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PHOTOLETTER TO THE EDITOR

# Subcutaneous ciliated Mullerian cyst

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## Abstract

Cutaneous ciliated cysts are benign lesions occurring primarily on the lower extremity of girls and young women. We present a case of a cutaneous ciliated Mullerian cyst arising in the lower leg of a 14-year-old girl, with brief discussion of etiology and diagnosis. This is a rare entity with approximately 50 cases in the literature. (*J Dermatol Case Rep*. 2015; 9(4): 116-117)

### Key words:

cutaneous ciliated cysts, Mullerian cysts, Mullerian heterotopias, Pax-8, nodule

Cutaneous ciliated cysts are benign lesions occurring primarily on the lower extremity of girls and young women. This is a rare entity with approximately 50 cases in the literature.

We present a case of a cutaneous ciliated Mullerian cyst arising in the lower leg of a 14-year-old female. A 14-year-old teenage girl presented with a right lower extremity subcutaneous cyst. Past medical history includes asthma and seizures. The unilocular cyst was clearly visible on examination, with bluish coloration and few overlying skin telangiectasias. It was fluctuant on palpation, and fixed to the overlying dermis. There were no signs of previous rupture or drainage. The cyst had developed over the course of a year and measured 2 cm in diameter at presentation. At surgical excision, the cyst was easily located within the subcutaneous fat, superficial to the deep fascia of the leg. It was well-circumscribed with a thin whitish capsule. On histology, the cyst lining showed a simple and pseudostratified cuboidal to columnar epithelial lining positive for estrogen receptor (ER) and Pax-8 immu-

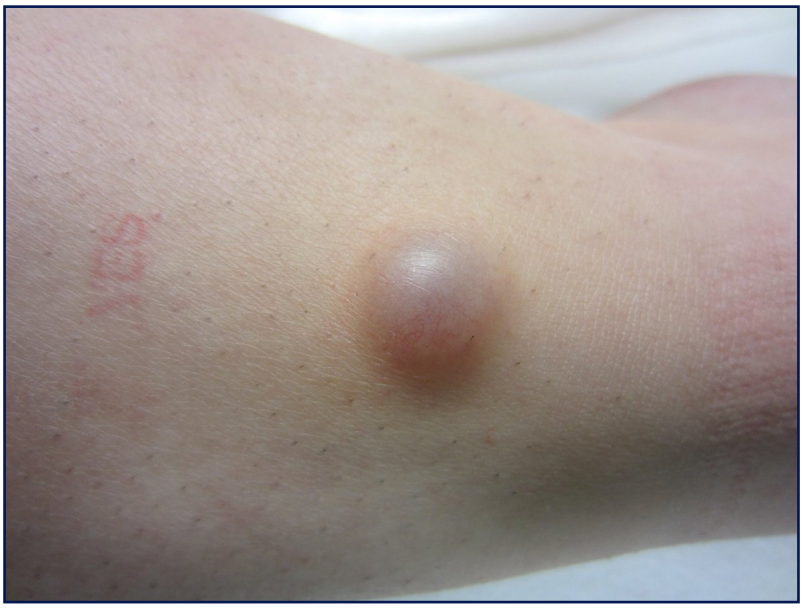

**Figure 1**

*Subcutaneous cyst of lower extremity, 2 cm.*

nohistochemical stains. A diagnosis of cutaneous ciliated Mullerian cyst was rendered. This is a rare entity with approximately 50 cases in the literature.[1,2]

Cutaneous ciliated cysts were termed so in 1978 by Farmer and Helwig in a series of 11 cases of lower extremity lesions in reproductive age women.[3] Since then, more benign cutaneous ciliated cysts in the dermis and subcutis have been described, but they are rare with scattered reports. Historically, the diagnostic nomenclature may have been vague or confused with inclusion cysts, lipomas, adnexal (eccrine) cysts, bronchogenic cysts, or pilonidal cysts. The cyst is lined by cuboidal to columnar simple and pseudostratified ciliated epithelium. The histology is uniform and bland; often reminiscent of fallopian epithelium.

The etiology has been explored recently utilizing immunohistochemistry. The cyst lining is positive for Pax-8, estrogen receptor (ER), progesterone receptor (PR), and Wilm's tumor gene-1 (WT-1).[1,2] Joehlin-Price *et al* suggested Mullerian differentiation in 2012, as Pax-8 expression is not typically found in skin or adnexal structures. Pax-8 is a paired box gene important in Mullerian development. However, the precise etiology may remain controversial to some authors.[1] In theory, migration of Mullerian rests occurs early in embryogenesis, and cysts later enlarge in response to hormonal development after puberty.[4,5] Examination of ultrastructure also corroborates a Mullerian origin.[4,5] There have been confounding reports of similar histologic findings arising in men and women with a supposed eccrine origin and ciliated metaplasia, but proper sampling and immunohistochemistry should render an appropriate diagnosis.[4,5] Distinction should be made from cutaneous ciliated eccrine cyst.[5]

In summary, cutaneous ciliated cysts are a rare entity primarily in the lower limbs of reproductive females, and should be reviewed with immunohistochemical studies for proper diagnosis.

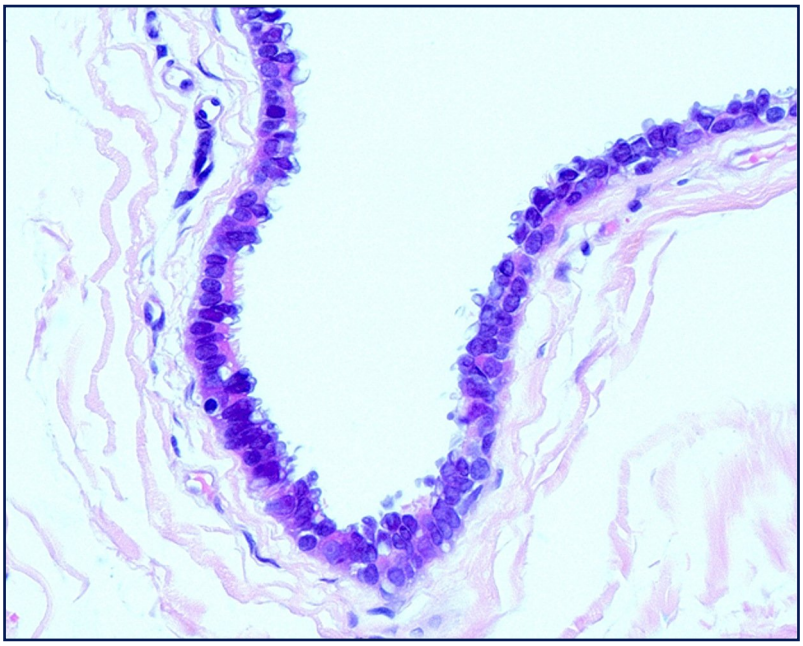

**Figure 2**

*Hematoxylin-Phloxine-Saffron stain, 400x magnification. Cyst lining showing ciliated cuboidal to columnar simple to pseudostratified epithelium.*

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