Placenta Accreta in a Rudimentary Horn Pregnancy

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Mullerian abnormalities serve as a fascinating framework with which to understand both the embryonic development and normal reproductive functioning. Uterine malformations are closely related to an abnormal uterine cavity, which is thought to impair the reproductive performance of patients. Pregnancy in a non- communicating rudimentary horn is extremely rare and, it is a life threatening condition because most cases terminate by uterine rupture by the second trimester. A case of such uterine anomaly, complicated by placenta accreta and ipsilateral renal agenesis in an 18-year-old primigravid is presented. Exploratory laparotomy with excision of left rudimentary horn uterus was performed. The hemorrhagic risk due to placenta accreta and that of spontaneous uterine rupture represent ample argument to recommend the immediate surgical removal of a pregnant rudimentary horn as soon as identification is carried out.

Key words: mullerian duct, placenta accreta, rudimentary horn, unicornuate uterus, transperitoneal sperm migration

Introduction

Mullerian duct abnormalities consist of a group of miscellaneous congenital anomalies of the female genital system. These result from arrested development, abnormal formation or incomplete fusion of the paramesonephric ducts. The true incidence of uterine anomalies in the general population is not accurately known[1] for the following reasons: 1) There is inaccuracy of the diagnostic methods employed, 2) Lack of a uniform system of classification, and 3) Some defects are asymptomatic, and therefore remain undiagnosed.[2] In a review of five studies with approximately 3000 cases, the mean overall incidence of uterine malformations in the general population and/ or the population of fertile women was 4.3% (Table 1).

Two paired mullerian ducts ultimately develop into the structures of the female reproductive tract. The structures include the fallopian tubes, uterus, cervix and upper two- thirds of the vagina. Complete formation and differentiation of the mullerian ducts into the segments of the female reproductive tract depend on completion of three phases of development: organogenesis, fusion and septal resorption.

During organogenesis, the mesonephric ducts regress and the paramesonephric ducts develop. This proceeds in a cephalad to caudal fashion. The more cephalad portions open directly to the peritoneal cavity and form the fallopian tubes. When one or both mullerian ducts do not develop, abnormalities such as uterine agenesis or hypoplasia, or unicornuate uterus result.

There are two kinds of fusion that occur. During lateral fusion, the two mullerian ducts fuse together to give rise to the epithelium and glands of the uterus and cervix. The medial aspects of the more caudal portions of the two ducts fuse, forming a median septum. Rapid cell proliferation occurs and a thick upper median septum is formed. When lateral fusion does not occur, this would result to either a bicornuate uterus or uterine didelphys. The formation of the vagina is completed by vertical fusion of the lower part of the Mullerian ducts that form the upper two thirds, and the ascending sino- vaginal bulb that forms the lower one third of the vagina. Complete

Table 1. Prevalence of uterine malformations in the general population or in the population of fertile women.

Study	Population Studied	Women (n)	Diagnosis	Uterine Malformations [n (%)]
Ashton, et al. (1988)	Transcervical tubal sterilization	840	HSG/ Hyst	32 (3.8)
Nesti, et al. (1990)	Varied indications	300	TVS	8 (2.7)
Maneschi, et al. (1995)	Abnormal uterine bleeding	322	Hyst	19 (5.9)
Acien (1997)	Consulted for contraception	241	TVS/ HSG/ Lap	21 (8.7)
	Without previous surgery	72		12 (16.7)
	With previous pregnancy and live newborns	131		6 (4.6)
	With previous pregnancy and some reproductive loss	38		3 (7.8)
Raga, et al. (1997)*	Fertile for tubal sterilization	1289	TVS/ HSG/ Lap	49 (3.8)
Total		2992		129 (4.3)

* Including the patients of a previous study from the same group (Simon, et al.)
HSG= hystserosalpingography; TVS= transvaginal ultrasonography; TDU= three-dimensional ultrasound; Hyst= hysteroscopy; Lap= laparoscopy

vertical fusion forms a normal patent vagina, while incomplete vertical fusion results in an imperforate hymen.

The third and final phase is septal resorption. After the lower mullerian ducts fuse, the median septum that was previously formed undergoes resorption, and a single uterine cavity and cervix are formed. Failure of resorption is the cause of septate or arcuate uterus.

Mullerian duct anomalies are the result of four major disturbances in the development, formation or fusion of the Mullerian ducts during fetal life: a) failure of one or both of the mullerian ducts to develop, leading to uterine or cervical agenesis, or unicornuate uterus; b) failure of the ducts to canalize leading to unicornuate uterus with rudimentary horns; (Figure 1) c) failure of abnormal fusion of the ducts, causing didelphys or bicornuate uterus; and d) failure of the resorption of the midline septum which leads to septate uterus or arcuate uterus.[1]

The American Fertility Society Classification of Uterine Anomalies was based on the proposed classification done by Buttram and Gibbons in 1979. The classification was based on the anomalies' degree of failure, similar clinical manifestations, treatments, and possible prognoses of their reproductive performance. The physician must be familiar with the consequences of the specific congenital anomaly on reproductive potential.[3]

One such anomaly is the unicornuate uterus, which results from the complete or almost complete arrest of the development of one mullerian duct. Several pertinent

Figure 1. Gravid rudimentary horn (left side of the picture) and the right hemiuterus, showing the thin fibrous band connection.

questions regarding this condition and its effect on pregnancy face the physician: a) How common are such abnormalities, and do these abnormalities necessitate further screening for other disorder?; b) How does an abnormality such as this occur, and what is the likelihood

that a patient's child will be affected by the same anomalies?; c) What is the probability that such a patient will deliver a healthy child, and what specific obstetric challenges will she face on the way to that desired endpoint?; d) Are there medical or surgical interventions that could improve the likelihood of having a healthy child?[1]

The Case

This is a case of L.B., 18-year-old primigravid, single, from Paranaque City, who was admitted for the first time at the Philippine General Hospital on November 4, 2008 for generalized abdominal pain.

Patient has an unremarkable past medical history as well as family history.

She is a high school undergraduate, currently unemployed, with no vices. She had her first coitus at 17 years old with one monogamous sexual partner. She has no history of oral contraceptive or IUD use. She has no history of sexually transmitted infections.

She is a primigravid and has had two prenatal check-ups at a local health center. Neither abdominal pain nor vaginal bleeding was noted. There were no baseline laboratories or ultrasound done.

She had her menarche at 12 years old with subsequent menses occurring at regular monthly intervals lasting for 3-5 days using 3 pads per day. Her last normal menstrual period was on May 22, 2008 giving her an amenorrhea of 23 weeks and 2 days.

History of present illness started one day prior to admission when the patient complained of intermittent epigastric pain accompanied by nausea and vomiting. No consult was done nor medications taken. Twelve hours prior to admission, pain was localized at the right lower quadrant with increasing intensity. Nausea and vomiting were still noted. Six hours prior to admission, there was persistence of the symptoms prompting consult at the emergency room. She was subsequently referred to OB service for co- management.

On review of systems, there was no headache, cough, colds, dysuria or change in bowel habits. No watery or bloody vaginal discharge was noted.

On physical examination, patient was conscious, agitated, stretcher- borne. She was hypotensive with a blood pressure of 60/40 mmHg, tachycardic at 120 beats per minute, tachypneic at 30 cycles per minute and she was febrile at 37.6 C. She had pale palpebral conjunctivae, anicteric sclerae, no anterior neck mass, no cervical lymphadenopathies. She had equal chest expansion, clear breath sounds, and no retractions. She had adynamic precordium, distinct heart sounds, and no murmurs. The abdomen was distended, with direct and rebound tenderness on all quadrants. Involuntary guarding was likewise noted. There were no fetal heart tones appreciated. Iliopsoas and obturator signs were noted. Internal examination was done which revealed normal external genitalia, nulliparous vagina, the cervix was smooth and closed. The corpus and adnexae were difficult to assess due to the abdominal distention and generalized guarding. On digital rectal examination, there was good sphincteric tone, intact rectal vault and no intraluminal masses. Bilateral parametria were smooth and pliable. Pararectal tenderness was noted.

Assessment at this time was pregnancy uterine 23 weeks and 6 days age of gestation, cephalic, not in labor; to consider acute appendicitis, probably ruptured; hemorrhagic shock probably secondary to an intraabdominal bleed; anemia secondary to acute blood loss.

The following laboratory examinations were done: complete blood count, blood typing, serum BUN, creatinine and electrolytes. ABG and ECG were also done.

Initial resuscitation was done which included IV fluids, NGT insertion and blood transfusion.

Patient was immediately brought to the operating room and underwent exploratory laparotomy, evacuation of hemoperitoneum and products of conception, and excision of left rudimentary horn uterus under general anesthesia.

Intraoperatively, there was note of 3 liters of hemoperitoneum. The General Surgery Service referred to Obstetrics - Gynecology intraoperatively after finding the appendix to be grossly normal. The right hemiuterus measures approximately 8cm x 5cm x 3cm. The rudimentary horn was attached to the right hemiuterus by a thin fibrous band measuring 5cm x 3cm x 1cm. There was no communication with the main uterine cavity. The right fallopian tube and ovary appeared normal. The left ovary appeared normal and the tube was normally attached to the rudimentary horn. Round ligaments were noted on the right hemiuterus as well as on the rudimentary horn. The rudimentary horn was noted to have a point of rupture at the (Figure 2) antero- fundal area measuring 6cm x 8cm from which the amniotic bag and fetus were already extruded into the abdominal cavity. The placenta was deeply penetrating the myometrial wall, with full thickness invasion (placenta percreta) over an area occupying approximately 50 percent of the rudimentary horn. The rudimentary horn measured 17cm x 15cm x 5cm. There was no appreciable gross fistula or communication between the cut sections of the ligament connecting the right hemiuterus and the rudimentary horn. (Figure 3)

Palpation of the retroperitoneal space revealed absence of the left kidney. The rest of the abdominal organs were grossly normal.

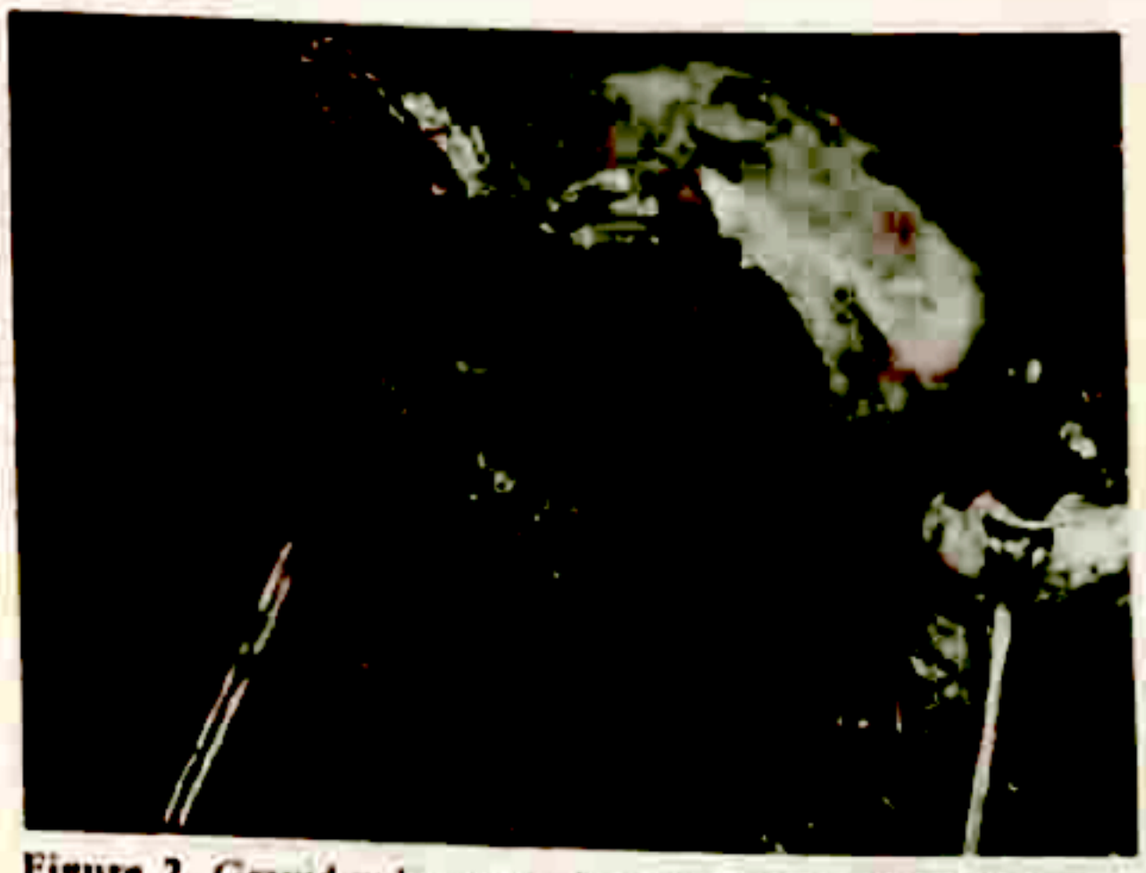

Figure 2. Gravid rudimentary horn with the point of rupture.

Figure 3. Cut section of the gravid rudimentary horn.

The male fetus weighed 400 grams, with a crown- heel length of 34 cm, approximately 25 weeks by fetal length. The umbilical cord length measured 17 cm. Intraoperative internal examination done revealed a single cervix on the right uterus. Estimated blood loss was 3.5 liters.

A total of four units packed red blood cell and one liter of colloids were transfused intraoperatively. Post-operative vital signs were as follows: blood pressure of 90/60 mmHg, cardiac rate of 110 bpm, and respiratory rate of 28 cpm, afebrile. She was eventually weaned off from the mechanical ventilator.

Patient had an unremarkable postoperative course. She was discharged on the 6th postoperative day, stable and improved. Final diagnosis was Unicornuate uterus with rudimentary horn pregnancy; placenta accreta (Figure 4); rudimentary horn pregnancy; renal agenesis, left, hemorrhagic shock secondary to massive intraabdominal bleed secondary to uterine rupture, resolved; anemia secondary to acute blood loss, resolved; G1P0 (0010).

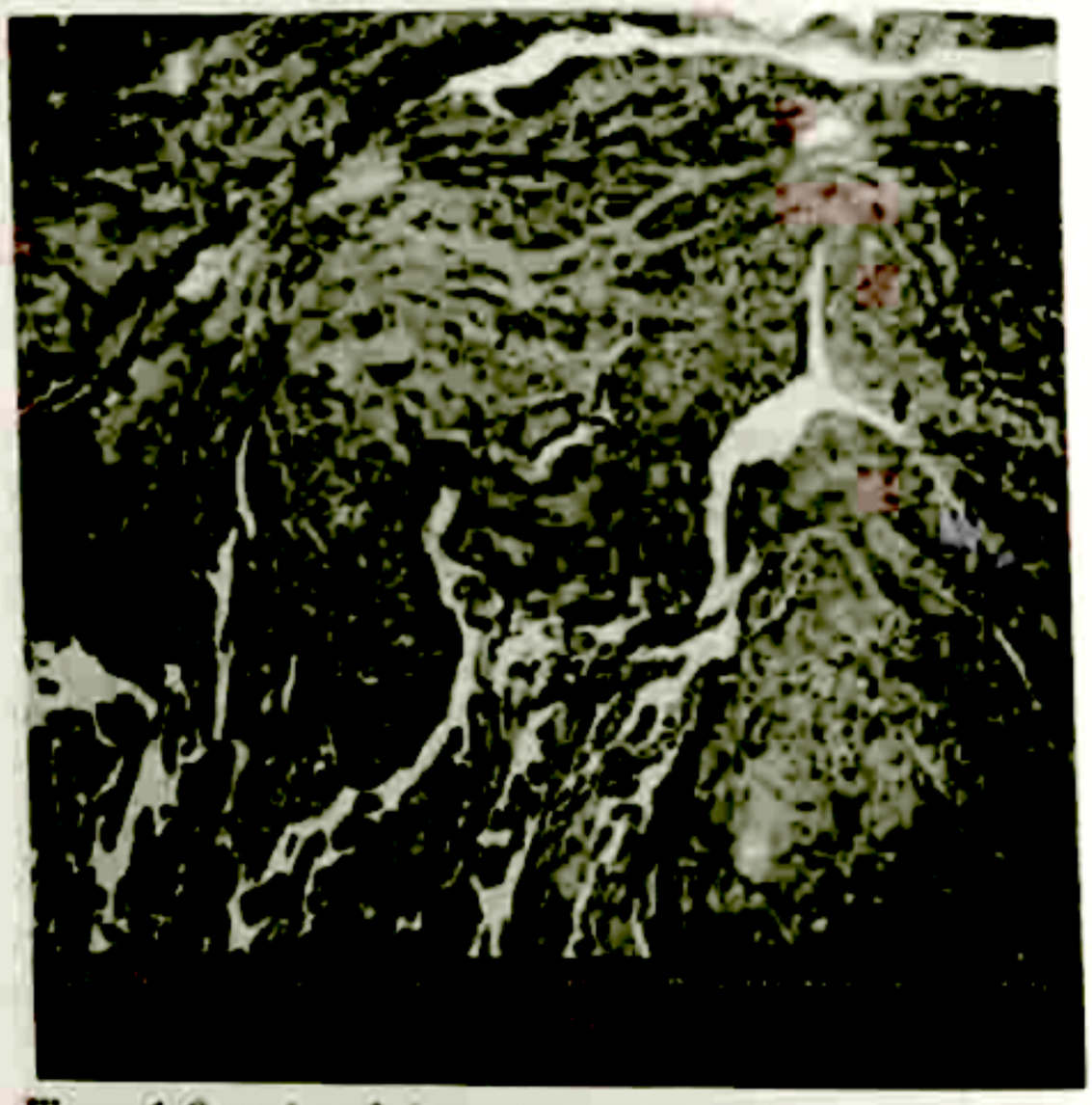

Figure 4. Scanning of placenta accreta showing the placental tissues adherent to the myometrium.

Discussion

The case presented is a peculiar one not only because of the rarity of the pathology but also due to the onset of circumstances and distinctive anatomical assemblage of the situation.

Embryology

The unicornuate uterus results from complete or almost complete arrest of the development of one Mullerian duct. Partial development of one of the ducts results in a

rudimentary uterine horn or anlage, whereas complete failure of the duct to develop leads to an isolated hemiuterus without a contralateral structure.[3] In 1979, Buttram and Gibbons proposed a classification based on the type and degree of failure of normal development of the female genital tract. Four variations of the unicornuate uterus were described: an isolated unicornuate uterus with no contralateral structure (type B), and three variations with an anlage, or rudimentary horn, is present contralateral to the unicornuate uterus. This rudimentary horn may have a cavity that is either in communication with (Type A1a) or sealed off from (Type A1b) the primary uterine cavity, or it may have failed to canalize entirely and is without a cavity (type A2).[3]

Incidence

The true incidence of the unicornuate uteri is difficult to determine since patients who do not develop obstetric or gynecologic complications may go unrecognized. Based on a meta-analysis study done by Nahum on 2008 regarding the incidence of the different types of uterine anomalies, unicornuate uteri constitute about 5 percent of the general population. Studies have indicated that the unicornuate uterus is almost always with rudimentary horn, and that the most uncommon type of all uterine anomalies was the unicornuate uterus without a rudimentary horn. The embryologic predominance of the unicornuate uterus to be on the right has not been explained.[4] In the case presented, intra- operatively, the patient had a ruptured non- communicating rudimentary horn.

Imaging Techniques

Before the advent of Magnetic Resonance Imaging (MRI) and ultrasound (US), the primary imaging modality for evaluating uterine anomalies in general was limited to hysterosalpingography. (HSG). The examination provides a morphologic assessment of the endometrial and endocervical canals and supplies important information regarding tubal patency.[4] Usually, the question of mullerian duct anomaly arises during HSG examination, if the typical trigone configuration of the cavity is not demonstrated.[14] Certain criteria are used to increase the level of confidence in diagnosing the type of anomaly, namely, the intercornual distance, the intercornual angle, and T- shaped cavity. However, overlaps exist between the different subtypes if they are on the above criteria. The major limitations of the procedure are the ability to characterize only patent canals and the ability to evaluate the external uterine contour adequately. HSG also entails exposure to ionizing radiation. The only anomaly in which HSG plays a significant role in diagnosis is the DES uterus, wherein the uterine cavity is clearly depicted but will only manifest as uterine hypoplasia in US or MRI. In the unicornuate uterus, the uterus is seen as being shifted from the midline, and filling of a small communicating rudimentary horn may be appreciated, although HSG cannot clearly delineate non- cavitary and non-communicating rudimentary horns.

Ultrasonography also helps to evaluate uterine anatomy. This should be performed during the secretory phase of the menstrual cycle, when the endometrial thickness and echo complex are better characterized. Images could be highly limited because of the patient's body habitus, uterine lie and shadowing from peristaltic bowel loops. The major limitation of US is that it is highly operator dependent. On US, the isolated unicornuate uterus appears small with deviation to one side of the pelvis. If a rudimentary horn is present, it may simulate a prominent cervix and confuse the findings. The identification of a cavitary uterine horn may be difficult to differentiate from other types of duplicated uterus. Three- dimensional US may help to further characterize the anomaly.[4]

MRI has a reported accuracy of up to 100% in the evaluation of mullerian duct anomalies.[4] It has the advantage of providing clear delineations of the internal and external anatomy in multiple imaging planes, and gives a reliable depiction of the external uterine contour. The unicornuate uterus appears curved and elongated, with the external contour assuming a banana shape. Uterine volume is reduced, and the configuration of the uterus asymmetric.[4] The appearance of the rudimentary horn is variable; if the endometrium is absent, the horn is of low signal intensity, with loss of normal zonal anatomy; if the endometrium is present, zonal anatomy is preserved.

Complications

The unicornuate uteri are susceptible to many gynecologic and obstetric complications. This was shown extensively in the study done by Heinonen in 1983. Based on his findings of 20 patients with unicornuate uterus, 14 (70%) patients complained of dysmenorrhea. Endometriosis was the most common finding in operative procedures. Tubal pregnancies in the fallopian tube of the rudimentary horn were also noted, with one case that presented with rupture.

In the same study, the obstetric outcome of pregnancies occurring in the unicornuate uterus was also noted. The challenge faced by the patients with unicornuate uterus has long been thought to be pregnancy maintenance rather than impaired fertility.[3] The unicornuate uterus has been implicated in intrauterine growth restriction, miscarriage, malpresentation of the fetus, preterm labor and cervical

incompetence.[1] Spontaneous abortion rates are reported to range from 41- 62%, premature birth rates range from 10- 20%, and fetal survival rates from 38- 57%.[4]

Three main etiologies have been suggested to explain such outcomes: diminished muscle mass, abnormal uterine blood flow, and cervical incompetence.[3] The gestational capacity is jeopardized by the presence of only half the full complement of the uterine musculature.[6] Not only are the walls of congenitally abnormal uteri thinner than normal, but also, their myometrium diminishes in thickness as gestation advances causing inconsistencies over different aspects of the uterus.[3] Another postulated etiology for the adverse outcomes is the anomalous vasculature supplying the uterus. This poor vascularity would result in impaired fetal nutrition, diminished fetal size, and higher incidence of first trimester abortion from compromised uteroplacental blood flow.[3] Lastly, the cervix is often believed to be at least a part of the difficulty in maintaining a viable pregnancy. This is due to the abnormal ratio of muscle fibers to the connective tissue in the uterine cervix.[3]

Another condition of the unicornuate uterus that leads to obstetric and gynecologic complications is the presence of a rudimentary horn. A small cavity within such a space may be home for a functional endometrium. Gynecologic problems may include the following: endometriosis, caused by retrograde menstruation and metaplastic conversion of omnipotential mesothelium to functional endometrium; hematometra, hematosalpinx and pelvic pain.[3] This uterine appendage can be a site of implantation that results in horn gestation.[3] From 1966 to 2003, only 156 cases of rudimentary horn pregnancy have been reported.[7] This is possible especially in the variant where communication exists between the unicornuate uterus and the rudimentary horn. However, horn gestation also occurs in the variant with no communication to the primary uterine cavity. It has been estimated that 90 percent of the unicornuate uteri with rudimentary horn are non- communicating.[8] When pregnancies occur within this 'blind' uteri that has no outlet to the contralateral hemiuterus or to the cervix, there is a necessary requirement for the occurrence of intraperitoneal transmigration of sperm in order to allow the pregnancy to implant in the uterine horn.[9] The first supposes that spermatozoons go up to the peritoneum by the permeable fallopian tube attached to the primary uterus, transmigrate intraperitoneally and fecundate the ovule that had been released by either of the ovaries.[10] Transperitoneal migration of sperm is a frequent occurrence in human reproduction, and has been well established in the literature.[9] In a study done by Nahum, et al. in 2004, they demonstrated that intraperitoneal sperm transmigration occurs in approximately 50 percent of all cases of human pregnancy, suggesting the free migration of sperm throughout the pelvic peritoneal cavity occurs routinely following intercourse. In this study of 88 patients with rudimentary horn pregnancy, they noted that 60 percent of the rudimentary horn pregnancies had its corpus luteum on the ipsilateral side. Thus, obligatory transperitoneal migration of the sperm must have occurred to produce this pregnancy.

In the same study by Nahum, et al. 40 percent of the pregnancies had the corpus luteum on the contralateral side. Two alternative gamete transmigration possibilities were presented. First is that there could have been independent transperitoneal migration of both the sperm and the ova to reach the contralateral fallopian tube, and that fertilization occurred in the contralateral tube, with subsequent tubal transport of the fertilized ovum to the 'blind' rudimentary horn for implantation. Alternatively, fertilization might have ooccurred within the peritoneal cavity with subsequent intraperitoneal transmigration of the resulting fertilized ovum and contralateral tubal pick up. Either of these mechanisms could have occurred in the index patient presented.

Rupture of a pregnancy in the rudimentary horn is a well-known severe implication of this uterine anomaly.[8] This is due to the underdevelopment and poor distensibility of the uterine muscular wall. Usually, the rupture of the uterine wall occurs in the second trimester.[11] This complication took place in the case presented. The patient came in with severe abdominal pain with other signs pointing to an acute abdomen. Exploratory laparotomy was immediately done and upon opening, a gravid ruptured rudimentary was noted.

Another complication prominent in the case was the presence of placenta accreta in the gravid rudimentary horn. Placenta accreta in rudimentary horn pregnancy was first reported by Heinonen in 1983.[8] It was proposed that the placenta grew through the wall of the rudimentary horn.[8] The muscle of the rudimentary horn is particularly delicate because it is so thin. Moreover, non- functional endometrium usually provokes abnormal placentation.[12] This is due to the fact that there is poor development of the decidua. Also, the reduced uterine volume and poor distensibilty contributed to the abnormal placentation. Since then, of the 51 cases with rudimentary horn pregnancy reported in literature, 7 were associated with placenta accreta, suggesting that rudimentary horn pregnancy is more likely to be associated with placenta accreta than an intrauterine pregnancy.[13]

The patient also presented with an absent ipsilateral kidney. It has been proposed that unicornuate uterus could also be caused by failure of the urogenital ridge to develop properly, an occurrence that would result in complete absence of the kidney, mullerian structures and gonad on the affected side. Such patients have been reported in literature, although this unique anatomical constellation

is extremely rare; the most common finding is for the paramesonephric duct to be affected in isolation, with the ovaries bilaterally present.[3]

Conclusion

Pregnancy in a non- communicating rudimentary uterine horn has been described in literature. It is however unfortunate that most of these patients presented with complications of the said uterine anomaly. Transperitoneal migration of the sperm and ovum has been proposed as the mechanisms by which these pregnancies occur.

Abnormal placentation usually occurs in this type of uterine anomaly. The hemorrhagic risk due to this and that of spontaneous rupture due to the thinness of the myometrium represent sufficient arguments to recommend immediate surgical removal of the pregnant rudimentary uterine horn.

It is of paramount importance that these patients have early diagnosis. A thorough, accurate counseling regarding their treatment options and ultimate likelihood of reproductive success is offered to these patients.

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