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Case Report

A rare case scenario of didelphic uterus with ovarian serous cystadenocarcinoma: an unusual clinical finding

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ABSTRACT

Uterine didelphys in women is uncommon. The improper development of the Mullerian ducts during the intrauterine period leads to congenital abnormalities of the female genital system. Mullerian duct canalization or improper fusion leads to a didelphic uterus. About 60% of ovarian neoplasms are epithelial in origin, and the surface epithelium is typically most affected. This case report describes the management of a patient who had previously undergone two caesarean sections for a rare gynecological case of didelphic uterus with unilateral serous cystadenocarcinoma of the ovary.

Keywords: Cystadenocarcinoma, Mullerian duct, Uterine didelphys, Ovarian lesion

INTRODUCTION

Congenital anomalies of the female reproductive system include abnormalities of the mullerian ducts. The cause is an aberrant mullerian duct formation during the embryological stage in the growing fetus. These anomalies in development take place during the time when an embryo is developing its organs. Mullerian duct development abnormalities, inadequate fusion of mullerian ducts, canalization failure, and incomplete reabsorption of the uterine septum are a few examples. These congenital defects might occur anywhere between 0.5 and 5.0 percent of the time.[1-4] WTJ, Pax2, WNT2, PBXJ, and HOX genes have a hereditary tendency for congenital uterine abnormalities.[5] Mullerian ducts and Wolffian ducts develop together, hence in 30 to 50% of cases, congenital mullerian abnormalities have also been associated with renal system anomalies such unilateral renal agenesis, hypoplastic kidney, horseshoe kidney, ectopic or duplicate ureters.[6] Spinal anomalies such extra or incomplete vertebral bodies have been linked to 10 to 12 percent of cases.[7] A particular kind of fusion defect known as uterine didelphys is defined by the complete failure of the Mullerian duct to fuse. Due to this, there are two distinct uterine cavities, two cervixes, and a potential for a double vagina. There might be a presence of longitudinal vaginal septum, it may be thin, thick or inelastic. With the absence of symptoms, these abnormalities remain undiagnosed and diagnosis is usually made during evaluation of obstetric or gynecological complaints with routine per speculum examination. According to ASRM classification for mullerian anomalies 2021, the mullerian anomalies have been classified into the nine basic categories: Mullerian agenesis, cervical agenesis, unicornuate uterus, uterus didelphys, bicornuate uterus, septate uterus, longitudinal vaginal septum, transverse vaginal septum and complex anomalies.[8] These abnormalities can be diagnosed with various diagnostic modalities such as hysterosalpingography, saline infusion sonography, hysteroscopy, laparoscopy and laparotomy.

In developing countries there is an increase in incidence of ovarian tumors.[9] In these countries 9 to 17 women per 1 lac women are affected with epithelial ovarian

neoplasms.[10] The overall incidence is markedly high in developed nations and its incidence increases with advancing age groups, especially in women belonging to age between 60-64 years. Epithelial ovarian cancer estimates around 5% of deaths related to cancer.[11] Risk factors such as pregnancy in younger age groups, nulliparous women and women with early menopause are linked with increased risk of developing epithelial ovarian tumors. Very little association and lower risk of ovarian cancer development has been seen in women on oral contraceptives.[12]

CASE REPORT

A 40-year-old female reported to the outpatient department of obstetrics and gynecology with the chief complaint of gradual increase in abdominal swelling for one year. Patient also experienced generalized pain covering the entire abdomen within eight months followed by increase in size of the swelling within the duration of four months with symptoms like breathlessness - insidious in onset which worsened on recumbency impacting sleep, generalized weakness, vomiting and difficulty in walking. Sensation of fullness over abdominal region was present and has lead in decreased appetite and incomplete voiding of urine and constipation was also experienced by patient. Patient reported two cesarean sections. Apart from it, no history of the past medical or surgical illnesses and no associated family history related to malignancy was noted. Despite of all these symptoms, the patient didn't seek any medical assistance before.

At the time when she had decreased appetite and increased abdominal swelling she was brought to the hospital. Abdominal examination revealed that abdomen was grossly distended. Lesion was palpable and from the pelvis to the diaphragm the abdomen was occupied. Veins were dilated and engorged. At the level of the umbilicus abdominal girth was found which was 42 inches. In the flanks, bowel sounds were heard. Large abdominal mass was corresponding to 30 weeks had smooth surface, consistency was cystic in nature, with a restriction in mobility, lower border of the mass could not be palpated easily. Past caesarean scar was healthy and well-healed. Vaginal vault was found healthy and bilateral fornices were obliterated due to presence of the mass on per speculum and per vaginal examination.

Postero-anterior radiographic view of chest was normal. Sonographic examination revealed a large well-defined large multiloculated multi-septate cystic lesion of 27×23×20 cm in size arising from pelvis, extending from right adnexa up to the epigastrium, liver and spleen were displaced supero-laterally, and posterior displacement of the kidneys and peripheral displacement of bowel loops. Gall bladder stone was present, 8 mm in size. On color doppler, vascularity was not seen within the septations. Continuous vascularity with low peripheral impedance was present. Two uteruses with 2 cervix and widely separated horns and ovaries were seen (Figure 1).

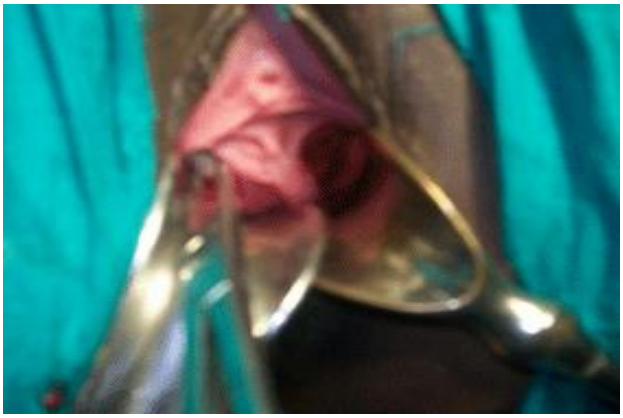

Figure 1: Per speculum examination showing 2 vagina and 2 cervices.

Kidney function tests and liver function tests were normal. Values of CA-125 were 50.61 IU/ml. Magnetic resonance imaging revealed large well defined thick irregular wall lesion arising from left ovary in pelvis and lying antero-superiorly to uterus and urinary bladder (Figure 2). Lesion showed superior displacement of bowel loops. From the lesion left ovary was not seen separately. Lesion demonstrated subtle hypo intense signal intensity on T2W and STIR images and hyper intense signal intensity on T1W and T1WFS images. Lesion approximately measured 29×25×15 cm (ap*ml*cc). Superiorly, the lesion was reaching up to infra-hepatic region.

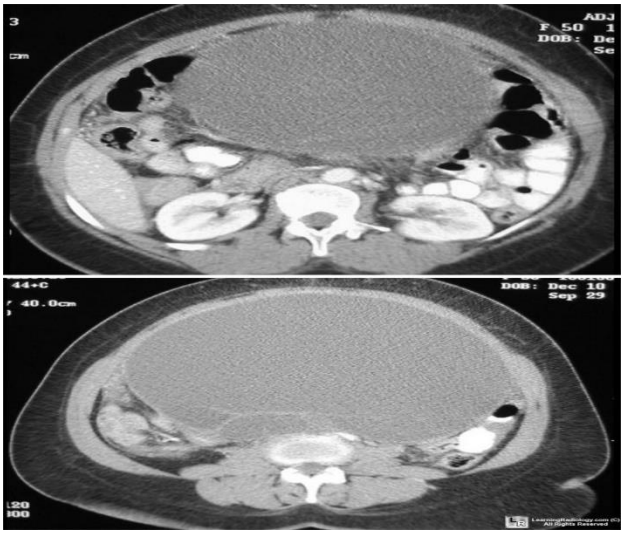

Figure 2: Magnetic resonance imaging showed large well defined thick irregular wall lesion arising from left ovary in pelvis.

Exploratory staging laparotomy was performed. Above the umbilicus level, an extending vertical midline incision was used to open the abdomen. A tensed smooth surfaced cystic mass of size 30×27×20 cm was found. The mass extended up to inferior surface of diaphragm and later it was delivered out. Fallopian tube found to be overstretching the tumor, no ascites was present. The left fallopian tube was found to be thinned out, completely

adherent, and stretched over the cystic lesion. Free fluid was found to be absent in the abdomen. Complete excision of the cyst was performed. A large multi-cystic lesion was arising from the left ovary. A multiloculated cyst displacing liver, spleen and bowel loops was seen. Surgical staging along with total abdominal hysterectomy along with salpingo-opherectomy was performed (Figures 3 and 4). Peritoneal washings and omental biopsies were taken and sent for histopathological examination.

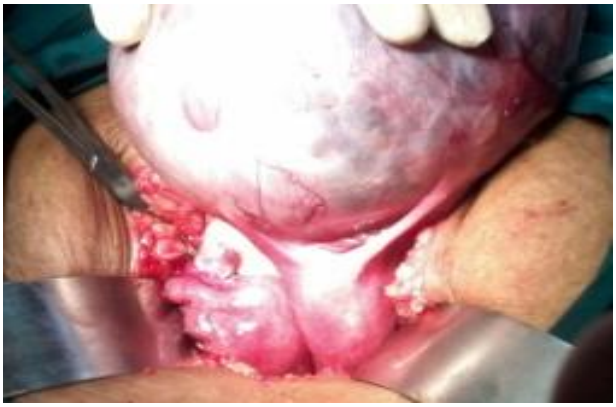

Figure 3: Exploratory staging laparotomy: tense smooth surfaced cystic mass.

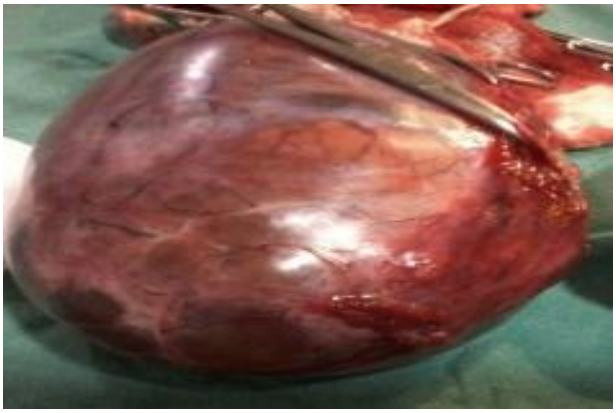

Figure 4: Specimen showing large serous cystadenocarcinoma of ovary.

Histopathologic investigations as shown in, revealed cyst thin-walled cyst with lining columnar epithelium (Figure 5). Nuclear atypia was absent and increased mitotic index was seen which suggested it to be benign serous cystadenocarcinoma. Postoperative period was uneventful. Malignancy was ruled out. cyst decompression was done with a controlled drainage procedure and around 3 liters of fluid was drained intra-operatively. Healthy ovarian tissue was not visualized separately. Intraoperative complications were absent with minimal blood loss. Biopsy of the omentum was taken and it was found negative. There was no involvement of lymph nodes. Histopathology revealed serous cystadenocarcinoma of ovary. The patient was discharged on the third day after operation. After a period of six months post-operative, the patient is healthy with no disease recurrence.

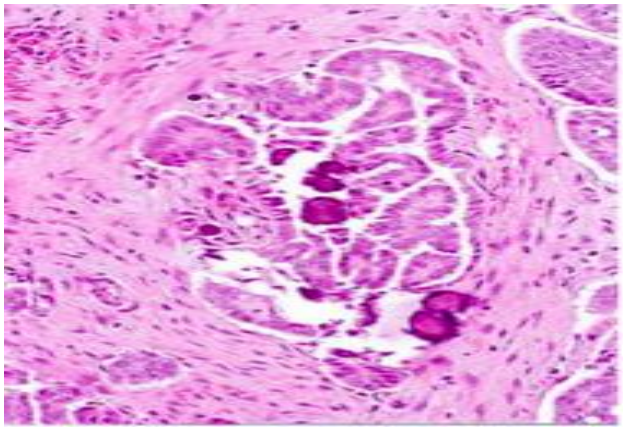

Figure 5: Psammoma bodies seen on histopathological report.

DISCUSSION

Congenital uterine anomalies are a rare clinical finding. These congenital anomalies result from the failure of fusion of two Müllerian ducts. The first case associated with uterine didelphys with renal agenesis was first reported in 1922.[13] Uterine didelphys is not usually associated with infertility, and the fetal outcome in these patients is good.[14] Müllerian duct abnormalities are usually associated with abnormalities of the genitourinary system.[15] This case report includes rarest of the Mullerian anomaly and an extensively large sized serous cystadenoma of the ovary, but the patient had no clinically significant findings from the gastrointestinal and genitourinary system. Ovarian tumors are usually age independent and can usually occur at any age.[16] Based on their cellular origin, they are classified into epithelial, stromal, and germ cell tumors. Serous cystadenomas are the more common benign cystadenomas, typically accounting for 75% of benign cystadenomas, and 25% of these are mucinous cystadenomas. CA 125 levels are used to detect malignancies of epithelial origin. Malignant tumors of epithelial origin of the ovary are identified using cancer antigen (CA)-125 levels. Diagnosis is usually made after microscopic examination of an ovarian mass. Serous cystadenomas are fluid-filled, usually thin-walled cysts with papillary projections inside.[17] These cysts are usually multilocular. Computed tomography (CT) scan and magnetic resonance imaging (MRI) are the best diagnostic methods for enlarged ovarian lesions.[18] Cysts are treated surgically by exploratory laparotomy, laparoscopic cystectomy, and removal of the ovary along with the cyst.[19] Examination of the contralateral ovary is mandatory.

CONCLUSION

We presented an uncommon case of a didelphic uterus with unilateral serous cystadenocarcinoma of the ovary. This is a rarity and often excluded from possible differential diagnoses within the included population. With early diagnosis and intervention being a pillar of this

gynecological condition, highlighting the possible incidence of such a case is significant. This case highlights the need to perform thorough clinical, radiological, pathological, and laboratory examinations on all the included study patients.

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REFERENCES

1. Heinonen PK. Uterus didelphys: a report of 26 cases. Eur J Obstet Gynecol Reprod Biol. 1984;17:345-50.
2. Heinonen PK. Clinical implications of the didelphic uterus: long-term follow-up of 49 cases. Eur J Obstet Gynecol Reprod Biol. 2000;91:183-90.
3. Raga F, Bauset C, Remohi J, Bonilla MF, Simón C, Pellicer A. Reproductive impact of congenital Müllerian anomalies. Hum Reprod. 1997;12:2277-81.
4. Grimbizis GF, Camus M, Tarlatzis BC, Bontis JN, Devroey P. Clinical implications of uterine malformations and hysteroscopic treatment results. Hum Reprod Update. 2001;7:161-74.
5. Hutson JM, Grover SR. Malformation syndromes associated with disorders of sex devdopment. Nat Rev Endocrinol. 2014;10:476.
6. Sharara FI. Complete uterine septum with cervical duplication, longitudinal vaginal septum and duplication of a renal Collecting system. A case report. J Reprod Med. 1998;43:1055.
7. Kimberley N, Hutson JM, Southwdl BR. Vaginal agenesis, the hymen, and associated anomalies. J Pediatr Adolesc Gynecol. 2012;25:54-8.
8. Ludwin A, Tudorache S, Martins WP. ASRM Mullerian anomalies classification 2021: a critical review. Ultra Obstet Gynaecol. 2022;59:1-8.
9. Parkin DM, Whelan SL, Ferlay J, Raymond L, Young J. Cancer Incidence in Five Continents. Int Agency Res Can. 1997;18:50-9.
10. Pisani P. Burden of cancer in developing countries. Int Agency Res Can. 1994;15:31-9.
11. Sung H, Ferlay J, Siegel RL. Global cancer statistics 2020: GLOBOCAN estimates of incidence and mortality worldwide for 36 cancers in 185 countries. CA Cancer J Clin. 2021;71:209-9.
12. Berek JS, Friedlander M, Hacker NF. Epithelial ovarian, fallopian tube and peritoneal cancer. Gynecol Oncol. 2020;7:15-32.
13. Scully RE, Young RH, Clements PB. Tumors of the ovary, maldevel-oped gonads, fallopian tube, and broad ligaments. Atlas Tumor Pathol. 1998;3:45-51.
14. Siegel RL, Miller KD, Jemal A. Cancer statistics, 2020. CA Cancer J Clin. 2020;70:7-30.
15. Negri E, Franceschi S, Tzonou A. Pooled analysis of 3 European case-control studies of epithelial ovarian cancer: I. Reproductive factors and risk of epithelial ovarian cancer. Int J Cancer. 1991;49:50-6.
16. Dhillon PK, Yeole BB, Dikshit R, Kurkure AP, Bray F. Trends in breast, ovarian and cervical cancer incidence in Mumbai,India over a 30-year period, 1976-2005: an age-period-cohort analysis. Br J Cancer. 2011;10:723-30.
17. Goff BA, Mandel L, Muntz HG, Melancon CH. Ovarian carcinoma diagnosis: results of a national ovarian cancer survey. Cancer Surv. 2000;89:2068-75.
18. Fealy J, Nelson JH. Adenocarcinoma in one-half of a uterine didelphys. Med Annals District Columbia. 1957;26:76-7.
19. Fanfanio F, Fagotti A, Restaino G. Endometrial cancer arising in the right cavity diagnosed by 2-dimensional sonography and magnetic resonance imaging. Gynecol Oncol. 2006;101:537-9.

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