



Pigmented Median Raphe Cysts on the Scrotum and Perianal Region: A Case Report

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Dear Editor:

Median raphe cysts (MRCs) are uncommon lesions occurring on the ventral median raphe, from the glans penis to the anus. These cysts represent a defect in the embryologic development of the male genitalia¹. The pathogenesis of these cysts has not been fully understood. Three different mechanisms have been described: fusion defect of the urethral folds, development of the ectopic periurethral glands of Littre, and development from the urethral columnar epithelium followed by separation². Pigmented MRCs (PMRCs), which contain melanin pigments and/or melanocytes, have rarely been reported.

The present case patient was a 4-year-old boy who presented with brown and black linear plaques on the scrotum and perianal region that had been present since birth (Fig. 1A~C). We received the consent form about publishing all photographic materials from parents of patient. Histopathological findings showed a cystic cavity lined by cuboidal epithelium and stratified squamous epithelium containing melanin pigments (Fig. 2A~D). On immunohistochemical analysis, melanocytes and melanin pigments in the epithelial layers were visualized with c-kit, S-100, Fontana-Masson, and Melan-A stains (Fig. 2E~H).

Although the cause of PMRC is not fully understood, it

may be due to the presence of lipochrome, the Tyndall phenomenon, or the presence of melanocytes that aberrantly migrated during the embryonic period³. It might also be related to factors stimulating melanocyte proliferation and differentiation, such as stem cell factors or c-kit⁴. c-kit plays a critical role in melanocyte physiology, influencing the melanogenesis, proliferation, migration, and survival of pigment-producing cells. The expression of c-kit could indicate melanocyte during their migration phase; thus, it may be considered a result of the mitogenic effect of melanocyte proliferation⁵.

Following grading, the immunohistochemical examination revealed high-intensity c-kit staining in the melanocytes of epithelial cells (Fig. 2E), strongly suggesting that c-kit con-

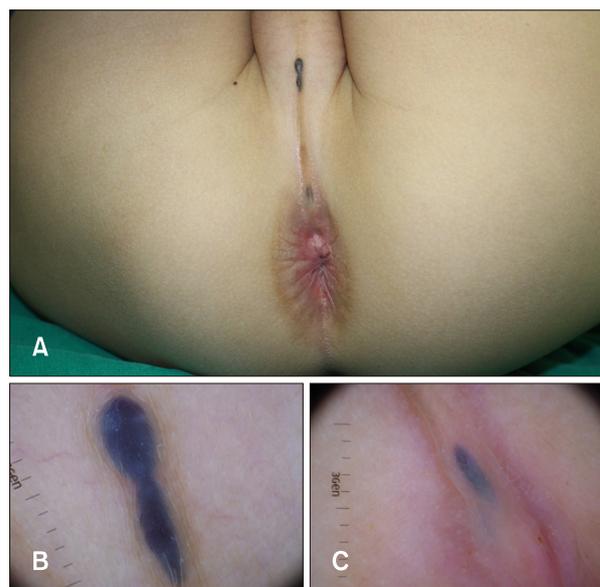


Fig. 1. (A) Brown and black linear plaques on the scrotum and perianal region. Dermoscopic findings of the dark blue to black plaque on the (B) scrotum and (C) perianal region.

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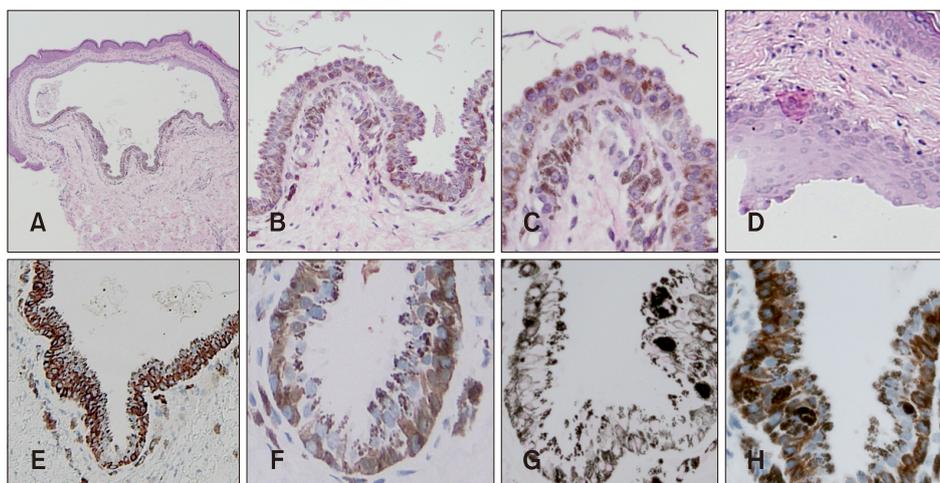


Fig. 2. Histopathologic findings showing a cystic cavity (A: H&E, $\times 40$) that is lined by cuboidal epithelium containing melanin pigments (H&E, B: $\times 100$, C: $\times 200$) and stratified squamous epithelium (D: H&E, $\times 100$). Immunohistochemical staining demonstrates diffuse staining of (E) c-kit ($\times 40$), (F) S-100 ($\times 200$), (G) Fontana-Masson ($\times 200$), and (H) 8 Melan-A staining ($\times 200$).

tributes to melanocytic colonization in MRC.

Our case has unique characteristics, including the unusual clinical features of pigmentation and the perianal localization. Other distinctive features were atypical histopathologic findings of not only a mixed epithelial lining of the cyst wall but also pigmentation.

From 1913 to 2010, 226 cases of MRC were reported. Melanin pigments and/or melanocytes on histology were reported in only seven cases, including the present case. Among these, ours is the only case demonstrating a mixed lining of cuboidal and stratified squamous epithelial cells. Moreover, this is a meaningful case because it suggests the first evidence supporting the hypothesis that c-kit may contribute to melanocytic colonization in PMRC.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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