Case Report
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A Remarkably Rare Position of a Cutaneous Ciliated Cyst in a 16 Month-old Female: A Case Report.

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Abstract
Objective. The aim of the work was to show a Cutaneous Ciliated Cyst (CCC) in an unusual location in a 16-month-old girl. **Case Report.** We present the case of a 16-month otherwise healthy girl presented to our hospital, with a report of a palpable mass in the left suprascapular region. Physical examination revealed a soft-textured, fluctuating, mobile and painless entity, with no further indications of local inflammation. The mass was totally excised, under general anesthesia, for both diagnostic and therapeutic purposes. According to the histopathological findings, the cystic lesion was covered by a pseudostratified ciliary epithelium, resembling the epithelium of a normal fallopian tube, surrounded by a smooth muscle layer. Immunohistochemical studies identified the cyst epithelium as having cytokeratin (CKAE1/AE3) expression, despite the negative immunostaining findings on Estrogen and Progesterone Receptors. **Conclusion.** Our case report concerns a CCC in an unusual position, in the suprascapular area. After a thorough review of the international literature, we concluded that this is the second published case regarding this specific location. To our knowledge our patient is the youngest ever diagnosed with CCC.

Key Words: Cutaneous Mullerian Cyst ▪ Unusual Position ▪ Paediatric Patient ▪ Müllerian Duct Remnants.

Introduction

The entity was first described by Hess, in 1890 and the term Cutaneous ciliated cyst (CCC) was established by Farmer and Helwig, describing a unique type of cyst arising on the lower extremities of young women (1). CCCs, also known as cutaneous Müllerian cysts, are cystic lesions, mostly found on the lower extremities of females. They are covered by a pseudostratified ciliary epithelium, resembling that of fallopian tubes. Usually the wrong impression is established of them being epidermoid or dermoid cysts.

We present a case of a CCC located in the suprascapular area. The unusual location, together with the young age of our patient were the stimuli for writing this study. There is no case of CCC described in the literature concerning such a young patient.

Case Presentation

A 16-month-old girl was referred to our Paediatric Surgery Clinic due to a painless lesion in her left suprascapular region. This growth was detected during the first month of her life. From then, it had remained asymptomatic and stable in its dimensions. There was no history of previous trauma or other remarkable medical problems. Clinical examination revealed an almond-sized mass, which was mobile, fluctuating, and had a soft texture on palpation. No macroscopic signs of local inflammation were detected, while there was no attachment to the subcutaneous fat. Ultrasonography showed a well-demarcated round mass, measuring 1.3 cm × 0.8 cm, with internal hyperechogenic regions.The formation was totally excised under general anesthesia (Figure 1).

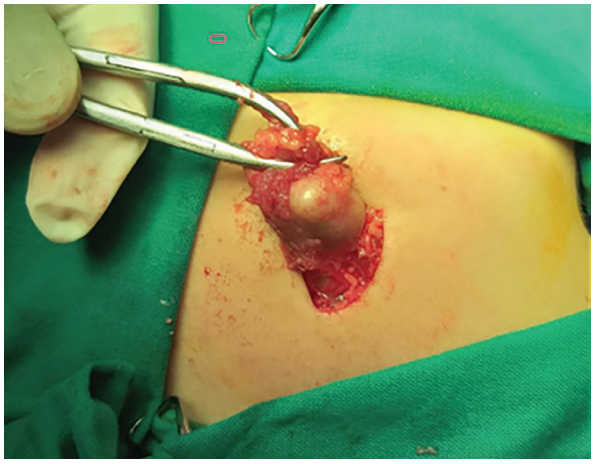

Figure 1. The gross appearance of the lesion, indicating the excised unilocular cyst.

The patient had an uneventful postoperative course and left our hospital the next morning. Microscopic investigation of the excised specimen identified a pseudostratified cilia columnar epithelium, clearly resembling a fallopian tube. A surrounding smooth muscle layer was also detected (Figure 2).

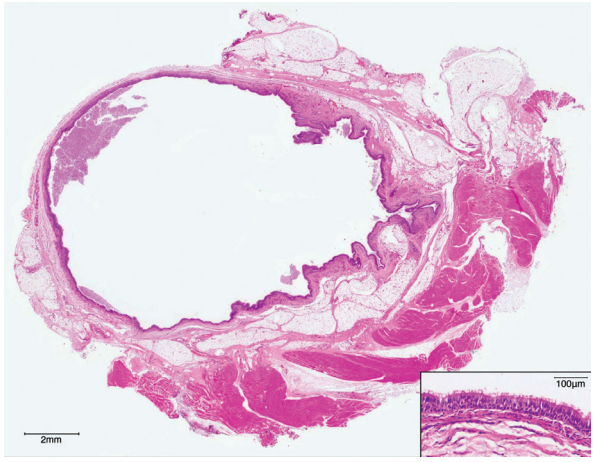

Figure 2. The pseudostratified columnar ciliary epithelium resembling the tubal epithelium (H&E stain). *Detail: The cilias are depicted on the surface of the cells.

The cytoplasmic membrane of the cyst had a positive reaction to Cytokeratin immunocytochemistry (CKAE1/AE3), but intranuclear staining for Estrogen and Progesterone receptors was not shown. The young patient remained asymptomatic during the one-year follow-up.

Discussion

A cutaneous Mullerian cyst is an extremely rare, benign entity, frequently located on the lower extremities of young women after puberty, during the second and third decade of life (1, 2). However, there are some published cases worldwide concerning men (2, 3). Different sites of location have also been reported, including the abdominal wall, the inguinal region, the umbilicus and the scalp (3). As far as our case is concerned, this formation was located in the patient's suprascapular area, which is an extremely rare position, as it is only the second case published worldwide in the literature, concerning such a young pediatric patient (3). These entities usually appear in a specific age group of the population. It is crucial to mention that our patient does not belong to this specific category (2, 3). Fabien-Dupuis et al. reviewed 60 patients with CCC, from 1890 to 2015. In only one 15-year-old patient (1/60) was the cutaneous Mullerian cyst located between the two scapular regions (1). Consequently, our case report is the second one referring to that uncommon location.

In the recent review by Yon Hee Kim et al. 31 patients were recorded, including both sexes, using the PubMed search. The youngest was 7 years old (4). This statement demonstrates that our 16-month girl is the youngest to be diagnosed with CCC described in the literature. CCCs immunohistochemically most often have identical epithelium to fallopian tubes, as they are covered by a cuboid or columnar pseudostratified epithelium (5). To the best of our knowledge, all case reports, published in the literature concerning women, have ER and PR positivity for nuclear cells (2). What makes our case special, is its negativity for these immunostains. The differential diagnosis for CCC also includes an unusual cystic entity known as the Bronchogenic Cyst (BC). The main difference lies in the thoracic location of the BC, apart from its predominance in male patients. The presence of smooth muscle, seromucous glands and, rarely, cartilage in the cystic wall are the main characteristics of a BC during histopathological examination (6). Concerning our patient, the data

excluded the diagnosis of BC. The CCC's epithelium had an obvious resemblance to fallopian tubes. Their nuclear positivity for antibodies of ER and PR, together with the epithelium, led to the hypothesis of Müllerian heterotopia (2). On the basis of that theory, the cysts derive from remnant cells of the paramesonephric duct during the embryological period, and especially between the 6th and 7th week of gestation (3). Cells that are not capable of being integrated with the mesoderm may migrate to the lower extremities.

Consequently, during menarche hormonal stimulation plays a role in the growth of those cystic formations (2-5). A second theory, concerning the etiological background of this entity, supports the cilia metaplasia of the eccrine glands. This concept was developed due to the published cases of CCCs appearing in men. Apart from that, more cases with ER and PR negativity for nuclear cells have been published over time, meaning the Müllerian duct hypothesis is void (3, 7). Finally, therefore, we believe that cutaneous ciliated cysts should be divided into two subgroups: cutaneous Müllerian cysts and ciliated cutaneous eccrine cysts. The cases presenting with ER and PR positivity belong to the first subgroup, in contrast to those that do not have positive immunostaining findings for Estrogen and Progesterone receptors. The first subgroup includes the majority of cases, specifically involving young women between the ages of 15 and 30 years. The cystic formations are usually located on the lower extremities, and their nuclear cells meet the above mentioned positivity for ER and PR. On the other hand, cases of uncommon locations, and also those found in male patients with ER/PR negativity, belong to the second subgroup of cutaneous ciliated cysts (2, 5, 7).

Conclusion

Consequently, our case refers to an eccrine cyst found in an exceptionally rare position. Our patient is the youngest reported in the literature. Surgical resection of the cystic mass is the only therapeutic solution. There was no technical difficulty in removing this entity (2, 3).

What Is Already Known on This Topic:

Cutaneous Ciliated Cysts(CCCs) are rare, cystic entities usually located on the lower extremities of young reproductive female adults. CCCs may also be found on the abdominal wall, in the inguinal region, the umbilicus and the scalp. The etiology is not fully known, but there are two dominant theories. The first is that they are ectopic Mullerian residues, while the second theory supports the ciliated metaplasia of eccrine glands.

What This Case Adds:

This case report presents an uncommon location of a Cutaneous Ciliated Cyst found in the suprascapular area of a 16-month girl. After a thorough review of the literature using Pubmed search, we found that our patient is the youngest presented from 1890 up to the beginning of this year. So, our case is novel, because the patient does not belong to the usual age group, and also in terms of the rare location.

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Conflict of Interest: The authors declare that they have no conflict of interest.

References

1. Fabien-Dupuis C, Cooper B, Upperman J, Zhou S, Shillingford N. Mullerian-Type Ciliated Cyst of the Thigh with PAX-8 and WT1 Positivity: A Case Report and Review of the Literature. Case Rep Med. 2016;2016:2487820.
2. Kim Y, Kim H. The Cutaneous Ciliated Cyst in Young Male: The Possibility of Ciliated Cutaneous Eccrine Cyst. Case Rep Med. 2015;2015:589831.
3. Doğan G, İpek H, Metin M, Özkayar Ö, Afşarlar ÇE. Cutaneous Ciliated Cyst in an Unusual Location: Between Two Scapulas. Case Rep Surg. 2018;2018:5961913.
4. Kim YH, Lee J. Cutaneous ciliated cyst on the anterior neck in young women: A case report. World J Clin Cases. 2020;8(19):4481-7.
5. Hung T, Yang A, Binder S, Barnhill R. Cutaneous ciliated cyst on the finger: a cutaneous mullerian cyst. Am J Dermatopathol. 2012;34(3):335-8.
6. Dini M, Lo Russo G, Baroni G, Colafranceschi M. Cutaneous ciliated cyst: a case report with immunohistochemical evidence for dynein in ciliated cells. Am J Dermatopathol. 2000;22(6):519-23. doi: 10.1097/00000372-200012000-00007.
7. Fontaine DG, Lau H, Murray SK, Fraser RB, Wright JR Jr. Cutaneous ciliated cyst of the abdominal wall: a case report with a review of the literature and discussion of pathogenesis. Am J Dermatopathol. 2002;24(1):63-6.