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

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

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

Figure 3. Melanic pigment in the cytoplasm of the epithelial cells and in the subepithelial melanophages (Masson-Fontana, ×200).
folds
debris originating from incomplete closure of the urethral
the second postulates that the lesion arises from epithelial
the urethral and genital folds, as a result of evagination of
during embryonic development, after primary closure of
pathogenesis of CRM. The first proposes that they occur
been reported.

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 postulates 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; and the third considers that these lesions could be
due to the presence of dilated ectopic periurethral
(Littre) glands. We agree with Nagore et al in that
these 3 mechanisms may be complementary and are not
necessarily exclusive.

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.

In addition to the above, some uncommon histological
variants of CRM have been reported. 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. 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.

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.

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.
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.

References