

Pigmented Cyst of the Median Raphe of the Scrotum in a Boy

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To the Editor:

Cysts of the median raphe (CMR) are uncommon lesions that result from abnormal embryologic development. In most cases, a urothelial-type or squamous epithelial lining is present, but other rarer histologic variants have been described. One of these, reported infrequently in the literature, is characterized by the presence of melanocytes and melanic pigment in the epithelial lining.

A 3-year-old boy was referred to our hospital for surgical removal of a cystic tumor that had been present in the median raphe of the scrotum for 2 years. The lesion was initially a single cyst, but further cysts subsequently appeared along the median raphe of the scrotum. These grew and became progressively pigmented. All cysts were less than 1.5 cm in diameter, soft, painless, and with no signs of inflammation. The lesion was completely excised. On sectioning the surgical specimen, there was a longitudinal canal measuring 0.2 cm in diameter, running the full length of the tissue sample, with several cystic dilatations along its path, the biggest measuring 1.5 cm. The canal and the cysts were filled with a yellow pasty material. Histologically, both the canal and the cystic dilatations showed an epithelial lining in which areas of pseudostratified cylindrical epithelium (Figure 1) alternated with keratinized stratified squamous epithelium (Figure 2). Many areas containing intracytoplasmic melanin were observed in the epithelium, both at the basal level and in upper strata (Figure 3). In addition, some vacuolated melanocytes could be observed in

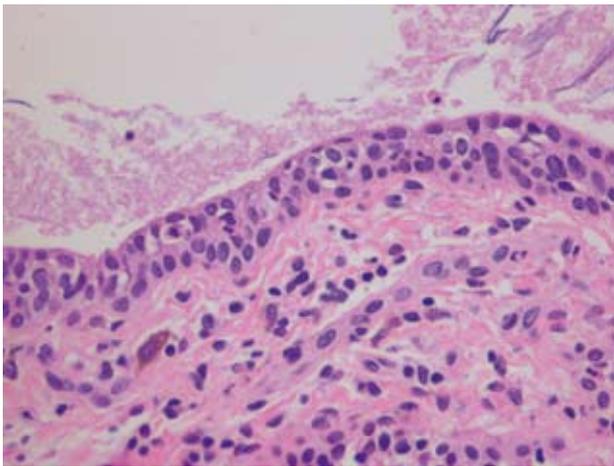


Figure 1. Areas lined with urothelial-type pseudostratified columnar epithelium (hematoxylin-eosin, $\times 200$).

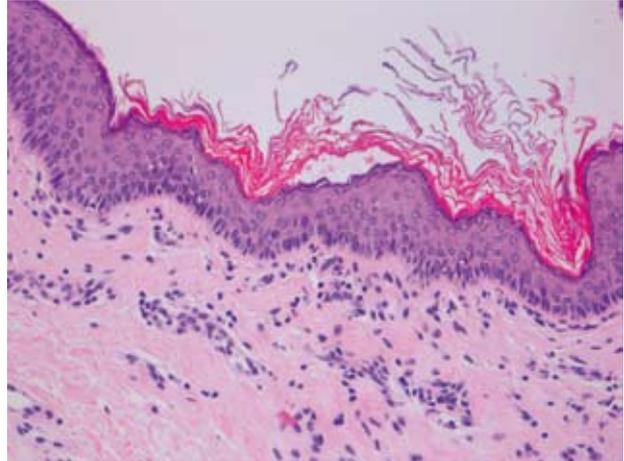


Figure 2. Areas lined with flat keratinized stratified squamous epithelium (hematoxylin-eosin, $\times 200$).

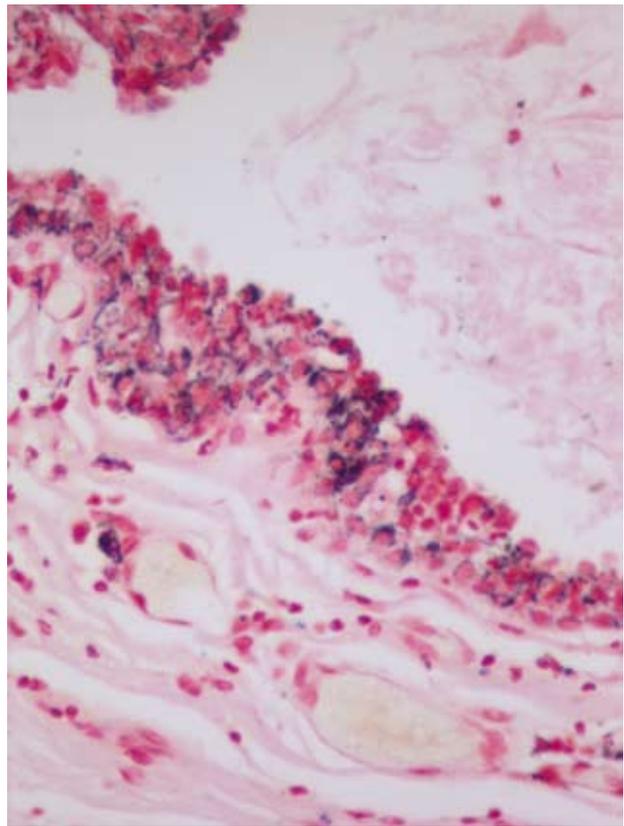


Figure 3. Melanic pigment in the cytoplasm of the epithelial cells and in the subepithelial melanophages (Masson-Fontana, $\times 200$).

the basal layer. In some areas, the presence of isolated subepithelial melanophages was detected along with a moderately-intense mixed inflammatory infiltrate. No decapitation secretion, mucosal secretion, or ciliated cells were observed and there were no myoepithelial cells in the cyst wall. Likewise, no atypia or mitosis was observed. The histopathological diagnosis was a pigmented variant of mixed-type CMR.

CMR is a rare lesion that may develop at any site between the anus and the urinary meatus.¹ The region of highest incidence is the ventral aspect of the penis, often near to the glans. CRM is generally diagnosed in patients under 30 years of age.¹ Three clinical forms have been described. The most common is a solitary cyst, but multiple cysts or a canal along the median raphe have also been reported.² Our case has the particular characteristic that the canal had multiple cystic dilatations along its path.

Three theories have been put forward to explain the pathogenesis of CRM. The first proposes that they occur during embryonic development, after primary closure of the urethral and genital folds, as a result of evagination of the urethral epithelium followed by subsequent growth; the second postulates that the lesion arises from epithelial debris originating from incomplete closure of the urethral folds^{3,4}; and the third considers that these lesions could be due to the presence of dilated ectopic periurethral (Littre) glands.³ We agree with Nagore et al⁵ in that these 3 mechanisms may be complementary and are not necessarily exclusive.⁵

Histologically, 3 patterns can be described:

1. Urethral type, lined by a pseudostratified columnar epithelium (70%)
2. Epidermoid type, with a stratified squamous epithelium (10%)
3. Mixed type (4.6%), as is the case in our patient, in which both types of epithelium are present.⁶

In addition to the above, some uncommon histological variants of CRM have been reported.^{7,8} The pigmented variant, as in the case we present here, is one of these, and to our knowledge, only 3 other cases have been published previously.^{9,10} Histologically, it shows a pseudostratified columnar, squamous, or mixed lining, with melanin granules in the cytoplasm of the basal cells and occasionally in the upper layers. Dendritic melanocytes are also observed interspersed among the epithelial cells, and subepithelial melanophages.^{9,10}

Although it was believed for a long time that, in superior warm-blooded vertebrates, melanocytes migrating from the neural crest were limited to the epidermis and

specialized organs, such as the eye or the pia mater, they have been shown to be present in other sites such as the esophagus, larynx, prostate, vagina, uterine cervix, and urothelial epithelium. The migration of undifferentiated melanoblasts from the neural crest to the urothelium explains the etiology of this variant of CRM.^{9,10}

The treatment for CRM recommended by most authors is simple excision followed by primary closure in order to prevent infections or symptoms associated with the site.⁴ In our patient, this procedure proved successful. Until present, no cases of malignant transformation of CRM have been reported.

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Conflicts of Interest

The authors declare no conflicts of interest.

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