

## A Case of Cutaneous Ciliated Cyst with Immunohistochemical Evidence for Müllerian Origin

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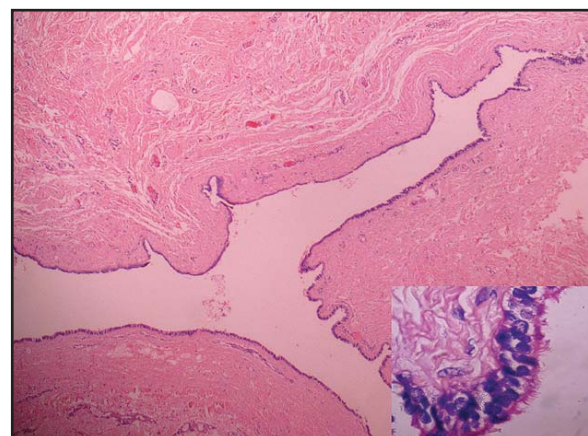
### Abstract

**Observation:** Cutaneous ciliated cysts (CCC) are rare developmental lesions that are generally located on the lower limbs of young females. As their lining closely resemble Fallopian tube epithelium, they are regarded as Müllerian heterotopias. Their estrogen receptor (ER) and progesterone receptor (PR) expressions also support this theory. Here we report an other example of CCC located on the left buttock of a 12-year-old girl that occurred as a small subcutaneous mass shortly after puberty. The cyst was lined by pseudostratified ciliated epithelium devoid of goblet cells and had a fibrous wall. There were no muscle bundles in the wall. The epithelium expressed EMA, ER and PR, but not CEA immunohistochemically.

### Introduction

Cutaneous ciliated cysts (CCC) typically occur in young women during their adolescent or reproductive ages [1]. They are usually less than 3 cm in diameter. The pathogenesis is unknown but strong association with the female gender, location on lower limbs, frequent growth in reproductive years and immunohistochemical expression of estrogen (ER) and progesterone receptors (PR) favor the heterotopia of the Müllerian epithelium theory [2, 3, 4, 5, 6, 7, 8, 9]. Ovarian hormones probably stimulate their growth and they become clinically apparent. They rarely occur postmenopausally and the hormonal imbalance may be the stimulating factor [7]. Ciliated metaplasia of eccrine glands is the alternative theory for the formation of the lesions those are deeply situated in different parts of the body in males with immunohistochemical expression of carcinoembryonic

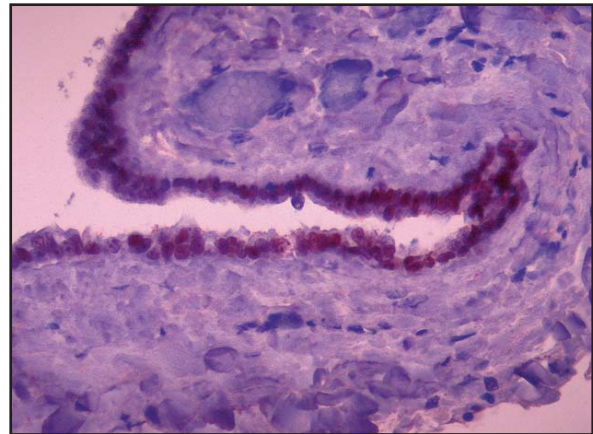
antigen (CEA) [9]. Bronchogenic cyst, branchial cleft cyst, thymic cyst, thyroglossal duct cyst, vulvar and perineal cysts, and cutaneous endosalpingiosis are encountered in the differential diagnosis [1].



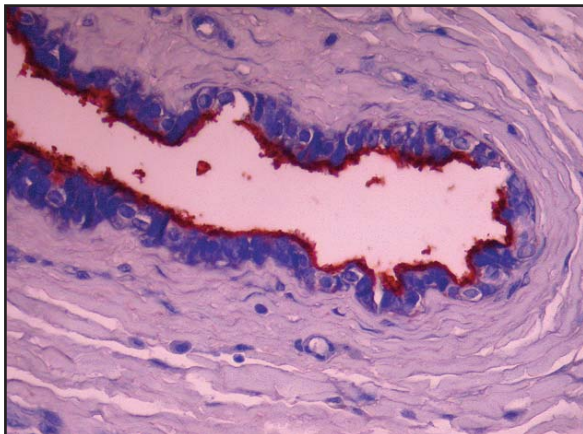
**Figure 1.** Cyst wall lined by tubal-type pseudostratified epithelium with cilia (HE, Original magnification is x40, inset x1000)



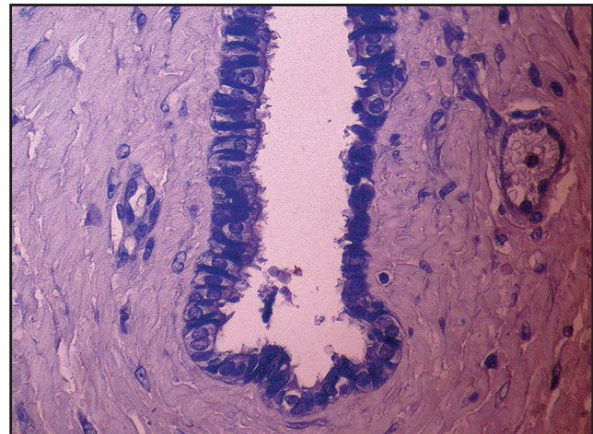
**Figure 2.** Positive nuclear immunostaining with PR (x400).



**Figure 3.** Positive nuclear immunostaining with ER (x400)



**Figure 4.** Positive immunostaining with EMA (x400)



**Figure 5.** No immunohistochemical CEA expression (x400)

### Case Report

A 12-year-old female presented with a mass in the left buttock skin. She recognised it in the past one week. On physical examination, it was non-tender and 2-3 cm in diameter. Her medical history was unremarkable except for an urinary infection and allergic symptoms. She had her first menstruation three months ago. The cyst was completely removed under local anaesthesia. Pathological examination revealed a uniloculated cyst containing colourless serous fluid with a diameter of 1 cm. The inner surface of the cyst was smooth. On microscopic examination the cyst was lined by cuboidal or pseudostratified columnar epithelium with cilia (**Figure 1**). It had a fibrous wall. There were no goblet cells, mucous glands, inflammatory infiltrate or smooth muscle. By immunohistochemistry, the epithelium showed positive staining with ER, PR and EMA (**Figure 2, 3, 4**). It was not

stained with CEA (**Figure 5**). The follow-up is uneventful for 2.5 years.

### Discussion

Cysts are usually classified on the basis of their pathogenesis. In the skin, most of them are derived from the dermal appendages as retention cysts. The developmental ones, which result from the persistence of vestigial remnants, are much less common [1].

The most common appendageal cysts are epidermal and trichilemmal cysts which are lined by squamous epithelium. The term 'cutaneous ciliated cyst' has been applied to the developmental cysts, but different names have been given according to their topographic localisation. Bronchogenic cysts, cutaneous ciliated cyst of the lower limbs, branchial, thyroglossal and thymic cysts, and cutaneous endosalpin-

giosis are included in this group [1]. They may appear anywhere in the course of the embryonic structures from the deep parts of the tissue up to the skin surface. They may be asymptomatic or may form subcutaneous masses.

All developmental cysts are uncommon. Cutaneous ciliated cyst is probably the least common one. There are less than 50 cases in the literature [2, 3, 4, 5, 6, 7, 8]. CCC was firstly described by Hess in 1890 [10] and later named by Farmer and Helwig in 1978 [8]. They are primarily observed in females in the second or third decade of the life. They are usually asymptomatic and present with an enlarging swelling cyst. They may occur anywhere but are primarily observed on the legs of females. Histologically, they are lined by cuboidal or pseudostratified epithelium. Mucinous cells are rare. The wall does not contain smooth muscle or glands. They may represent migratory Müllerian duct structures related to the fallopian tubes, the uterus, and the upper part of the vagina. The occurrence in males may be the result of vestigial Müllerian structures, or the cyst may be the result of a different genesis like ciliated metaplasia of eccrine glands [1]. Female hormones may play a role in stimulating the ciliated epithelium. Treatment is surgical excision of the lesion. If it is secondarily infected, antibiotics are needed.

Other cutaneous cysts are encountered in the differential diagnosis [1]. Bronchogenic cysts are lined by pseudostratified ciliated columnar epithelium with occasional goblet cells and collection of mucous glands and smooth muscle bundles in the wall. Branchial cleft cyst are lined mostly by stratified squamous epithelium, but deeper parts have a lining of ciliated columnar epithelium. A heavy lymphoid infiltrate invests the cyst, and mucinous glands and cartilage are occasionally present in the wall. Thyroglossal cysts are deep lesions in the midline of the neck. Thymic cysts are also deeply situated cysts. They are lined by one or more of squamous, columnar, cuboidal or pseudostratified columnar epithelia. The wall characteristically contains Hassal's corpuscles and there may be lymphoid tissue, cholesterol granulomas and sometimes parathyroid tissue. Ciliated cyst of the vulva is the most similar lesion with CCC and expresses ER and PR immunohistochemically. It is regarded as CCC by some authors [11, 12].

Cutaneous endosalpingiosis present as small unilocular cysts filled with granular material around the umbilicus following salpingectomy.

Some authors found the term 'cutaneous ciliated cyst' confusing and Hung et al. suggested the term 'cutaneous Müllerian cyst' for the ER/PR positive ones resembling simple fallopian tube epithelium, and 'cutaneous ciliated eccrine cyst' for the ER/PR negative lesions usually occurring in males and immunohistochemically compatible with an eccrine origin [2, 7].

Here, we reported a rare case of cutaneous ciliated cyst. It showed ER and PR expressions immunohistochemically, compatible with a possible Müllerian origin.

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