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Case report

Incidental vaginal müllerianosis

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ABSTRACT

Müllerianosis is the term used to designate lesions composed of an admixture of two or three types of müllerian-derivation glands in heterotopic location. In this report, we describe a case of incidental vaginal müllerianosis in a 59-year-old woman who underwent rectosigmoidectomy for rectal adenocarcinoma. In the vaginal cuff removed for neoplastic invasion, a separate multilocular mass measuring 1.5 cm was found. The microscopic examination of the vaginal wall revealed endosalpingeal, endocervical and endometrial dilated or cystic glands with predominance of the endosalpingeal epithelium. Müllerian epithelium showed positivity for cytokeratins 7 and 8/18, high molecular weight cytokeratin, estrogen receptor alpha, and androgen receptor. The periglandular stroma was condensed and reactive for smooth-muscle actin, h-caldesmon, and CD10. To the best of our knowledge, a case of vaginal müllerianosis has not been previously reported. This lesion should be differentiated form vaginal adenosis and primary well-differentiated vaginal adenocarcinoma. The vagina should be added to the list of locations in which müllerianosis can be observed.

1. Introduction

Müllerianosis is a rare entity consisting of an organoid admixture of two or three types of müllerian-derivation glands (cervical, endometrial, or tubaric) in heterotopic location [1,2]. This benign tumor-like lesion has mostly been found in the urinary bladder [2,3], but diverse reports have documented its presence in other locations, such as pelvic peritoneum [1,4], mesosalpinx [5], ureter [6], inguinal lymph nodes [7], and spinal cord [8]. To the best of our knowledge, a case of vaginal müllerianosis has not been reported.

We herein present a unique case of incidental vaginal müllerianosis in a patient operated on for rectal adenocarcinoma and discuss the clinical implications and pathogenesis.

2. Case report

This report presents the case of a 59-year-old woman with a past medical history significant for uterine leiomyomas for which she underwent hysterectomy nine years earlier, and breast carcinoma treated with lumpectomy. The patient was referred to the colorectal surgery unit with a diagnosis of adenocarcinoma of the high portion of the rectum for surgical treatment. She underwent rectosigmoidectomy (Hartmann's procedure) with complete resection of the mesorectum, removal of a vaginal cuff infiltrated by the neoplasia, and lymphadenectomy, including inguinal lymph node dissection.

The surgical specimen received consisted of a segment of rectosigmoid that measured 29 cm in length. It showed an infiltrating poorly differentiated tubular adenocarcinoma of the rectum pT4b, pN2b of the AJCC classification.

The vaginal portion measured 5 cm × 2 cm and showed a 1.5-cm multicystic mass in its wall. The cystic mass on cutting showed locules of varying size filled with clear fluid (Fig. 1).

Microscopic examination of the vaginal segment showed an infiltrating poorly differentiated (rectal) adenocarcinoma and a separated multicystic lesion. The latter lesion was ill-defined. Glands were embedded in a dense stroma which differed from that of the vaginal wall (Fig. 2A). Glands and cysts were lined by variable types of epithelium, the predominant being ciliated columnar epithelium of the tubal type (Fig. 2B). Focally endocervical-type (Fig. 2C) and endometrial-type (Fig. 2D) glandular and cystic structures were seen. The endometrial glands often showed a cuboidal epithelium. Mild glandular cellular atypia was occasionally observed. No mitotic activity was seen. The cystic spaces were often filled with eosinophilic secretion (Fig. 2B). The epithelial structures were surrounded by a condensed stroma. The rest of the interglandular stroma was variable in nature. There were fibrous, myofibromatous, and occasional densely cellular areas resembling

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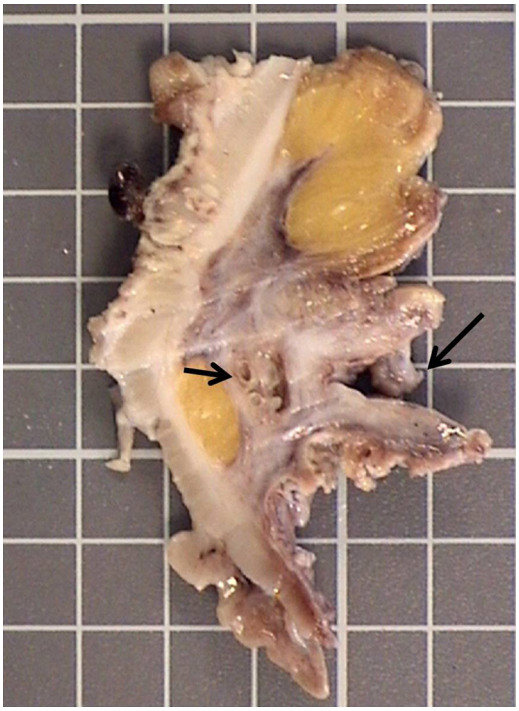

Fig. 1. Gross section of the recto-vaginal septum showing a multiloculated cystic mass in the vaginal wall (short arrow). A long arrow points out to the vaginal mucosa.

endometrial stroma. The immunohistochemical study revealed that the müllerian epithelium showed positivity for cytokeratin 7 (Fig. 3A), cytokeratin 8/18, high molecular weight cytokeratin, estrogen receptor alpha (Fig. 3B), and androgen receptor; and negativity for cytokeratin 20 and carcinoembryonic antigen (CEA). The periglandular stroma displayed diffuse positivity for smooth muscle actin (Fig. 3C), h-caldesmon, and CD10 (Fig. 3D), and focal positivity for estrogen receptor alpha, progesterone receptor, and androgen receptor. Table 1 summarizes the antibodies used in this study.

Postoperatively, she developed a pelvic abscess that was drained. Three months after the surgery, she is asymptomatic and doing well.

3. Discussion

We have reported a case of müllerianosis involving the wall of the vagina in a woman who had undergone hysterectomy several years earlier. Normally, the vagina is devoid of glands. The cystic and glandular structures in the vaginal wall of our patient showed three types of müllerian epithelia, namely tubal, endocervical, and endometrial. This feature is in accord with the term müllerianosis we have used. However, it is noteworthy to mention that endosalpingiosis was the predominant component of the lesion.

The clinical presentation of our case was dominated by the rectal adenocarcinoma. The vaginal müllerianosis was an incidental finding associated with the neoplastic invasion of the vaginal wall. However, both lesions were completely separated. On the other hand, despite the tumor-like appearance, the infiltrative pattern of the müllerian lesion and the focal mild cytologic atypia of the epithelium, the close resemblance of the structure to endosalpingiosis, endocervicosis, and endometriosis, and the absence of mitotic figures facilitated the correct diagnosis.

Surgical excision of the müllerianosis is considered curative, and no recurrence should be expected.

As far as we are aware, there have been no prior descriptions of müllerianosis of the vagina. However, endocervicosis or endometriosis has been reported in this location. Thus, Martinka et al. [9] described a case of endocervicosis presenting as a painful vaginal mass in a 36-year-old woman, and Choi et al. [10] reported a post hysterectomy vaginal vault endometriosis in a 45-year-old woman.

The differential diagnosis of vaginal müllerianosis includes vaginal adenosis and primary well-differentiated vaginal adenocarcinoma. Vaginal adenosis differs from the lesion described herein in that it does not form a tumor-like mass, the glands are confined to the mucosa or superficial stroma, and these glands lack an infiltrative pattern and a periglandular stromal response [11]. Primary vaginal adenocarcinoma includes clear cell, endometrioid, mucinous, intestinal, and mesonephric types. These neoplasms show variable degrees of architectural and cytologic atypia, and usually a high proliferative index. Nonetheless, nonclear-cell primary vaginal adenocarcinomas are rare.

Endocervicosis may become malignant, but this complication is very uncommon [12,13]. The vagina is a rare location of endometriosis. Stern et al. found that only 2% of 1323

Table 1
Antibodies used in this study.

Antibody	Source	Clone	Dilution	Retrieval solution pH (Dako[a])
CK 7	Dako[a]	OV-TL 12/30	FLEX RTU[b]	High
CK 8/18	Dako[a]	EP17/EP30	FLEX RTU[b]	High
High molecular weight CK	Dako[a]	34-beta-E12	FLEX RTU[b]	High
CK20	Dako[a]	K_S20.8	FLEX RTU[b]	High
Estrogen receptor alpha	Dako[a]	EP1	FLEX RTU[b]	High
Progesterone receptor	Dako[a]	PgR636	FLEX RTU[b]	High
Androgen receptor	Dako[a]	AR441	1:50	High
Carcinoembryonic antigen	Dako[a]	II-7	FLEX RTU[b]	High
Smooth muscle actin	Dako[a]	1A4	FLEX RTU[b]	High
h-Caldesmon	Dako[a]	h-CD	FLEX RTU[b]	High
CD10	Dako[a]	56C6	FLEX RTU[b]	High

CK, cytokeratin.
[a] Glostrup, Denmark.
[b] Ready-to-use.