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A Case of Multiple Canaliform Median Raphe Cysts Showing a Mixed Type Lining of Epithelium: A Case Report and Review of the Literature

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Dear Editor:

A 7-month-old boy presented with linearly arranged, multiple pinhead-sized to rice-sized whitish cysts along the median raphe from the perineum to the penile shaft (Fig. 1), which were present from birth and progressively increased in size and number. The lesions spread along the perineal raphe and had a cordlike appearance. The infant showed no signs of pain, tenderness, pruritus, or other symptoms. He had no history of medical disease or congenital anomaly, and the rest of the skin was normal. A skin biopsy was performed at the upper cystic portion of the lesion (Fig. 1), and the biopsy specimen showed the cysts located in the dermis with variable lining. Some parts were lined by stratified squamous epithelium with well-formed granular layer and keratin flakes. Other parts consisted of cuboidal urothelium-like epithelium with mucinous cells (Fig. 2). Thus, considering the site and the histologic features, a diagnosis of mixed-type median raphe cysts was made.

Median raphe cysts are rare, benign congenital lesions that can develop anywhere along the midline of the ventral side of the male genital area. They can form from the urethral meatus to the anus and the perineum along the perineal raphe[1]. Histologically, they are classified into four types. The urethral type, the most common, consists of an urothelium-like epithelium, with a layer of columnar cells overlaid with several stratified layers of uniform small cells. The epidermoid type consists of stratified squamous cell epithelium. The glandular type consists of a well-formed intraepithelial glandular structure in the lining of the urethral epithelium. The mixed type, the second most common and the type of this case, consists of more than one type of epithelium, including the urethral epithelium with squamous metaplasia, urethral epithelium with mucinous cells, or a combination of these three[1].

Histomorphological features of the median raphe cyst are considered to be related to its embryonic origin and pathogenesis[2]. The pathogenesis is unclear, but several hypotheses have been proposed. First, the 'tissue trapping'

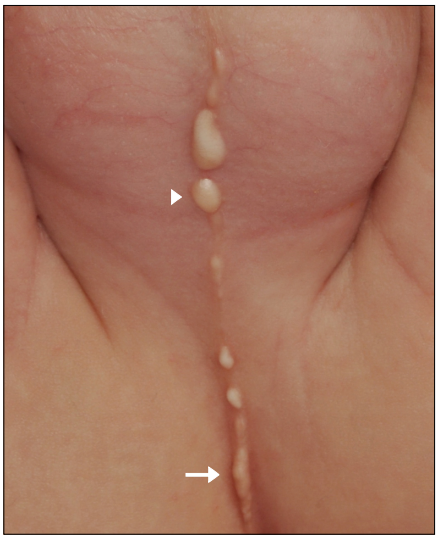

Fig. 1. Linearly arranged, multiple, whitish cysts along the perineal raphe. The arrow indicates the canalicular appearance of the cysts. The arrowhead indicates the biopsy site.

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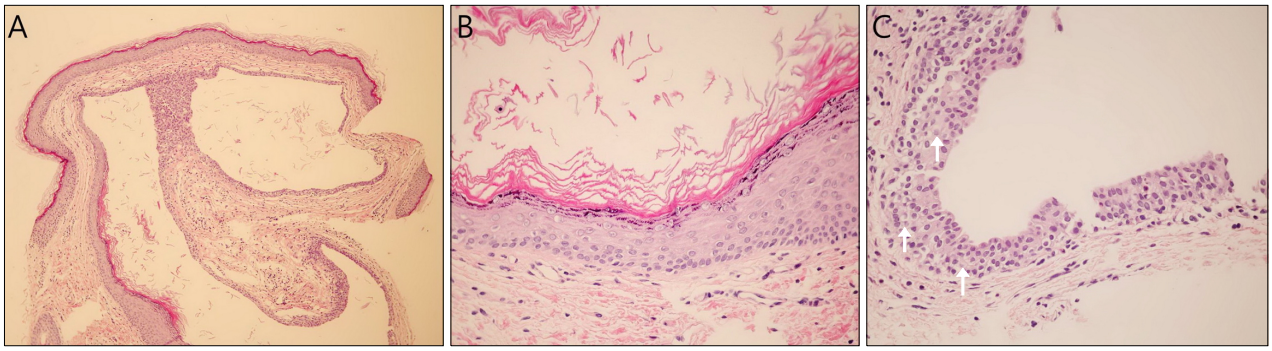

Fig. 2. (A) The biopsy revealed cysts located in the dermis that showed variable lining (H&E, ×100). (B) Focal part of the cyst wall lined by stratified squamous epithelium with well-formed granular layer and keratin flakes (H&E, ×400). (C) Focal part of the cyst wall lined by cuboidal urothelium-like epithelium with mucinous cells (H&E, ×400). The arrows indicate the mucinous cells.

theory states that median raphe cysts are caused by either a defective fusion of the urethral folds or an anomalous outgrowth of the epithelium during the development of the urethra[2]. Second, median raphe cysts may result from the anomalous developmental rest of the periurethral glands of Littre. This hypothesis may help explain glandular type median raphe cysts and the involvement of mucinous cells in some cases[3]. A third hypothesis suggests that blockage of the paraurethral ducts may underlie the development of median raphe cysts[4].

The canaliform median raphe cysts demonstrated in the present case are an uncommon presentation of this rare condition, and reports of these cysts are rare. Most patients present with a single isolated from or a few cysts. The patient described here presented with multiple, continuous cysts. This unique morphology makes diagnosis confusing due to its rarity and particular clinical features. Thus, histologic confirmation is often required[5].

Complete local excision is recommended for the symptomatic lesions to prevent possible complications. Furthermore, whether associated congenital anomalies are present should also be assessed.

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