Pigmented Median Raphe Cysts of the Penis

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Two cases of median raphe cysts of the penis with melanosis are presented. The presence of melanocytes was observed in the lining of the cysts by light and electron microscopy. The possible mechanism of the embryological development of the cysts is discussed in the context of the published literature.

Key words: median raphe cyst; pigmentation; melanocytes.

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Median raphe cyst of the penis is an uncommon entity. It is located on the ventral surface of the penis. Herein, we report 2 cases of this entity that displayed diffuse melanosis of the lining of the cysts. Pigmentation in the median raphe cysts due to the presence of melanocytes has rarely been documented.

CASE REPORTS

Case 1

A 4-year-old Japanese boy presented with cystic lesions on the ventral midline of the penis from birth. Examination of his genitalia revealed multilobulated, freely mobile, cystic papules, 2 – 4 mm in diameter, on the ventral surface of the penis from the frenulum to the anus. These cysts were translucent and brown – black in color (Fig. 1). One large cystic lesion was surgically removed for histopathological examination. An electrodesiccation was carried out for the remainder of the lesions.

Histopathology

The histopathological changes in Cases 1 and 2 were similar and are described together. An irregularly shaped empty cystic space was observed beneath the overlying surface epidermis. No connection with the epidermis was evident. On examination, 1 lesion was 1 mm in diameter in the ventral midline and the other was 5 mm in diameter beside the frenulum preputi penis. These cysts were brown – black, translucent cystic lesions and were connected with a sinusal cord. One large cyst in the ventral midline was excised.

CASE REPORTS

Case 2

A 5-year-old Japanese boy presented with 2 small cystic lesions on the ventral surface of the glans penis from birth. His mother denied any history of trauma or infection. On examination, 1 lesion was 1 mm in diameter in the ventral midline and the other was 5 mm in diameter beside the frenulum preputi penis. These cysts were brown – black, translucent cystic lesions and were connected with a sinusal cord. One large cyst in the ventral midline was excised.

Histopathology

The histopathological changes in Cases 1 and 2 were similar and are described together. An irregularly shaped empty cystic space was observed beneath the overlying surface epidermis. No connection with the epidermis was evident. The wall of the cysts consisted of a pseudostratified columnar epithelium, which varied from 1 to several layers thick. Some of the cells lining the cysts contained a variable amount of colloidal iron stain-positive and periodic acid Schiff stain after digestion with diastase-positive material in the cytoplasm on the luminal side of the cells and in the lumen of the cysts. No myoepithelial cells or decapitation secretion were observed. The basal layer of the epithelia was filled with dark brown granules that were also noted throughout the epithelial lining. Scattered among the epithelial cells were large vacuolated cells. The pigment in the epithelial cells and large vacuolated cells was negative for iron, but strongly positive with Fontana–Masson stain for melanin. Numerous dendritic melanocytes were interspersed between the epithelial cells (Fig. 2). The surrounding stroma contained a number of melanophages.

An electron microscopic examination was performed in Case 2 and showed 2 types of cells: secretory cells and basal cells. The secretory cells were columnar and showed large basal nuclei and occasional prominent nucleoli. Mucus secretion was noted in some of the columnar epithelial cells. The luminal surface of the columnar cells displayed well-developed microvilli (Fig. 3). No secretory apocrine-type granules, which show a densely packed and uniform internal structure, were noted in these cells. The basal cells were located beneath the secretory cells and the basement membrane and dendritic melanocytes were interspersed with the basal cells. Interspersed with the epithelial cells were numerous melanocytes. These were recognizable due to the presence of a number of cytoplasmic melanosomes and premelanosomes and the lack of intercellular junctions with the adjacent columnar cells. A prominent feature was the presence of apical caps over the free surface of the cells. These consisted of dome-shaped portions of cytoplasm that projected into the lumen and contained a large number of compound melanosomes as well as numerous vesicles and vacuoles. In some areas, the lumen showed detached fragments of cytoplasm that appeared to pinch off fragments of the apical cytoplasmic caps. The separated caps contained numerous compound melanosomes, cytoplasmic vesicles.
DISCUSSION

Median raphe cysts usually arise in young men and are located on the ventral surface of the penis, most commonly near the glans. Median raphe cyst of the penis has been erroneously reported as apocrine cystadenoma of the penis (1, 2). The present cases showed clinical and histopathological features consistent with median raphe cysts of the penis as the epithelial lining, observed by light and electron microscopy, failed to show evidence of apocrine-type secretory activity or myoepithelial cells as seen in apocrine cystadenoma. Apocrine cystadenoma is pigmented more often than is realized: up to 50% of the reported cases are clinically pigmented (3). However, median raphe cysts with melanosis are uncommon. Although the cause of pigmentation is not fully understood, this pigmentation may be due to the presence of lipochrome (4), the Tyndall phenomenon (5) or the presence of melanocytes (6). The distinctive feature in our cases was the presence of pigmentation in the lining of the cysts. It was concluded that this pigment was due to the presence of melanocytes and melanosomes based on the light and electron microscopic results. Hitti et al. (6) reported the presence of melanocytes using electron and light microscopy.

Some theories have been proposed to explain the origin of the median raphe cyst. The first possible explanation is embryologic defective closure of the median raphe (7). The second theory is anomalous development of ectopic periurethral glands of Littre (8). The third theory, proposed by Paslin (9), hypothesizes an anomalous budding and separation of the urethral columnar epithelium from the urethra. Fetissof et al. (10) studied 4 cases of median raphe cyst of the penis and found that in 1 of the 4 cases numerous melanocytes but no serotonin-storing cells were observed on the lining of the cyst. Because the glans portion of the urethra, which is ectodermal in origin, was devoid of endocrine cells and contained melanocytes the authors concluded that this cyst was probably an ectodermally-derived cyst. Romani et al. (11) reported a case of this entity with the peculiar feature of ciliated cells. These ciliated cells are thought to have an ectodermal origin. Most reports of median raphe cyst of the penis do not give details of melanin stains. To our knowledge, the present cases are only the second and third detailed reports of median raphe cyst of the penis with prominent melanosis. Median raphe cyst of the penis has a broad spectrum of histopathological appearances. It is suggested that some cysts arise from the endodermal part of the urethra (10). Further studies may have a bearing on the embryological development of the close inter-relationship between the melanocytes and columnar epithelial cells in this cyst.

REFERENCES


Fig. 2. Case 1: dendritic melanocytes are identified at the base of the epithelial lining and dark brown granules are noted in the pseudostatified columnar epithelium. Fontana – Masson stain (original magnification × 400).

Fig. 3. Low magnification electron micrograph showing secretory cells with microvilli. Note the detached cells in the lumen. Original magnification × 2,000.