ABSTRACT

Mullerian duct malformations delineate a miscellaneous group of congenital anomalies that result from the arrested development, abnormal formation, or incomplete fusion of the mesonephric ducts. Unicornuate uterus with rudimentary horn is a rare mullerian duct anomaly of female genital tract. The frequency of this pathology is 1/100 000. It is responsible for many obstetrics and gynecological complications during reproductive life of women. These uterine anomalies are either diagnosed incidentally or the patient may present with obstetrical or gynecological problems. These patients present with dysmenorrhea, dyspareunia, and rarely chronic pelvic pain because of endometriosis and rarely with acute abdominal symptoms following distention and torsion of the non communicating rudimentary horn. We present a case of pelvic endometriosis due to non communicating but functional rudimentary horn of a uterus in a multiparous woman.
Introduction
Mullerian duct malformations delineate a miscellaneous group of congenital anomalies that result from arrested development, abnormal formation, or incomplete fusion of mesonephric ducts. [1] The incidence of uterine anomalies in a fertile population is reported to be around 3.2%. [2] Unicornuate uterus with a rudimentary horn is a very rare congenital malformation of female genital tract. The frequency of this pathology is approximately 1/100,000. A rudimentary horn usually develops following incomplete development of one of the mullerian ducts. [3] Besides gynecological complications such as endometriosis, primary infertility and hematometra, anomalies of the urinary system and obstetric problems such as malpresentations, habitual abortions and premature births can occur.[4]

Case Report
This patient 40 year old , para3 with 3 living issues was presented to the gynecology outpatient department with history of progressive pain in left lower abdomen from last 6 months to 1 year duration. Her menstrual cycles were of 30-35 days with slightly decreased flow since last 5-6 months. There was no other menstrual complaints, no history of dyspareunia or bowel and bladder disturbances. She was receiving antispasmodics and NSAIDS for her pain for last one year. Her per abdominal examination were normal. Her clinical pelvic examination revealed a mass of about 6 by 6 cms in the left fornix ,soft to firm in consistency, adherent to uterus. Uterus was normal in size with slightly restricted mobility. There was tenderness over the mass on bimanual examination. Pelvic ultrasound examination was done which revealed 7cm by 6 cm multiloculated space occupying lesion in the left adenexal region. MRI pelvis revealed a large well defined lobulated solid cystic lesion with no obvious septations of approximately 10cm by 7.6 cm in size seen in left iliac fossa extending into the left side of pelvis. Left ovary was not seen separately from the lesion .Right ovary and uterus was normal.MRI findings were suggestive of complex ovarian cyst or neoplastic ovarian mass. Her hemoglobin was 11 gms percent and all her serological test were normal. Her CA -125 levels was 243.3u/ml.

Patient was posted for laprotomy with the possibility of large left ovarian dermoid cyst, tubo ovarian mass or sub serous fibroid. On laprotomy a large cystic lesion was found on the left side of pelvis which was adherent to uterus and also to lateral pelvic wall. Cyst was separated from all sides and from omental and bowel adhesions by blunt and sharp dissections till the base of pedicle was reached which was clamped and removed. Thick endometriotic fluid was present in cyst. On right side another uterus was found with the right tube and ovary. The right round ligament arose from the right uterine cornual region. The left round ligament arose from another mass of about 4 cm in size which was considered a rudimentary horn. The left uterine tube arose from the superior portion of rudimentary horn . So it was diagnosed as a case of Unicornuate uterus with non communicating rudimentary horn with functional endometrium. (Fig 1). Cut section of rudimentary horn showed small cavity with thin endometrial lining. (Fig 2). The decision for hysterectomy was taken in view of perimenopausal age group of patient ,complete family size, risk of recurrence of chronic pelvic endometriosis and refusal of patients consent for fertility conserving surgery. Patient withstood the procedure well and her post op period was uneventfull.On discharge patient was kept on monthly injections of GnRH analogue upto 3 months. Histopathology of cyst wall confirmed the diagnosis of endometrosis.Histopathology of uterine horn confirmed the diagnosis of its functional endometrium.
Discussion
The true incidence of Unicornuate uterus is not known. Current estimates are based only on those few cases which are actually diagnosed and subsequently reported in peer review journals. The reported incidence of uterine anomalies in fertile population is around 3.2%.[2] Unicornuate uterus with rudimentary horn is a rare type of mullerian duct malformation representing only 1-2% of congenital mullerian anomalies.[5,6] It results from defective fusion of the malformed duct with the contra lateral duct.[7] The uterine anomaly covers a wide range of anatomical variability and is divided into four subgroups according to American Fertility Society classification of mullerian anomalies: (IIa) rudimentary horn with cavity communicating to Unicornuate uterus, (IIb) with cavity non communicating, (IIc) with no cavity,(IId) with no horn. [8] Type IIb is the most common and clinically significant type. A fibrous or fibro muscular band usually connect the horns, but in 80-90% of cases there is no communication.

This condition is often asymptomatic due to lack of functional endometrium.[9] However when the horn is lined with functional endometrium, the resulting obstructed menstrual flow may cause severe cyclic pain shortly after menarche.[10] Retrograde menstruation is thought to initiate and potentiate endometriosis in women with non communicating uterine anomalies. Endometriosis seen in these cases supports the retrograde menstruation theory. The pain in endometriosis in these cases is usually severe and results in severe dysmenorrhea, chronic pelvic pain and dyspareunia.[11] The cause of pain in these cases may also be because of hematometra, causing distention of uterus. Another problem with the non communicating rudimentary horn is rudimentary horn pregnancy. [4] Since myometrial tissue is thin in a rudimentary horn of uterus, uterine rupture is seen frequently in rudimentary horn pregnancies. [4] The presence of gestation in a non communicating rudimentary uterus can be explained by transperitoneal migration of sperm. Although this is uncommon, these pregnancies may lead to serious complications. Urinary tract abnormalities are commonly associated with mullerian anomalies. This anomaly is associated with ipsilateral renal agenesis (67%) or ipsilateral pelvic kidney. [12]

The diagnosis of rudimentary horn is not made until the reproductive age when ruptured rudimentary horn pregnancy or pelvic pain occurs or sometimes it may be diagnosed accidently during laparotomy. The marked lower abdominal pain and pelvic pain usually bring patients for imaging in the form of ultrasound, CT scan or MRI which demonstrate a pelvic mass. In order to facilitate the surgical procedure, it seems important to be prepared for either presentation. Recent literature has suggested that MRI provides a considerably improved and accurate means of diagnosing and identifying mullerian anomalies. [13] Recently, three dimensional sonography has been introduced into clinical practice and offers advantages over two dimensional scanning as it provides fine anatomical anatomic details, useful for preoperative planning. [14]

The traditional surgical approach to treatment of this problem has been through laparotomy and removal of the dilated non communicating horn. Since the first report by Cannei et al (1990), laparoscopic resection of rudimentary uterine horn has rapidly become the standard treatment of such mullerian dysgenesis, specially to prevent severe complications as ectopic pregnancy or extensive endometriosis.[15] Though in our case decision for hysterectomy was taken, in considering the perimenopausal age, complete family and refusal for consent for fertility preserving surgery.

Conclusion
Non communicating horn of uterus is a rare cause of pelvic endometriosis. Women in reproductive age groups with dysmenorrhea or lower abdominal and pelvic pain with adenexal mass, the rudimentary horn with functional endometrium should be kept as differential diagnosis. Laparoscopic resection of non communicating rudimentary horn is the preferred treatment. It gives benefit to the patient of shorter hospital stay, significantly reduced postoperative morbidity and speedy recovery.

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