CASE REPORT

CILIATED VULVAR CYST IN A 13-YEAR-OLD GIRL- CASE REPORT

Aneta Sima[1], Vesna Naunova[2], Slagana Simeonova Krstevska[1], Drage Dabeski[1], Lazo Jovcheski[2], Toni Risteski[2], Blagica Krsteska[3]

[1] *University Clinic for Gynecology and Obstetrics; Faculty of Medicine, Ss. Cyril and Methodius University in Skopje, North Macedonia*

[2] *University Clinic for Pediatric Surgery; Faculty of Medicine, Ss. Cyril and Methodius University in Skopje, North Macedonia*

[3] *Institute of Pathology, Faculty of Medicine, Ss. Cyril and Methodius University in Skopje, North Macedonia*

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***Correspondence:** Aneta Sima, University Clinic for Gynecology and Obstetrics, Skopje; Republic of North Macedonia

E-mail: aneta_sima@yahoo.com

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Abstract

Vulvar cysts are rare, mostly benign lesions in childhood, originating from Miller's embryonic tissue and associated with progesterone action. The interior of the cysts is lined with columnar epithelium with cilia. They can be asymptomatic until they reach a size that causes heviness and pressure on the surrounding tissue and aesthetic deformity of the vulva. Case report: A 13-year-old girl presented with a large elongated cystic structure originating from the right labia majora of the vulva, with a length of about 20 cm, without significant symptoms, except for discomfort in the external genitalia. The cyst was surgically removed and the histopathological findings confirmed the existence of a ciliary vulvar cyst, originating from Miller's structures. After the operation, the cosmetic result was excellent and the young patient felt relieved. Conclusion: Vulvar cysts are a rare finding in childhood and puberty, but they can also reach large sizes. The treatment is surgical excision, which provides a cure, correction of the normal anatomy of the vulva, and histological confirmation of the diagnosis.

ПРИКАЗ НА СЛУЧАЈ

ЦИЛИЈАРНА ЦИСТА НА ВУЛВА КАЈ 13- ГОДИШНО ДЕВОЈЧЕ

Анета Сима[1], Весна Наунова[2], Слаѓана Симеонова- Крстевска[1], Драге Дабески[1], Лазо Јовчески[2], Тони Ристески[2], Благица Крстеска[3]

[1] *Универзитетска клиника за гинекологија и акушерство; Медицински факултет, Универзитет „Св. Кирил и Методиј" во Скопје, Република Северна Македонија*

[2] *Универзитетска клиника за детска хирургија; Медицински факултет, Универзитет „Св. Кирил и Методиј" во Скопје, Република Северна Македонија*

[3] *Институт за патологија; Медицински факултет, Универзитет „Св. Кирил и Методиј" во Скопје, Република Северна Македонија*

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Клучни зборови: цилијарни цисти, Милерови структури, вулва

***Кореспонденција:** Анета Сима, Универзитетска клиника за гинекологија и акушерство, Скопје, Република Северна Македонија.

E-mail: aneta_sima@yahoo.com

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Печатарски права: ©2024. Анета Сима, Весна Наунова, Слаѓана Симеонова- Крстевска, Драге Дабески, Лазо Јовчески, Тони Ристески, Благица Крстеска. Оваа статија е со отворен пристап дистрибуирана под условите на нелокализирана лиценца, која овозможува неограничена употреба, дистрибуција и репродукција на било кој медиум, доколку се цитираа торигиналниот(ите) автор(и) и изворот.

Конкурентски интереси: Авторот изјавува дека нема конкурентски интереси.

Извадок

Вулварните цисти се ретки, најчесто бенигни промени во детската возраст, со потекло од Милеровите ембрионални структури и се поврзани со дејството на прогестерон. Внатрешноста на цистите е обложена со колумнарен епител со трепки. Можат да бидат асимптоматски сé додека не постигнат големина за да предизвикаат тежина и притисок врз околното ткиво, како и естетски деформитет на вулвата. Приказ на случај: Тринаесетгодишно девојче дојде на преглед со голема цистична структура која потекнуваше од десната голема усна на вулвата, со должина од околу 20 см, без значајни симптоми, освен дискомфорт во надворешни гениталии. По сооветна подготовка, цистата беше хируршки отстранета- хистопатолошкиот наод потврди постоење на цилијарна вулварна циста, со потекло од Милеровите структури. Постоперативниот тек помина уредно, со одличен козметички резултат и олеснување за младата пациентка. Заклучок: Вулварните цисти се редок наод во детска возраст и пубертет, но можат да достигнат и големи димензии. Третманот се состои во хируршка ексцизија со која се постигнува лекување, корекција на нормалната анатомија на вулвата, како и хистолошка потврда на дијагнозата.

Introduction

Vulvar cysts are a unique entity in pediatric gynecology with an often confusing clinical appearance. Although relatively rare in pediatric population, they can have significant implications on the health and well-being of young patients and therefore require special attention due to their anatomical and physiological characteristics[1,2]. The exact etiology of the vulvar cysts with cilia remains to be further investigated. Current research is focused on the most probable origin from epithelial remnants of the Mullerian ducts, the embryonic precursors of the female reproductive tract. However, it is widely believed that these result from incomplete involution or fusion, abnormalities of Mullerian ducts during embryogenesis. This incomplete regression can lead to the formation of cystic structures, typically located in the vulvar region[3,4]. Furthermore, hormonal influences can also play a role in developing vulvar ciliary cysts. Changes in hormone levels, especially estrogen, can affect the growth and maintenance of these cystic structures. This is especially relevant in adolescent patients, where fluctuations in hormonal levels are common[5,6].

Case report

We describe the case of a 13-year-old patient who presented with a large elongated cystic formation localized on the right labia majora(Figure1), 20cm long and filled with liquid content. The cystic formation did not fluctuate in size and was connected to the skin of the labia majora with a narrow stalk. According to the patient's history, the cystic formation appeared 3 years ago and gradually grew without causing any particular symptoms. However, she complained that it became so large that she felt uncomfortable wearing jeans and a bathing suit in public. She was afraid and shy, so she didn't share any information with her family and physician, at least not until the cyst grew significantly. She had her first menarche at the age of 12 years, and she has a regular menstrual cycle. On clinical examination, the inguinal region was free without signs of hernia and the hymen was intact. Surgical excision was indicated and she was scheduled for elective removal of the lesion.

The patient underwent surgery with general anaesthesia. She was placed in a modified frog-leg position. The excision boundaries were marked and the entire cyst was excised at the level of the stalk-skin attachment. Right labial skin edges were sutured with continuous interlocking suture with monofilament, absorbable suture (Figure 2). The patient tolerated the procedure well and there were no complications.

The entire cyst was sent for histopathologic examination After complete processing of the pathohistological and immunohistochemical examinations, the cut section of the specimen revealed a cystic space filled with serous fluid (Figure 4). The wall of the cyst was lined by one layer of pseudostratified ciliated epithelium with focal squamous metaplasia. There was no significant inflammation in the cyst wall.

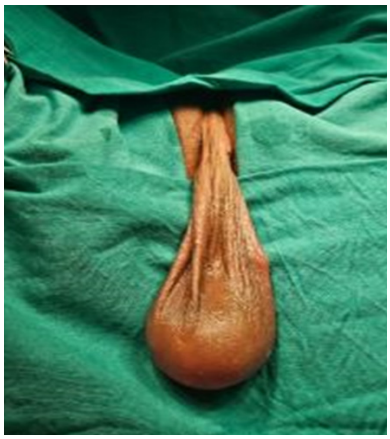


Figure 1. Before operation

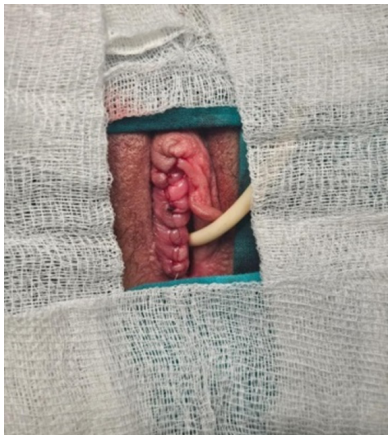


Figure 2. Post-operation

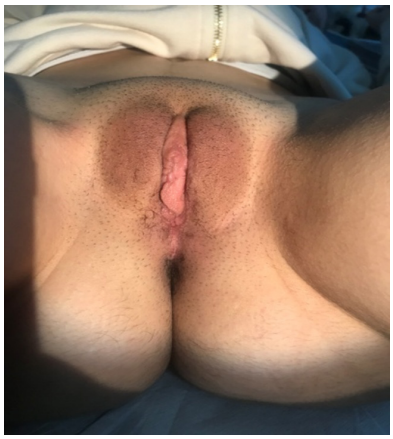
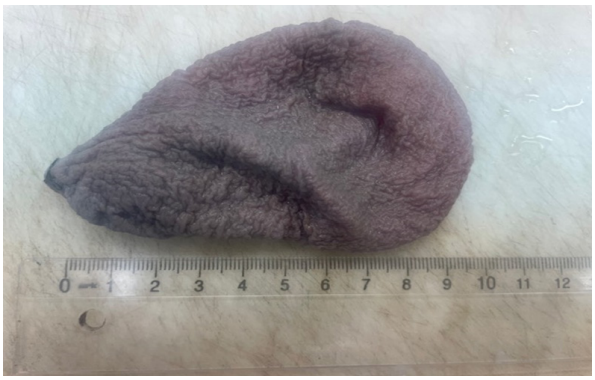


Figure 3. One month after



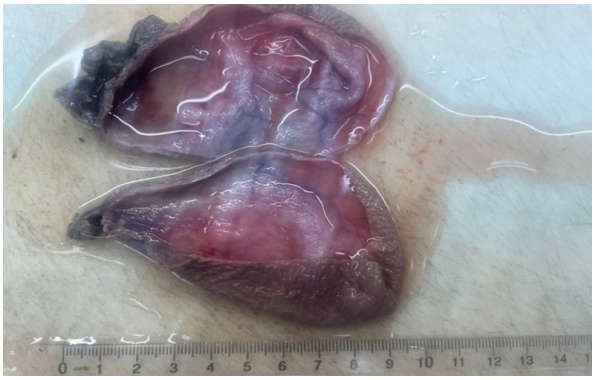
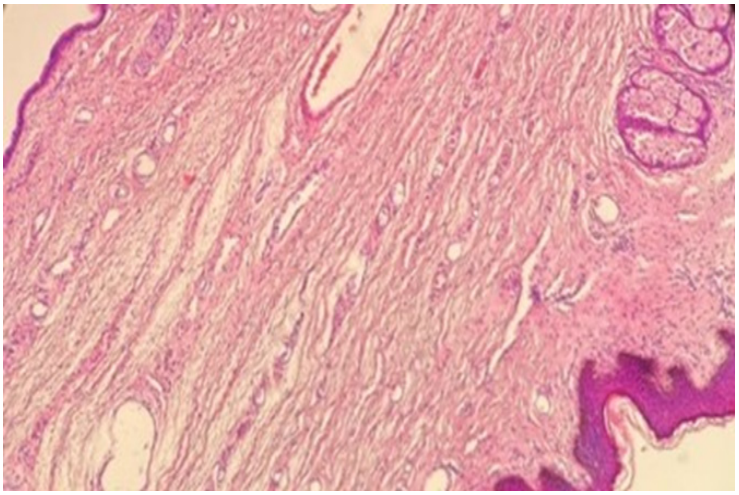


Figure 4. The cut section of the specimen revealed cystic space filled with serous fluid

Teratoma elements were not identified. Immunohistochemical analysis showed that the ciliated cells were positive for estrogen receptor (ER), progesterone receptor (PR), cytokeratin 7 (CK7), PAX8 and WT1 which confirmed the Mullerian origin (Figure 5).



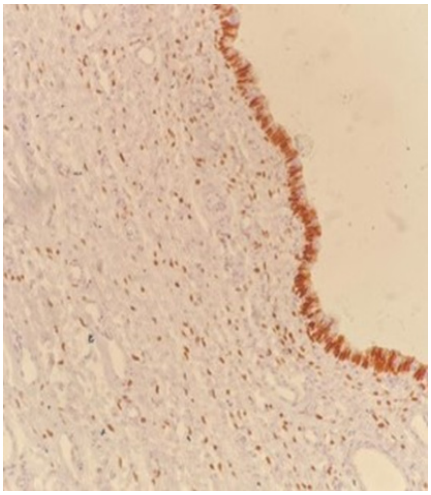


Figure 5. A benign Müllerian cyst lined with ciliated epithelium with ER positivity

No complications were observed postoperatively, and at the 4-week follow-up visit, the patient was in a good clinical condition (Figure 3). She was very satisfied with the esthetic outcome and was psychologically relieved.

Discussion

The clinical presentation of vulvar cysts can vary greatly, which often leads to challenges in the diagnosis. Various pathohistological entities can present clinically with a similar appearance as a vulvar "polypoid cystic lesion". These cysts may be asymptomatic and discovered incidentally during routine physical examinations or tests for other conditions. However, when they are symptomatic, they can manifest in several ways: vulvar mass or swelling, pain or discomfort, dyspareunia, localized redness or inflammation, urinary symptoms and recurrent infections[7]. Diagnosis can be confirmed by physical examination of the vulvar region as the first step in the evaluation of a suspected vulvar ciliary cyst. Palpation is necessary to assess the size, location and consistency of the cystic mass[8]. Transabdominal or transperineal ultrasound is often the modality of choice for evaluating cystic lesions of the vulva. The ultrasound can provide valuable information about the size, location, contents and characteristics of the cyst, helping to differentiate it from other vulvar masses. In the cases where ultrasound findings are indistinct or require additional characterization, magnetic resonance imaging can offer detailed imaging of the cystic lesion and its relationship with the surrounding structures. A biopsy of the cystic lesion is rarely performed, although in some cases it can be necessary. Histopathological examination after complete surgical excision is a gold standard in the diagnosis[4,6].

Differential diagnosis- considering the varied clinical presentation of vulvar ciliary cysts, it is essential to consider several differential diagnoses when evaluating a pediatric patient with a vulvar mass8. The following are some situations that can imitate this situation: cyst or abscess of the Bartholin's gland, hemangioma or vascular malformations, epidermal inclusion cyst, lipoma, sebaceous cyst, inguinal hernia(in pediatric patients, an inguinal hernia can manifest as a bulge or swelling in the groin or labial region). This condition requires rapid evaluation and surgical intervention if symptomatic[5,6,9].

Conclusion

Comprehensive clinical evaluation and wisely chosen imaging techniques can help in the differentiation of vulvar cysts. Histopathological examination is the most preferred way of distinguishing vulvar ciliary cysts from other vulvar masses in pediatric patients. Therefore surgical excision is the preferred approach to treatment, it is curative while also providing significant restoration of the appearance of external genitalia.

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