Cutaneous Ciliated Cyst

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Cutaneous ciliated cysts are rare benign lesions that typically appear on the lower extremities in young females, shortly after puberty.

We report a seventeen-year-old female who presented with a gradually increasing painless swelling over the anterior aspect of the right ankle. On examination, a non-tender 3.7x3.7 cm soft cystic swelling was found. Examination of the skin above the cyst showed no clinical finding. Ultrasound revealed a cystic lesion. Histopathological examination confirmed a rare cutaneous ciliated cyst.

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Cutaneous ciliated cysts are rare benign lesions typically found on the lower extremities in young females shortly after puberty. The morphological similarity between the lining cells and the fallopian tubes epithelium suggests their Müllerian origin[1-3].

The aim of this report is to present a rare case of cutaneous ciliated cyst on the ankle in a 17-year-old female.

THE CASE

A seventeen-year-old female presented with a gradually increasing painless mass in the anterior aspect of her right ankle. On examination, a 3.7x3.7 cm solitary, soft, non-tender, fluctuant mobile cystic lump was found. The overlying skin was unremarkable. The ankle joint ultrasound revealed a well-defined, subcutaneous lobulated cystic lesion measuring 3.7x1.1x3.7 cm with a clear volume of 7.2 ccs. Fine internal septa were seen but no calcifications or increased vascularity were noticed. The lesion was not attached to any nearby muscles or tendons. Clinical examination and ultrasound findings were suggestive of a ganglion cyst, hence, excised and sent for histopathological evaluation.

Gross examination revealed that the cystic lesion measured 25x25x15 mm, formed by a firm fibrofatty wall. The cut surface revealed a unilocular cavity filled with a clear serous fluid. The cyst wall was thin, smooth and grayish-white in color. Delicate and incomplete septa were also noted. Light microscopy confirmed the cystic nature of the lesion and the cyst was lined by a single layer of ciliated epithelial cells, the lining cells were cuboidal to columnar with scattered intraluminal papillary projections resembling that of a fallopian tube, see figures 1 and 2. The cyst wall was formed by vascularized fibro-collagenous tissue and no inflammatory cells were seen.

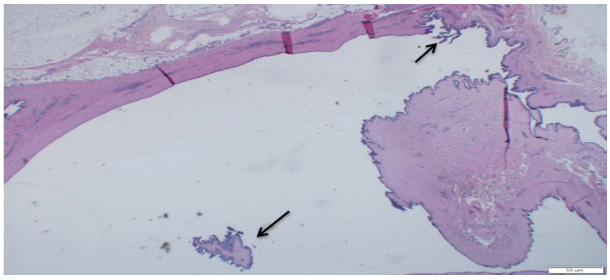

Figure 1: Unilocular Cyst with Incomplete Fibrous Septa and Some Papillary Projections (Arrow) [H&E, LPF]

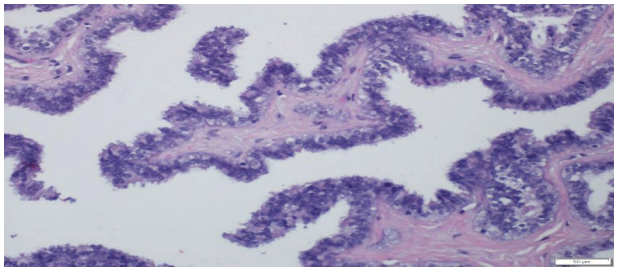

Figure 2: The Cyst is Lined by a Single Layer of Ciliated Cuboidal or Columnar Cells, Resembling that of a Fallopian Tube Lining [H&E, HPF]

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Immunohistochemical staining revealed strong epithelial nuclear positivity for estrogen (ER) and progesterone (PR), see figure 3. Pan-cytokeratin (Ckae1/ae3) was diffusely positive, while carcinoembryonic antigen (CEA) was negative.

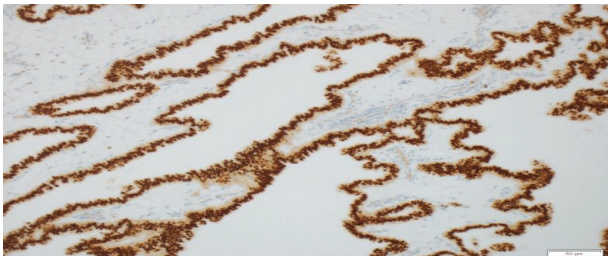

Figure 3: Strong, Diffuse Nuclear Positivity for Estrogen (ER) Antibody [IHC/ER, LPF]

The final diagnosis was a cutaneous ciliated cyst and the patient routine postoperative follow-up was uneventful.

DISCUSSION

Müllerian cysts are rare benign lesions, typically asymptomatic, presenting on the lower extremities in young females in their second or third decade of life. It is usually a simple cyst lined by a single layer of cuboidal to columnar ciliated epithelial cells[3]. Immunophenotypic findings correspond to the theory of heterotopia or heterotopic origin of the ciliated epithelium from the Müllerian epithelium[4].

Müllerian cyst was first described by Hess in 1890. Up to 2015, only 60 cases have been reported, of which 50 were females and 10 were males. While most reported cases occurred in the lower extremities and the surrounding region, other locations such as the back, neck, scalp, cheek and abdominal wall were reported[5].

Various theories were proposed; however, the origin remains debatable until today. These theories include Müllerian heterotopia, metaplasia of sweat gland and embryonic remnants. The evidence that supports heterotopic Müllerian origin concludes that Müllerian tissue was sequestered during embryonic development causing a hormone responsive Müllerian rests to form Müllerian-type cysts whenever there is an increased hormone production after puberty or during pregnancy[5-7].

In 1982, Leonforte studied the similarities and relationship of the cyst to the sweat glands, he described two features of apocrine sweat glands seen in these ciliated cysts: either the presence of PAS-positive granules or epithelial apical caps. Leonforte considers the epithelial change which appears similar to fallopian tube as metaplastic change due to chronic irritation of the pluripotent cells[8].

A similar case of a cutaneous ciliated cyst of the perineum from a 60-year-old male has been reported by Sidoni et al, in which they proposed that the cyst origin was from embryonic remnants of the cloacal membrane, especially the primitive caudal gut[9].

Regardless of the different suggested theories, the Müllerian heterotopia theory remains the most appealing due to the morphological similarities to the fallopian tube epithelium and the presence of nuclear positivity for estrogen and progesterone[5,7].

Other differential diagnoses should be considered: branchial cleft cyst, teratoma, bronchogenic cyst and thyroglossal cyst[10,11]. However, the location of the lesion in the lower extremity and the absence of mucous gland, together with the immunohistochemical positivity for ER, PR and negative CEA confirms the diagnosis of cutaneous ciliated cyst.

The recommended treatment for this cystic lesion is surgical removal for which recurrence after surgery has not been reported in the literature[12,13].

CONCLUSION

Despite its rarity, ciliated cyst should be included in the deferential diagnosis of a leg cyst in a young female.

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