Median raphe cysts are rare congenital anomalies that were first reported in 1895 by Memert.\textsuperscript{1} Median raphe cysts can be found along the distal penis and scrotum, towards the perineum in a midline position.\textsuperscript{2} Although the embryologic etiology of median raphe cysts has not been clearly established, they are believed to occur during embryologic development of the male genitalia.\textsuperscript{3-5} Median raphe cysts can be diagnosed by assessment of patient history and physical examination. However, it can be difficult to diagnose this condition in childhood if it remains asymptomatic. We report the case of a median raphe cyst along the length of the scrotum and perineum with recurrent infection and purulent discharge in a 28-year-old man.

**CASE REPORT**

A 28-year-old male patient presented with recurrent swelling along the midline of the perineum and scrotum. He complained of purulent discharge at the proximal end of a linear lesion on the scrotum and perineum median raphe. Acute swelling of the linear skin lesion with purulent discharge and spontaneous healing occurred two to three times a year. The patient and his mother thought that the lesion was a recurrent skin infection when the symptoms first developed at the age of 5 years. He had no history of trauma or other remarkable medical or family history. Physical examination showed no swelling of the lesion,
and the scrotum and perineum seemed to have an almost normal shape except for the fistula opening on the mid-scrotum (Fig. 1A). To identify the lesion, we injected normal saline through the fistula opening. Several 5-7 mm cystic lesions with linear alignment were observed from the mid-portion of the scrotum to the perineum (Fig. 1B). Ultrasonography showed no malformation of the genitourinary tract. The lesion was excised under spinal anesthesia after administration of prophylactic empirical antibiotics using second generation cephalosporin. We identified the midline cystic structures between the dartos fascia and dermis in the midline of the scrotum and perineum (Fig. 2A). The lesion did not communicate to other sites such as the urethra or anal canal. Histology revealed multiple canalized cysts lined with squamous epithelium (Fig. 2B). The epidermoid type of median raphe cyst between the scrotum and the perineum was diagnosed finally. The postoperative course was uneventful with no sign of recurrence at the 6-month follow-up. We are planning to follow up annually through physical examination on external genitalia.

**DISCUSSION**

Median raphe cysts usually present as solitary or multiple cystic lesions, or canal-like lesions along the midline of the genital area. The lesion is typically located along the midline from the penile area to the perineal area. Although various shapes and sizes have been reported, median raphe cysts never communicate to the urethra and surrounding tissues. In our case, we observed coexistence of cystic lesions and canal-like lesions on the scrotum and perineum.
This congenital anomaly was easily diagnosed based on the patient’s medical history and physical examination results. However, the median raphe cysts were usually diagnosed in adults as this anomaly is usually silent in childhood unless accompanied by infection, ulceration, or traumatic injury.\cite{1,2,3} Therefore, median raphe cysts are likely under-reported and under-recognized for pediatric and adult urologists. In some cases, the patient may present for cosmetic reasons or for obtaining a diagnosis without symptoms. Although asymptomatic until adulthood, this lesion can become traumatized or infected by Neisseria gonorrhoeae or Staphylococcus aureus producing swelling, tenderness, and purulent discharge.\cite{6}

Median raphe cysts have a wide histological classification that includes stratified, pseudostratified, and squamous columnar epithelium, resembling the epithelium of the male urethra.\cite{7} Three representative histologic patterns have been described: (1) urethral type lined with pseudostratified columnar epithelium; (2) epidermoid type with squamous epithelium; and (3) mixed type associated with both types of epithelium.\cite{8,9} In our case, the inner layer of the lesion was composed of squamous epithelium and hence it was classified as the epidermoid type.

Although the etiology of median raphe cysts has not been clearly established, they are believed to be a congenital defect that occurs during embryologic development of the male genitalia.\cite{3,4,5} There are two major theories regarding its etiology. The first theory suggests that the median raphe cyst is a result of embryologic outgrowth of the epithelium after primary closure of the urethral or genital folds. The second theory suggests that the lesion develops from epithelial remains caused by incomplete closure of the folds.\cite{10}

The differential diagnosis of a median raphe cyst includes urethral diverticulum, molluscum contagiosum, syringoma, sebaceous cystoma, glomus tumor, and pilonidal cyst. However, accurate diagnosis can be obtained based on clinical findings and histologic features.

Observation of the patient may be a reasonable choice in asymptomatic cases. In patients presenting with symptoms or for cosmetic reasons, surgical excision is recommended to prevent symptoms derived from its location, size, and secondary infection.\cite{2,4,9}

We report a case of median raphe cysts of the scrotum and the perineum presenting with recurrent infection. In conclusion, median raphe cysts are rare findings that are likely under-reported and under-recognized. Therefore, we believe that it is important that urologists be aware of this condition and its appropriate management.

**CONFLICT OF INTEREST**

No potential conflict of interest relevant to this article was reported.

**REFERENCES**