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Case Report
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Uterine Mullerian Cyst in a Pregnant Woman

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Objective

We describe a rare case of a pregnant woman with mullerian cyst of the uterus.

Case Report

A 27-year-old woman pregnant lady was found to have a cystic adnexal mass at anomaly scan. This was suspected to be a left sided ovarian cyst. This was her second pregnancy and she had an emergency caesarean section which was reported to be uncomplicated. Her pregnancy remained uneventful and she opted for a caesarean section at term as she did not want trial of scar. An informed consent was obtained to perform ovarian cystectomy at caesarean section.

At caesarean section, minimal adhesions were encountered and baby was delivered as per routine. After closure of uterine incision, we explored the ovaries and tubes which were normal. However, a large benign looking cystic mass was seen arising from posterior uterine wall (Figure 1).

The mass was 9 cm × 10 cm in size with two loculi separated by a thin septum containing clear fluid. The cystic mass was resected and histological examination revealed a benign mullerian cyst of uterus lined with mucinous epithelium (Figure 2).

No complications were observed postoperatively and she was discharged home in stable condition. The patient had a follow up pelvic ultrasound scan after 3 months which was normal.

Discussion

Mullerian cysts are rare neoformations of paramesonephric duct, with peak incidence between the third and fourth decades of life1. Prevalence is still unclear but estimated to be 1% to 5% [1-3]. These cysts are generally small, midline, cystic masses which are asymptomatic and therefore, do not need to be treated. Sometimes these cysts become large and warrant surgical removal. Uterine cyst of mullerian origin are even rare with very few cases reported in literature. Pathogenesis is unclear but few theories have been proposed to justify their origin from peritoneal mesothelial cells or from primary mullerian epithelium [4]. Yang et al. described a case of 31-year-old woman with endosalpingiosis who was the youngest of all reported cases [4]. Our patient was 27 years and is therefore the youngest of all cases of Mullerian uterine cysts reported so far.

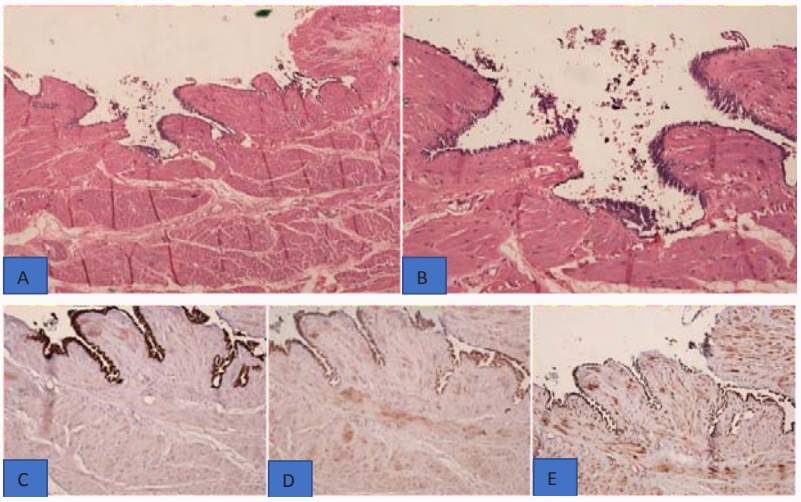

Figure 1: Hematoxylin and Eosinophil (H&E) stained sections of cyst lined by bland cuboidal epithelium and surrounded by myometrium (A & B). Immunostains CK-7 (C), WT-1 (D), and PAX-8 (E) highlighting the lining epithelium.

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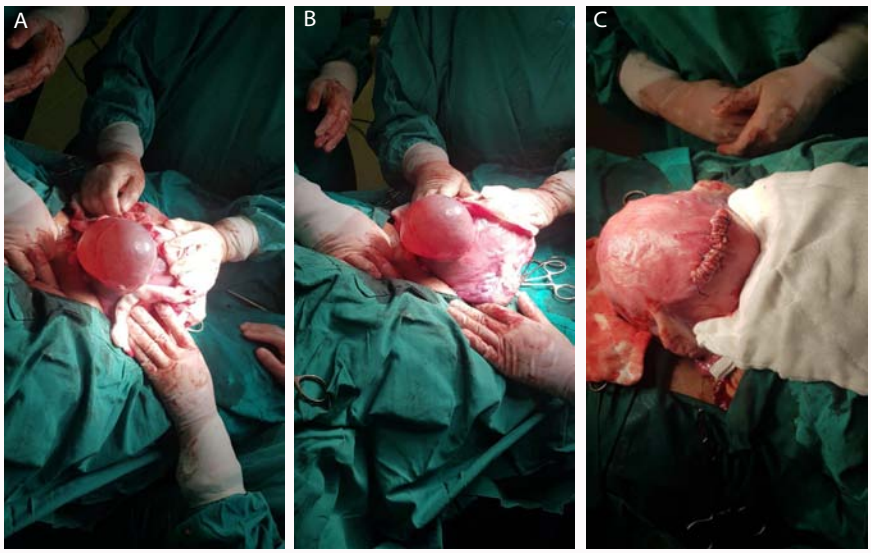

Figure 2: Histological examination.

It is important to differentiate mullerian cysts from ovarian cysts before surgical intervention. However, definitive diagnosis of mullerian cyst is challenging without histopathological assessment [5]. Ultrasound is the first line investigation tool, however, MRI provides excellent visualization and resolution of the cyst and surrounding structures so it should be carried out where expertise and facilities are available.

There are few case reports where patient was operated as a case of ovarian cyst but surgical assessment and histopathology confirmed diagnosis of mullerian cyst [6,7]. Datti et al. performed laparotomy for huge ovarian cyst that proved to be mullerian cyst on histopathology [8]. Literature reports three cases of retroperitoneal cysts of mullerian origin that presented initially as pelvic masses [8,9]. Rashmi and Montella et al. reported a case of enterocele and anterior vaginal wall cyst respectively that turned out to be Mullerian cyst [10,11]. Similarly, Lui and Nakae et al. reported two cases of pelvic/adnexal masses in perimenopausal women that came out as uterine cysts of mullerian origin on histology [12-14]. These findings were consistent with our case report where we suspected an ovarian cyst while histopathological examination confirmed a mullerian cyst.

There is a close correlation between mullerian cysts and benign endosalpingiosis. It is a condition where multicystic masses are seen involving female reproductive system, lined by ciliated columnar epithelium closely resembling tubal mucosa [3,13]. There are few case reports in literature where endosalpingiosis has been described to arise from primary mullerian epithelium instead of tubal mucosa [15]. Endosalpingiosis of uterus may present with pain, menorrhagia, infertility and pelvic mass in reproductive age group. A thorough clinical and diagnostic assessment is required to differentiate it from endometriosis, endocervicosis and mullerian tumors, leiomyoma ad inclusion cysts [12,16]. Due to scarcity of information on this topic, these benign conditions are seldom reported with accuracy preoperative and definitive diagnosis is dependent upon histopathology [17-19].

Conclusion

Mullerian cyst of uterus is a rare pathology, and the available evidence is based only a few case reports. Our patient is the youngest of all cases reported so far. Due to the rarity of this condition, preoperative diagnosis is challenging, therefore, definitive diagnosis is made at histology. Further research is needed for implications of this diagnosis on the woman's health in future and the best imaging modality.

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