cysts are a rare, benign condition of uncertain etiology. Treatment consists on simple excision.



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Resumen.- *OBJETIVOS:* Presentar dos casos de quistes de rafe medio, uno de localización peneana y otro perineal.

MÉTODO: Se diagnostican y describen dos casos de quiste de rafe medio, a uno se aplicó tratamiento quirúrgico y el segundo no precisó de tratamiento alguno.

RESULTADOS: La intervención quirúrgica del paciente operado no tuvo complicaciones y a los dos años de la misma estaba asintomático y sin recidiva.

CONCLUSIÓN: Los quistes de rafe medio son poco frecuentes; es una patología benigna y su etiología no está completamente aclarada. Su tratamiento consiste en la exéresis simple.

Palabras clave: *Quistes de rafe medio. Restos embrionarios.*

INTRODUCTION

Median raphe cyst was first described in detail by Mermet in 1895 (1,2). Such lesions are uncommon and can occur in the midline, following the raphe, from the distal tip of the penis to the margins of the anus, although the region usually affected is the distal area near the glans (3).

Case 1

A 32-year-old patient reported perineal bultoma that had been present from childhood as long as he could remember (Figure 1). The examination revealed a mass at the midline of the perineum; the mass was soft, elastic, and nonpainful, and did not appear to be adhered to deep surfaces. The voiding and retrograde cystourethrography was normal. The magnetic resonance imaging study of the pelvis and perineum showed the morphology, fluid content, and absence of any communication between the mass and other structures or organs (Figure 2a, 2b, 2c); the size was 5x2x1.5 cm. The cyst was excised under spinal block; perioperative examination of the surgical specimen revealed that the cyst was multilocular (Figures 3 and 4). The histologic study showed a multilocular cystic cavity, with pale brown mucus-fluid content, partially lined by pseudostriated respiratory epithelium with mucosecretory cells positive for periodic acid-Schiff staining and ciliated cells, as well as nonkeratinizing squamous epithelium. Cytokeratin (CK) immunohistochemistry revealed co-

lumbar cells and apical cells of squamous epithelium with immunophenotype positive for CK 7, epithelial membrane antigen, and carcinoembryonic antigen, and negative for CK 20.

At the time of writing, 2 years after surgery, the patient is asymptomatic with no recurrences.

Case 2

A 68-year-old patient was referred by his primary-care physician for moderate prostatism and a prostate-specific antigen level of 19. The examination revealed a median raphe cyst of the penis (Figure 7); the patient reported that the cyst had been present since birth and he had never experience any discomfort, even during sexual relations. A digital rectal examination showed a small, fibrous prostate. The ultrasound-guided transrectal prostate biopsy and histologic study revealed adenocarcinoma; the patient was treated with radiotherapy and hormonal therapy. The cyst of the penis was not treated.

DISCUSSION

The lesions are congenital and readily diagnosed clinically, but usually diagnosed in adulthood rather than childhood because they are often not evident for years and typically silent, particularly when small, and only grow and cause symptoms in the case of traumatic injury or infection (3,4). Other terms apart from median raphe cyst have been used: mucus cyst of the penis, genitoperineal cyst of the medium raphe, paramental cyst, hydrocystadenoma, apocrine cystadenoma, and urethroid cyst (1,2,5,6), although these cysts are now



FIGURE 1. Perineal median raphe cyst.

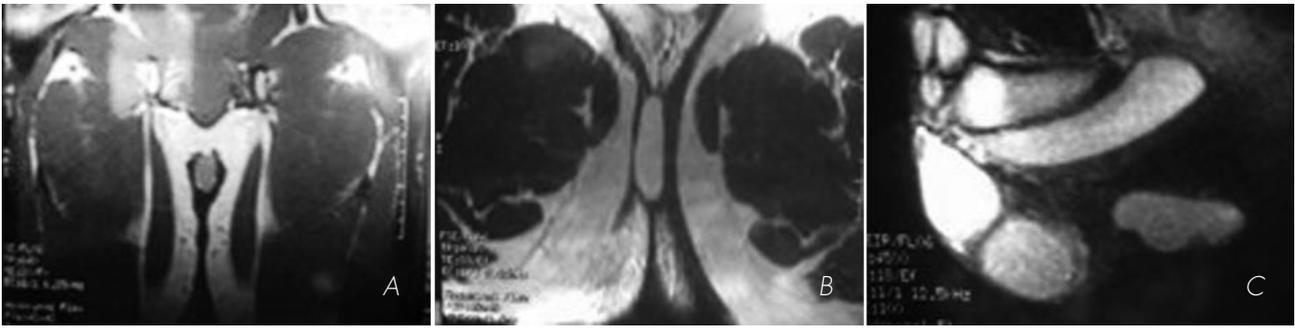


FIGURE 2A, 2B y 2C. Magnetic resonance imaging of the lesion in the pelvic and perineal region.

known referred to as median raphe cysts because they have similar clinical and histologic characteristics.

Only a few cases have been published, mostly in dermatology journals rather than urology publications. The cysts are usually asymptomatic; hence, many cases are probably not reported.

Cyst formation is explained by embryologic anomalies or closure defects of the raphe, sometime due to the persistence of embryonic rests during invagination and fusion of the urogenital folds, the presence of embryologic epithelium protruberances trapped before fusion of the folds, or abnormal development of the Littre's (ectopic periurethral) glands, with intraepithelial mucous glands observed in the latter cases (5,7,8,9).

Clinical symptoms are usually rare, although cystic infection or ulceration sometimes occurs (9). Discomfort (e.g., during sexual relations) may occur if the cyst is large and urinary symptoms may be present if the cyst

is near the urethral meatus (9). In some cases, the patient consults for cosmetic reasons or to obtain a diagnosis or recommendation, even though no symptoms are present.

The differential diagnosis should be performed with epidermal cysts, steatocystoma, glomus tumors, dermoid cyst, urethral diverticulum, and pilonidal cyst (3,6,7,9). A histologic study will also clarify the diagnosis if the lesions are small and resemble molluscum contagiosum or condylom (8).

The lesion may produce a wide spectrum of results in the histopathology study, including stratified, pseudostratified, and squamous columnar epithelium, resembling the epithelium of the male urethra (8,10). Three histologic patterns have been described: 1) urethral type, present in 70% of cases with epithelium similar to the urethra; 2) epidermoid type, composed of squamous epithelium (which accounts for 10% of cases); and 3) mixed type formed by a combination of the above (5).

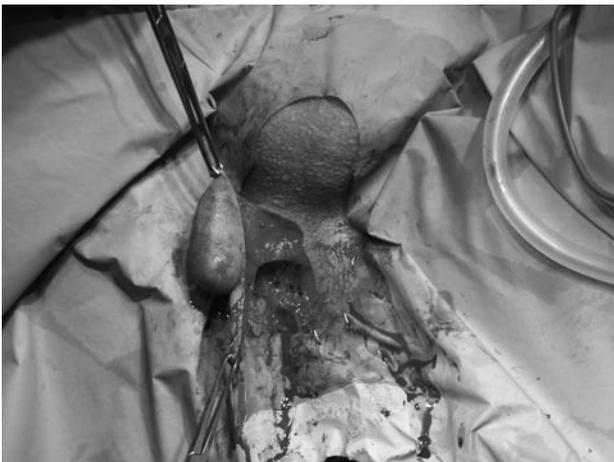


FIGURE 3. Surgical excision of the cyst.



FIGURE 4. Surgical specimen.

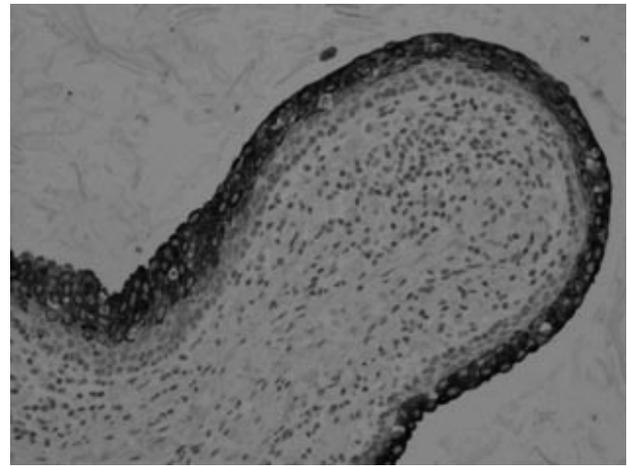
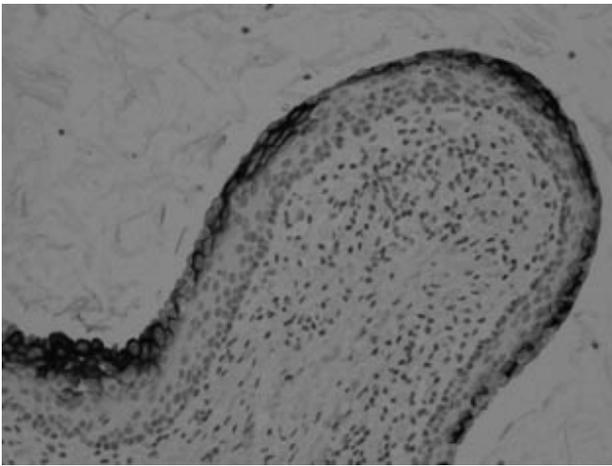


FIGURE 5. Positive immunohistochemistry reactions of apical cells for cytokeratin 7 and carcinoembryonic antigen.

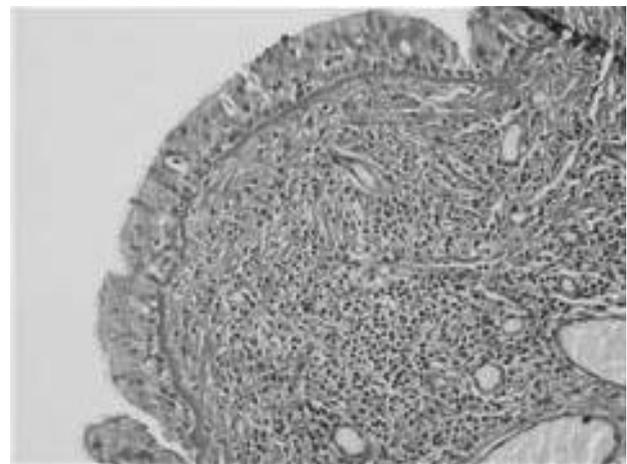
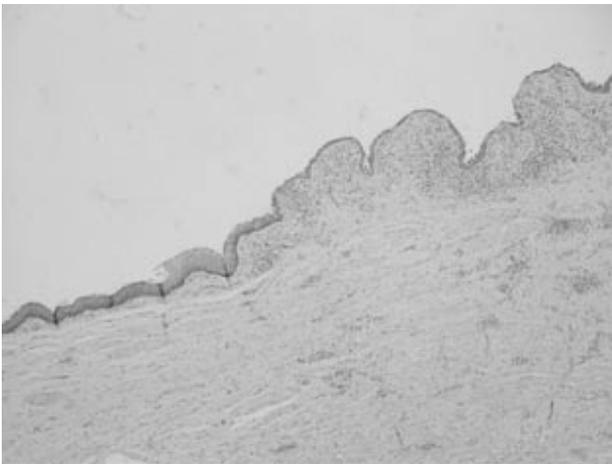


FIGURE 6. Cyst partially lined by respiratory-like pseudostratified epithelium with mucus secretory cells (positive periodic acid-Schiff staining) and nonkeratinizing squamous epithelium.



FIGURE 7. Median raphe cyst of the penis.

The histologic study usually reveals the presence of an irregular cystic cavity, covered with an epithelium that corresponds to that of the urethra portion from which it originates, i.e., nonkeratinizing squamous stratified epithelium in the distal portion (ectodermic origin) and columnar pseudostratified epithelium when it originates from the rest of the urethra (endodermic origin) (3,6).

Median raphe cysts are positive for periodic acid-Schiff stain, colloidal iron, and Alcian blue (4,7). Immunohistochemical studies based on immunostaining with cytokeratins, particularly CK 7, will confirm the urothelial origin of these cysts. The carcinoembryonic antigen immunostaining observed particularly in columnar cells probably indicates the dysembryogenic cloacal nature in cyst genesis (2,10). The lack of immunoreactivity for CK 20 indicates the benign, nondysplastic nature of these lesions (2,6).

Treatment consists of surgical excision in the case of symptoms or for cosmetic reasons. If the lesions are small and asymptomatic, the best option is to take a wait-and-see approach.

REFERENCES AND RECOMENDED READINGS

(*of special interest, **of outstanding interest)

- **1. Memert P. Congenital cysts of the genitoperineal raphe. *Rev Chir* 1895; 15:382-435.
- **2. Dini M, Baroni G, Colafranceschi M. Median raphe cyst of the penis: a report of two cases with immunohistochemical investigation. *Am J Dermatopathol* 2001; 23(4):320-324.
- *3. Nagore E, Sanchez-Motilla JM, Febrer MI, Aliaga A. Median raphe cyst of the penis: a report of five cases. *Pediatr Dermatol* 1988; 15(3): 191-193.
4. Lopez-Candel E, Roig Alvaro J, Lopez-Candel J, Fernandez Dozagarat S, Soler J, Hernandez Bermejo JP, et al. Quistes de rafe medio perineal en la infancia. *An Esp Pediatr* 2000; 52(4): 395-397.
5. Otsuka T, Ueda Y, Terauchi M, Kinoshita Y. Median raphe (parameatal) cysts of the penis. *J Urol* 1998;159(06):1918-1920.
- *6. Cardoso R, Freitas JD, Reis JP, Tellechea O. Median raphe cyst of the penis. *Dermatol Online J* 2005; 11(3): 37.
- *7. Cole LA, Helwig EB. Muroid cysts of the penile skin. *J Urol* 1976; 115(04): 397-400.
8. Romani J, Barnadas MA, Miralles J, Curell R, De moragas JM. Median raphe cyst of the penis wiht ciliated cells. *J Cutan Pathol* 1995; 22(4): 378-381.
9. Menendez Lopez V, Funez R, Osman Agha G, Lopez Almansa M, Garcia E. Quistes de rafe medio. *Actas Urol Esp* 1997; 21(8): 800-802.
10. Sagar J, Sagar B, Patel AF, Shak DK. Ciliated median raphe cyst of perineum presenting as perianal polyp: a case report with immunohistochemical study, review of literature, and pathogenesis. *Scientific World Journal* 2006; 6:2339-2344.