Case Presentation

Unusual location of median raphe cyst presenting as perianal polyp: a case report

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Abstract

Median raphe cysts (MRC) are uncommon, benign congenital lesions that may present anywhere in the midline between the urinary meatus and the anus, with the shaft of the penis and the glans penis being the most common sites. We report a 52-year-old man with median raphe cyst unusually located in perianal region and treated by surgical excision.

Key Words: Median raphe cyst, perianal polyp, cyst, perianal region

Introduction

Median raphe cysts (MRC) are uncommon, benign congenital growths that may present anywhere in the midline between the urinary meatus and the anus, with the shaft of the penis and the glans penis being the most common sites. Swelling, tenderness and purulent discharge can be seen when cysts become damaged and infected [1]. The differential diagnosis includes epithelial cysts, sebaceous cysts, glomus tumors, dermoid cysts, urethral diverticulae, and pilonidal cysts [2]. In the presence of symptoms or if there is cosmetic concern, treatment of MRC entails surgical excision, whereas small and asymptomatic lesions might be followed [3]. We report herein a man with median raphe cyst unusually located in perianal region and treated by surgical excision.

Case synopsis

A 52-year-old man presented with a mass in the perianal region. This was a relatively large lump and the patient said that it was present in that location for about 10 years without causing any symptoms except for slight discomfort. The lesion was a 3.5 x 4 x 4 cm sized mobile, non-tender, cystic, well-defined swelling, which was located at the posterior midline in the perianal region (at 12 o’clock in jack-knife position) (Figure 1). During surgical removal, the lesion was disrupted at its base, resulting in the release of cloudy pale yellow fluid. Under spinal anesthesia, the entire mass was excised by making an elliptical incision and the wound was closed primarily. No complications occurred and the patient recovered uneventfully. During 12 month follow-up no recurrence was observed.
Histopathological Findings

Macroscopic findings

Polypoid lesion was 3x0.7x3 cm sized and covered by anal skin. It contained a unilocular cyst with cloudy pale brown fluid. Thickness of the cyst wall was 0.3 cm.

Microscopic findings

A cystic lesion was located in dermis and covered by non-keratinized squamous stratified or pseudostratified columnar epithelium not associated with any dermal structure (Figure 2, 3). Lining epithelium showed squamous metaplasia in some regions; no mucinous glands were associated with the cyst wall.

Figure 1. Cystic lesion in perianal region at 12 o’clock in jack-knife position.

Figure 2. Epidermis below covering polyp and columnar epithelium above covering cyst.
**Immunohistochemistry**

Immunohistochemistry study showed an epithelial lining that stained positive for cytokeratin 7 (CK7) in the columnar cells, but stained negative in squamous cells. The cytokeratin 20 (CK20) was negative throughout all fields. There was strong staining for carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA). S-100 and estrogen receptor (ER) staining was slightly positive and smooth muscle antibody (SMA), desmin, and progesterone receptor (PR) were negative for squamous epithelium.

**Discussion**

Median raphe cyst is an uncommon congenital lesion occurring anywhere along the midline of the ventral side of the male genital area to the anus and the perineum. They are usually found in the first 3 decades of life [4]. The diagnosis of MRC is difficult and the differential diagnosis includes epidermal cyst, sebaceous cyst, glomus tumor, dermoid cyst, urethral diverticulum, and pilonidal cyst [1]. The pathogenesis of MRC has not yet been well understood although there have been some theories on its development [5].

MRC may present as a perianal polyp especially in the elderly, which makes their diagnosis difficult since hypertrophied papilla, viral warts, and hemorrhoids may have a similar macroscopic appearance and location [4]. Thus, median raphe cysts should be considered in the differential diagnosis of polyp-like lesions in perianal region. The epithelial lining of the MRC, as in our case, includes columnar stratified, pseudostratified, or squamous cells resembling the histological features of the different portions of the male urethra.

Hara et al. reported that MRC can express both estrogen and progesterone receptors [6]. However, in our case only estrogen receptor was expressed indicating that MRC may differ in immunohistochemical features. According to Shao et al’s classification, our case was classified as mixed type of MRC [7].

All of the MRCs reported previously the literature were those located between the anus and the scrotum [3,4,8], whilst the MRC in our case was found located between the anus and the sacrococcygeal region at 12 o’clock position in the jack-knife position, resembling a hemorrhoidal lesion. This difference was the unique feature of our case.

**Conclusion**

Median raphe cysts are uncommon benign congenital lesions that should be kept in mind in the differential diagnosis of growths located in perianal region.

**References**
