What is your diagnosis?

A 30-year-old nulligravida woman presented to the gynaecology outpatient department, with the chief complaint of progressive, painless swelling in the perineal region for the last three months. The swelling was initially asymptomatic, but for the previous 15 days, it had started to protrude out on straining and caused difficulty during urination and defecation. There was no associated pain or fever. Her menstrual cycles were regular, and she had no dysmenorrhea, dyspareunia or chronic pelvic pain. On local examination, there was a boggy, non-tender swelling of 3x2 cm just inside the introitus, on the left side, located in the subcutaneous tissue of the labia majora, at the junction of upper two-thirds and lower one-third. The patient also complained of a similar swelling at the same site six months earlier, which was tense, painful and associated with fever. At that time, it ruptured spontaneously with the discharge of foul-smelling purulent fluid. A clinical diagnosis of Bartholin cyst was made, and the cyst was completely excised. Of note, the cyst had a thick wall and was densely adherent to the underlying structures. The bed was very vascular, leading to a blood loss of approximately 250 mL during excision, which was quite unusual for a Bartholin cyst. The post-operative period was uneventful. A one-month follow-up visit showed complete healing of the surgical scar, and the patient was symptom-free.

Received: 05 January, 2023 **Accepted:** 17 March, 2023

Answer

Grossly, the cyst measured 3x2.5x2.5 cm, with wall thickness varying from 3-5 millimetres. The cavity was filled with solidified brownish material (Figure 1a). Microscopic evaluation confirmed the diagnosis of Bartholin cyst. The cyst wall was composed of fibro-muscular tissue lined by inflamed transitional epithelium. The normal Bartholin gland was seen in juxtaposition to the cyst wall (Figure 1b-d). However, in a few areas, the lining epithelium showed the presence of ciliated cells (Figure 1e,f), which is an extremely unusual finding and has been documented only once previously (1). Interestingly, the ciliated epithelium was negative for Periodic acid-Schiff (PAS) stain.

In contrast, the non-ciliated epithelium displayed a patchy magenta-coloured PAS-positive reaction, similar to the native glandular epithelium (Figure 1g-i). Multiple areas of the cyst wall were processed because of the resemblance of cavity contents to that of a chocolate cyst, which showed similar histo-morphology. There was no evidence of endometriotic cysts, such as endometrial gland or stroma, hemosiderin-laden macrophages or areas of haemorrhage.

Non-neoplastic cysts of the vulva and vagina can usually be easily diagnosed based upon the location, clinical features and subsequent histopathological characterisation of the lining epithelium (Table 1) (2,3). Although the literature regarding ciliated cysts is sparse, they are thought to arise due to Mullerian heterotopy, that is a Mullerian tissue that has been displaced from its original position during embryogenesis. These cysts are usually an incidental finding, but rarely can present as large masses distorting the normal anatomy. As the Mullerian tissue is sensitive to hormones, these cysts have a tendency to increase in size during pregnancy or due to exogenous hormone administration (2). They constitute a tight differential diagnosis in the present case. Clinically, a ciliated cyst can present as a cystic swelling in the postero-lateral aspect of the vulva (the specific location of a Bartholin cyst), and histopathology can also show ciliated lining epithelium (2,3). In the present case, a diagnosis of Bartholin cyst with ciliated/tubal metaplasia was favoured because of the presence of Bartholin glands and smooth muscle fibres in the cyst wall. The ciliated cyst of the vulva is described as being lined by ciliated, mucinous or non-mucinous columnar cells without smooth muscle cells in the cyst wall (4,5).

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DOI: 10.4274/jtgga.galenos.2023.2022-12-7

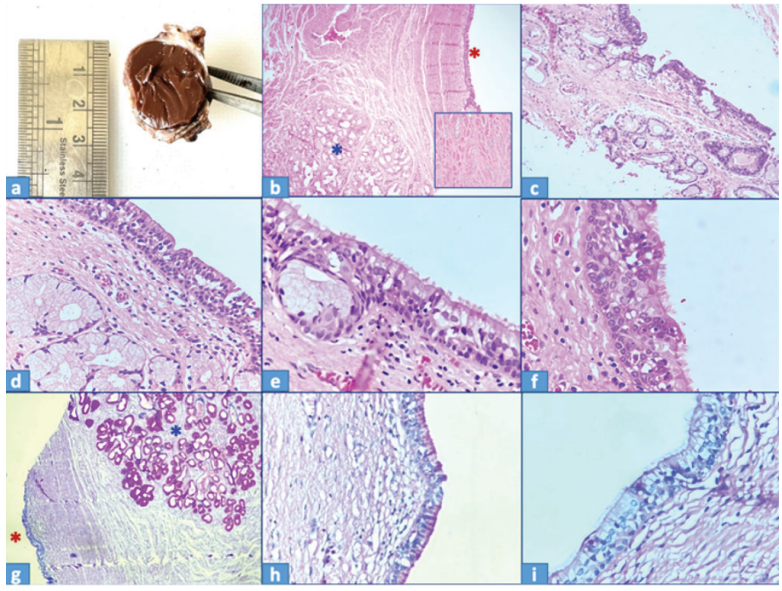

Figure 1. (a) Gross picture of the cyst showing solidified, chocolate-brown material in the cavity; (b) Cyst wall composed of fibromuscular tissue (inset) and native Bartholin gland (blue asterisk) and lined by transitional epithelium (red asterisk) [hematoxylin & eosin (H&E), x40]; (c) The lining epithelium of the cyst shows connection with the underlying glandular tissue (H&E, x100); (d, e) At places, the lining epithelium was pseudostratified to multi-layered with the presence of cilia (H&E, x400); g) Intense Periodic acid-Schiff (PAS)-positive reaction noted in the native Bartholin gland (blue asterisk) but not in the lining epithelium (red asterisk) (PAS, x40); h) Patchy PAS-positive reaction noted in the transitional epithelium (PAS, x100); i) The ciliated epithelium is PAS-negative (PAS, x400)

Table 1. Diagnostic key points of non-neoplastic cystic lesions of vulvovaginal region (2,3)

S. no.	Entity	Usual location	Histopathology
1	Bartholin cyst	Posterolateral to the opening of vagina	- Cyst lining may be stratified squamous, transitional, mucinous (PAS, AB, mucicarmine positive) or flattened - Fibromuscular wall
2	Gartner's cyst (mesonephric like cyst/ Wolffian duct like cyst)	Anterolateral wall of vagina lateral aspects of the vulva	- Lined by non-mucinous, non-ciliated cuboidal cells - Thin fibromuscular wall - Usually contains clear fluid
3	Epithelial inclusion cyst	Frequently seen on vulva as superficial lesions of varied dimensions	- Stratified squamous epithelial lining - Cavity contains keratinous material - May be associated with foreign-body reaction to keratin.
4	Mucous cysts	Usually are seen within the vestibule	- Mucus-secreting (PAS, AB, mucicarmine positive), cubo-columnar epithelial lining - Squamous metaplasia may be noted - No smooth muscle fibres in cyst wall
5	Ciliated cyst (Mullerian-type cyst)	Can be seen anywhere within vagina and vulva, although more common in the anterolateral wall	- The lining epithelium can be endocervical type (mucinous, columnar) or endometrial type (non-mucinous, columnar) or tubal type (ciliated, columnar) - No smooth muscle fibres in cyst wall
6	Cyst of canal of nuck (mesothelial cyst)	Found in inguinal canal or superior aspect of labia majora	- Lined by a single layer of flattened mesothelial cells
7	Endometriotic cyst	Usually seen in association with pelvic disease and can be seen anywhere in the vulvovaginal area.	- Any 2 of the following 3 findings: endometrial glands, stroma and hemosiderin laden macrophages.

PAS: Periodic acid-Schiff; AB: Alcian blue

The present case highlights that ciliated epithelium or tubal metaplasia can also be encountered in a Bartholin cyst. A definitive diagnosis can easily be achieved by clinical correlation, finding the acini of the Bartholin gland in the cyst wall and the presence of smooth muscle fibres. As various metaplasias are commonly encountered in the female genital tract, it is quite plausible that ciliated/tubal metaplasia in a Bartholin cyst is an under-reported entity rather than a very uncommon entity.

The authors certify that an appropriate patient consent form has been obtained.

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