

Ciliated Cyst of the Vulva: Oestrogen and Progesterone Receptors

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Accepted March 5, 2008.

Sir,

Cases of cutaneous cysts lined by ciliated epithelium have rarely been documented, and only a few cases of such cysts in the vulvar vestibulae have been reported (1–3). The origin of the cysts is controversial because the location is so anatomically specific. In this report, we describe a 14-year-old girl with a ciliated cyst of the vulva (CCV), the lining cells of which were positive for oestrogen and progesterone receptors, and discuss the pathogenesis of ciliated epithelium.

CASE REPORT

A 14-year-old Japanese girl was referred to our outpatient clinic for the evaluation of a 2-month history of a pedunculated mass on the vulva. The mass was elastic hard, measuring 2 cm in diameter, and was pedunculated on the mid-portion of the right labium minus. It had a slack skin surface and was slightly pigmented (Fig. 1). The patient was healthy and laboratory investigations showed no abnormal haematological results and no history of either oestrogen or progesterone treatment. Under local anaesthesia, the patient underwent complete resection of the vulvar mass. Histopathological examination revealed that the mass was a submucosal cyst lined by a single layer and partly pseudostratified by 4–5 layers of cuboidal and columnar epithelium with ciliated cells, resembling Müllerian epithelium, with occasional papillary projections (Figs 2 and 3). No smooth muscle was identified in the supporting tissues beneath the lining epithelium. Only the luminal border of the cells stained faintly with Alcian-blue and Periodic acid-Schiff (PAS) stain even after diastase digestion. Immunohistochemically, the lining epithelium was positive for epithelial membrane antigen (EMA), oestrogen receptor (ER), progesterone receptor (PR) and cytokeratin (CK) 7, and negative for S-100 protein, carcinoembryonic antigen (CEA), CK 20 and α -smooth muscle actin. The patient was diagnosed as having a CCV.



Fig. 1. Clinical appearance of ciliated cyst of the vulva.

DISCUSSION

Cases of CCV (also known as paramesonephric cysts) are reported in the 25–35-year-old age group of young women and are occasionally associated with pregnancy or exogenous progesterone. Only a few case of CCV in the vulva vestibulae have been reported (1–3). Clinically, the cysts contained fluid, occurred on the superior portion of the labia minora and measured 1–3 cm. No cases of either multifocal lesions or recurrences have been reported. Excision of the lesion should be curative. The aetiology is unknown; however, heterotopia (sequestration and migration of Müllerian tissue during embryogenesis), dysontogenesis (defective embryonic development), prosoplasia (abnormal development resulting in organization), and metaplasia have been suggested as mechanisms for the origin of the cyst. Of these hypotheses, Müllerian heterotopia has been most commonly proposed to be the basis of similar histopathological features of the epithelium (4, 5).

The origin of the ciliated epithelium of CCV is unclear, although there are 3 theories: (i) displacement of Müllerian duct remnants, (ii) ciliated epithelial cell metaplasia

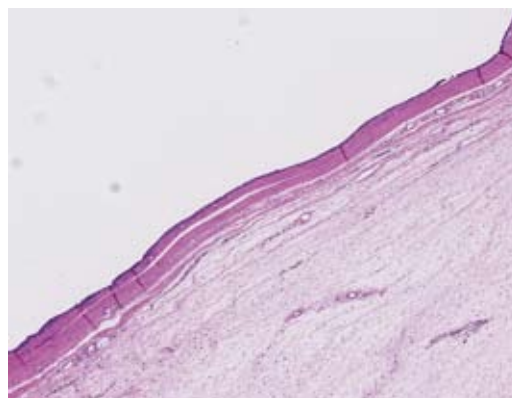


Fig. 2. The wall of the lesion was composed of both epithelial and loose connective tissue elements (haematoxylin-eosin stain (H&E), $\times 40$).

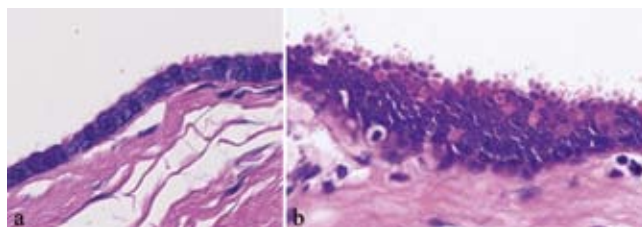


Fig. 3. The lining epithelium of the cyst is composed mainly of (a) a single row and (b) pseudostratified cuboidal and columnar cells intermingled with ciliated cells (H&E stain, $\times 400$).

Table I. Immunohistochemical analysis for ciliated cysts

Diagnosis of the cyst	EMA	S-100	CEA	ER	PR	CK7	CK20	Reference
Cutaneous ciliated cyst	+	+ (<10%)	-	+ (<10%)	NT	NT	NT	Yokozaki et al. (10)
Cutaneous ciliated cyst	+	-	-	+	NT	NT	NT	Fontaine et al. (8)
Ciliated cyst of the vulva	+	-	-	NT	NT	NT	NT	Kang et al. (2)
Ciliated cyst of the vulva	+	-	-	-	NT	NT	NT	Hamada et al. (3)
Peritoneal Mullerian cyst	+	NT	-	+	+	+	-	Konishi et al. (14)
Parafallopian tube transitional cell carcinoma	NT	NT	NT	+	+	+	-	Paner et al. (15)
Ciliated cyst of the vulva (the present case)	+	-	-	+	+	+	-	

EMA: epithelial membrane antigen; S-100: S-100 protein; CEA: carcinoembryonic antigen; ER: oestrogen receptor; PR: progesterone receptor; CK 7: cytokeratin 7; CK 20: cytokeratin 20; NT: not tested.

of the sweat gland, and (iii) urogenital sinus origin. There are the following possibilities: heterotopia (embryonic displacement of tissue in other areas) dysontogenesis (defective embryonic development of individuals), prosoplasia (progressive transformation), and ciliated epithelial cell metaplasia. Among these, the possibility of heterotopia (misplacement of Müllerian duct tissue) is considered high (1, 5). The epithelium of the fallopian tube consists of a single layer of ciliated cells, non-ciliated columnar secretory cells, and intercalated cells.

The cyst wall in the present case was lined by a single row or pseudostratified columnar or cuboidal cells which exhibit cilia on the luminal aspect, showing similar features to the fallopian tube histologically, and was hypothesized to be oestrogen-dependent Müllerian heterotopia during embryogenesis. In fact, ciliogenesis produced by oestrogen administration has been reported in various animals (6).

In contrast, cutaneous ciliated cyst (CCC) is found very rarely in females as a single lesion, largely in the lower extremities, and even more rarely in males and on the back (7, 8). CCC was reported to rapidly increase in size during pregnancy, suggesting that these lesions are sensitive to hormonal stimulation (9). Epithelial cells in CCC demonstrated nuclear staining for oestrogen receptor (7, 8, 10). CCC is considered by some authors to be the same entity as CCV (11). The detection of intranuclear ER and PR within the CCC lends strong support that hormonal stimulation plays a role in the development of these lesions.

Leonforte (12) reported a case of cutaneous ciliated cystadenoma with columnar ciliated cells and apical caps in the heel of a man that was thought to have originated as sweat gland metaplasia. However, the present case might not originate from sweat gland metaplasia on the basis of negative staining for CEA and S-100, and the absence of histological features such as decapitation or apical formation. Therefore, there is only a slight possibility that CCV originated from ciliated metaplasia of eccrine or apocrine glands in the present case.

Robboy et al. (13) reported 11 mucinous and ciliated cysts on the vulva, suggesting urogenital sinus origin (4, 13). Previously, the presence of ER on the lining epithelium of CCV was evaluated only by Hamada et al. (3), whose case was negative for ER, and CEA; therefore, their case might have been of urogenital sinus origin.

In contrast, both reported cases of retroperitoneal Müllerian cyst (14) and parafallopian tube transitional cell carcinoma (15) showed that its lining cells were positive for CK 7, EMA, ER and PR, and negative for CK 20, CEA on immunohistochemical study, the same as the present case, suggesting that the lesion originated from the developing Müllerian apparatus (Table I).

The present case is the first case of CCV with the lining epithelium positive for ER and PR. In conclusion, these histopathological and immunohistochemical findings strongly support the hypothesis that CCV is Müllerian heterotopia.

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