ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
Direttore Nicola Picardi

Pigmented median raphe cyst of the scrotum.

A rare pediatric case and review of the literature

Ann Ital Chir, 2022; 11 - March 14
pii: S2239253X22037422
Online Epub

Cristina Martucci*, Francesca Diomedi Camassei**, Alessandro Borsellino***

**General Surgery Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy*
***Pathology Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy*
****Andrological Surgery Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy*

Pigmented median raphe cyst of the scrotum. a rare pediatric case and review of the literature

AIM: *Median raphe cyst are uncommon malformations of male genitalia, in which are rarely described melanin pigments or melanocytes; less than ten cases have been reported in literature. The aim of our study is to describe a rare ormations, case of pigmented median raphe cyst of the scrotum, successfully treated in our hospital.*
CASE EXPERIENCE: *A 6-years-old boy underwent surgical removal of a melanocytic lesion of the ventral surface of the scrotum in Day Surgery regimen. He reported no surgical complication or recurrence.*
RESULTS: *Histology showed multiple cystic nodules, lined by squamous and pluri-stratified columnar epithelium, some of which contained melanic deposits and were anti-MART-positive.*
DISCUSSION: *Even though the first case has been reported in 1985, the etiology of median raphe cysts remains unclear. Infrequently associated with trauma or infections, these lesions seem to origin from an abnormal development of the periurethral glands or atypical closure of the median raphe. Rarely melanin pigments or melanocytes are described in the histological examination, and the cause of the pigmentation is still unknown.*
CONCLUSION: *Median raphe cysts present a non-negligible variety of clinical presentations and histological features. Pigmented ones represent the rarest form: further studies may be necessary to clarify their pathogenesis and describe their clinical evolution.*

KEY WORDS: Median raphs, Male genitalia, Malformations

Introduction

Median raphe cysts are rare lesions of the male genitals, which can arise along the midline from the perineum to the penile meatus. They are mostly recognized during childhood or adolescence and totally asymptomatic. Although the pathogenesis of the disease is still not well understood, many authors suggest their origin from the urethral mucosa or gland (Littre's gland) [1].

The presence of melanin or melanocytes in these lesions is very uncommon and only few cases have been described [2]. We reported a rare pediatric case of a pigmented median raphe cyst of the scrotum.

Case Experience

A 6-year-old boy presented at our Unit with a brownish multi-cystic lesion of the ventral surface of the scrotum and base of the penis. The parents noticed it fifteen days before the visit; no trauma or other significant pathologies were referred.
On physical examination, the lesion was measuring 3x0.5 cm, slightly elevated, soft and movable, not painful. The nodule was resected as a Day-Surgery admission; no local recurrence or surgical complication was encountered at 5-years follow-up.

Pervenuto in redazione Novembre 2021. Accettato per la pubblicazione Gennaio 2022
Correspondence to: Dr. Cristina Martucci, Andrological Surgery Unit, Bambino Gesù Children's Hospital, Piazza Sant'Onofrio 4, 00165 Rome (RM), Italy (e-mail: Martucci.cristina@gmail.com)

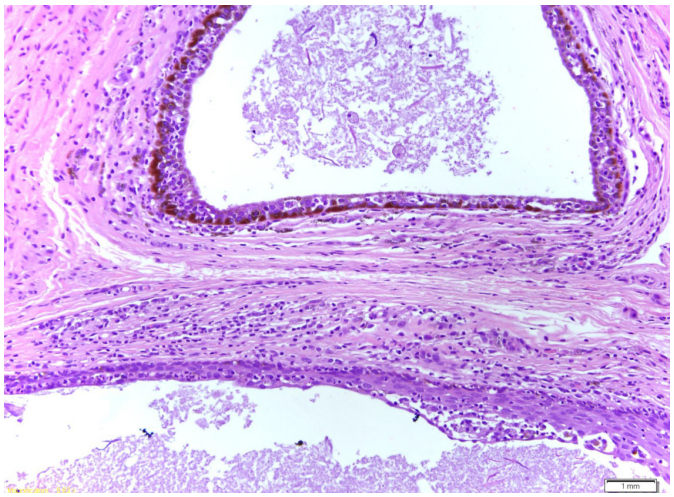

Fig. 1: Cystic wall lined by both squamous and pluristratified columnar epithelium. Some epithelial cells contained melanic deposits.

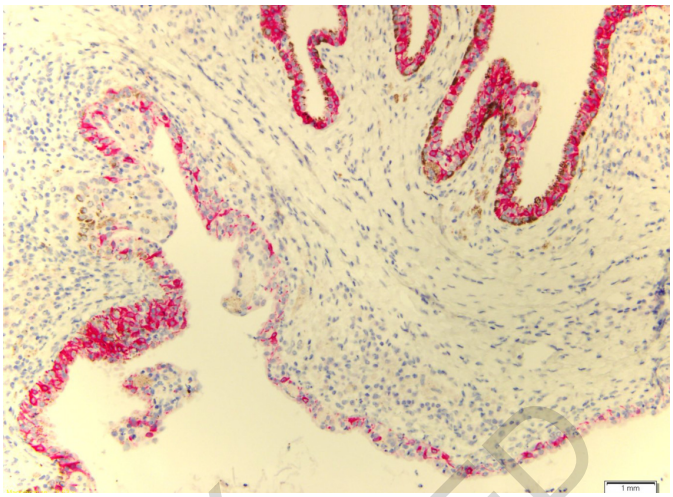

Fig. 2: Immunostain for MART-A showed clusters of positive cells, indicating focal melanocytic phenotype of epithelial lining.

Histological examination showed multiple cystic nodules (maximum diameter: 7 mm), lined by squamous and pluri-stratified columnar epithelium (Fig. 1). Some of the lining cells contained melanic deposits and were anti-MART-positive (Fig. 2).

Discussion

Median raphe cysts are benign uncommon entities that arise, during childhood or adolescence [2,3] and they can occur at any site on the ventral side of the genital area, including the parameatus, glans penis, penile shaft, scrotum or perineum.

They are usually solitary, asymptomatic and not interfering with sexual or urinary function [4]. They are frequently unrecognized during childhood but may progress later and become symptomatic during adolescence or adulthood [4], appearing as a solitary and movable cystic nodule on the ventral surface of the penis.

This could explain the bimodal distribution of the presentation: the patients were either presented to the physicians by their parents during childhood or they visited the physician themselves as young adults because they developed symptoms or, more frequently, for cosmetic reasons.

Several terminologies have been used, in the past literature, to describe it, including median raphe cyst, 'mucoid cyst of penile skin' and 'genitoperineal cyst of the medium raphe'.

Although referred to as median raphe "cysts," they can also present, in rare cases, as a cordlike or canaliform induration on the median raphe.

The first case was reported in 1895 from Mermet but only a few have been reported to date, mostly in case reports. The clinic pathogenesis of the disease are not well understood by physicians and many theories have been proposed to explain their aetiology: an abnormal development of the periurethral glands of Littre, anomalies in the process of closure of the median raphe or during the separation of the urethral columnar epithelium from the urethra.

The cysts should be differentiated from other genital lesions like the glomus tumor, pilonidal cyst, dermoid cyst, epidermal inclusion cyst, urethral diverticulum and steatocystoma. These lesions can rarely be associated with acquired factors (recent surgical procedures, infections and trauma) [5,6].

Shao et al [7], suggested a classification of the median raphe cysts into four categories: urethral, epidermoid, glandular and mixed. The urethral type (55% of the cases) is lined by urothelium-like epithelium, with multiple layers of small cells over covering a layer of columnar cells.

The epidermoid type (5% of the cases) is characterized by squamous epithelium, while glandular type (3%) presents well-differentiated glandular structure, surrounded by urethral epithelium. The mixed type (37%) contains more than one type of epithelial lining. According to Nagore et al [4], the histopathological aspect of the median raphe cysts may reflect their embryonic origin. Melanin pigments or melanocytes in median raphe cyst represent a rare finding, with only less than 10 cases reported in literature [3,8-13]. The cause of the pigmentation of these lesion is still unclear [14], but it may be determined by the presence of lipochrome, the Tyndall phenomenon or the presence of melanocytes [8]. Our case presented multiple MART-positive cells containing numerous melanic granules. Neither ciliated cells nor transitional epithelium were present in the specimen, as described by other authors [15], so it belonged to the rare epidermoid type.

No malignant potentiality has ever been described, during long-term follow-up, in the literature [7].

Surgical excision followed by primary closure, which establishes an accurate hemostasis and prevents infection or cosmetic sequelae represents the optimal treatment [7].

Conclusion

Median raphe cysts show a considerable variety of clinical presentations and histological features. Further studies are necessary to clarify their embryological origin and the pathogenesis of the pigmented ones.

Riassunto

SCOPO DELLO STUDIO: Le cisti del rafe mediano rappresentano una rara malformazione dei genitali maschili; in pochissimi casi (meno di 10 riportati in letteratura) è presente nella lesione una componente melanocitica o pigmenti di melanina. Lo scopo del nostro studio è quello di descrivere un raro caso di cisti pigmentata del rafe mediano dello scroto, trattata con successo presso il nostro nosocomio.

CASO CLINICO: Il paziente, di 6 anni di età, presentava una lesione cistica e melanocitica della superficie ventrale dello scroto; è stato sottoposto ad asportazione della cisti in regime di Day Surgery e non ha mostrato, con un follow-up di 5 anni, alcuna recidiva né complicanza chirurgica.

RISULTATI: L'analisi istologica della lesione ha mostrato multipli noduli cistici, caratterizzati da un epitelio squamoso pluristratificato, che conteneva depositi di melanina ed esprimeva positività per anti-MART.

DISCUSSIONE: Nonostante il primo caso sia stato descritto nel 1985, l'eziologia delle cisti del rafe mediano risulta ancora ignota. Raramente associati ad infezioni o trauma, tali lesioni sembrerebbero originale da un anomalo sviluppo delle ghiandole periuretrali o da una chiusura patologica del rafe mediano.

Solo in pochi casi sono presenti melanociti o pigmenti di melanina all'esame istologico e la causa di tale pigmentazione non è ancora chiara.

CONCLUSIONI: Le cisti del rafe mediano mostrano un'ampia gamma di presentazioni cliniche e di aspetti istologici. Le forme pigmentate rappresentano una minima percentuale: ulteriori studi sono necessari per chiarirne la patogenesi e descriverne la naturale evoluzione clinica.

References

1. Cole LA, Helwig EB: *Mucoid cysts of the penile skin*. J Urol, 1976; 01:115(4):397-400.

2. Mermet P: *Congenital cysts of genitoperineal raphe*. Rev Chir, 1895; 15:382.

3. Hitti IF, Vuletin JC, Rapuano J: *Giant median raphe cyst of the penis with diffuse melanosis of its epithelial lining*. Urol Int, 1989; 44(2):121-124.

4. Nagore E, Sanchez-Motilla JM, Febrer MI, Aliaga A: *Median raphe cysts of the penis: A report of five cases.* Pediatr Dermatol, 1998; 01:15(3):191-93.

5. Ramon Quiles D, Betlloch Mas I, Jimenez Martinez A, Verdeguer Miralles JM, Botella Anton R, Castells Rodellas A: *Gonococcal infection of the penile median raphe.* Int J Dermatol, 1987; 01:26(4):242-43.

6. Sharkey MJ, Grabski WJ, McCollough ML, Berger TG: *Postcoital appearance of a median raphe cyst*. J Am Acad Dermatol, 1992; 01:26(2 Pt 1):273-74.

7. Shao IH, Chen TD, Shao HT, Chen HW: *Male median raphe cysts: Serial retrospective analysis and histopathological classification*. Diagn Pathol, 2012; 4:7:121-1596.

8. Urahashi J, Hara H, Yamaguchi Z, Morishima: *Pigmented median raphe cysts of the penis*. Acta Derm Venereol, 2000; 01:80(4):297-98.

9. Colmenero I, Sastre N, de Prada I, Hernandez-Martin A: *Pigmented cyst of the median raphe of the scrotum in a boy*. Actas Dermosifiliogr, 2009; 01:100(7):633-34.

10. Romani J, Barnadas MA, Miralles J, Curell R, de Moragas JM: *Median raphe cyst of the penis with ciliated cells*. J Cutan Pathol, 1995; 01:22(4):378-81.

11. Koga K, Yoshida Y, Koga M, Takeshita M, Nakayama J: *Median raphe cyst with ciliated cells of the penis*. Acta Derm Venereol, 2007; 87(6):542-43.

12. Nishida H, Kashima K, Daa T, Yada N, Arakane M, Matsuda K, et al: *Pigmented median raphe cyst of the penis*. J Cutan Pathol, 2012; 01:39(8):808-10.

13. Ishida M, Iwai M, Yoshida K, Kagotani A, Okabe H: *Pigmented median raphe cyst of the penis with consideration of the possible mechanism of melanocytic colonization: A case report*. Oncol Lett, 2014; 01:7(2):342-44.

14. Malhotra R, Bhawan J: *The nature of pigment in pigmented apocrine hidrocystoma*. J Cutan Pathol, 1985; 01:12(2):106-109.

15. Fetissof F, Lorette G, Dubois MP, Philippe A, Tharanne MJ, Jobard P: *Endocrine cells in median raphe cysts of the penis*. Pathol Res Pract, 1985; 180(6):644-46.