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Case Study

Mullerian Anomalies Manifesting as Haemorrhagic Shock: A Series of Rare Cases

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ABSTRACT

Introduction: Congenital anomalies of the reproductive tract may involve the uterus,cervix, fallopian tubes or vagina. Uterine anomalies are the most common of the mullerian anomalies. They are mainly diagnosed during work up for infertility, recurrent pregnancy loss, and cyclic pelvic pain and at times they may be detected incidently. But sometimes these anomalies results in life threatening haemorrhagic shock. In the present series of case reports all the patients presented in haemorrhagic shock and diagnosed as cases of mullerian anomalies not known before. **Material & methods**: this is a retrospective study; all the patients presented in shockdue to mullerian anomalies were recruited for the study. Shock is graded as per protocols. **Case reports**: we are reporting a series of five patients all of different mullerian anomalies but the presentation was common i.e. haemorrhagic shock. All the patients presented with different degree of shock. All the patients were operated with a provisional diagnosis but intraoperatively diagnoses were completely changed. **Conclusion**: we need to be more attentive while managing cases of haemorrhagic shock regarding presence of mullerian anomalies.

Keywords: congenital, haemorrhage, mullerian.

1. INTRODUCTION

Normal development of the female reproductive tract wends through a complex series of events: mullerian duct elongation, fusion, canalization and septal

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resorption. Faliure at any step results in congenital anomaly which may involve uterus, cervix, fallopian tubes or vagina. Uterine anomalies are the commonest variety reported in 0.16 -10 % of cases. They occur in 3-4% of fertile and infertile women, 5-10 % of women with early recurrent pregnancy loss and upto 25% of women with late first or second trimester pregnancy loss or preterm delivery.2 Mullerian anomalies are classified according to American Society for Reproductive Medicine(ASRM)classification in seven types.

Although mullerian duct anomalies have a wide range of symptoms in reproductive age group from silent course to life endangering bleeding. The gold standard diagnostic modality for mullerian anomalies is magnetic resonance imaging (MRI) as it can distinguish between bicornuate, didelphic and septate uterus and can determine the extent of a uterine and vaginal septum. Other modalties include ultrasonography (USG) and hysterosalphingo-graphy(HSG)2.We are reporting five cases of mullerian anomalies where patients were unaware of these anomalies despite undergoing earlier investigations and even previous caesarean deliveries. These patients presented to us in various stages of shock due to haemorrhage as a consequence of these anomalies. Case 1 and case 2 in present series with such presentations are very rare and no such case reports have been reported so far in literature despite thorough medline search.

2. AIMS & OBJECTIVES

1. To highlight the need for a more attentive attitude of sonologists and clinicians regarding diagnosis of mullerian anomalies.

2. To highlight the spectrum of presentation of various mullerian anomalies for the knowledge of young budding clinicians.

Materials and methods:

Here is a retrospective analysis of five patients presented with haemorrhagic shock due to mullerian anomalies in the deptt. of Obst. & Gynae in a tertiary care government hospital. These patients were resuscitated and operated according to their ailment and were diagnosed intraoperatively only. Shock was graded according to standard guidelines as demonsterated in table 1. In the patient of torn vaginal septum with double cervix final diagnoses was made with hysterosalphingography. On follow up ultrasonography, hysterosalphingography was done for associated anomalies.

Table 1: Grading of shock (used in present case series for defining shock in different patient).

	Compensated (grade 1)	**Mild(grade2)**	**Moderate (grade3)**	**Severe (grade4)**
Lactic acidosis	+	++	++	+++
Urine output	Normal	Normal	Reduced	Anuric
Level of consciousness	Normal	Mild anxiety	Drowsy	Comatose
Respiratory rate	Normal	Increased	Increased	Laboured
Pulse rate	Mild increase	Increased	Increased	Increased
Blood Pressure	Normal	Normal	Mild hypotension	Severe hypotension

(Taken from book"Bailey&Loves Short Practice of Surgery Chap2 Shock &Blood transfusion" pg15, 25th edition.)

3. OBSERVATION

Table 2 presents the details of patient in tabular form. Further description of patients provisional diagnosis and intraoperative findings are given below.

Case 1:

A 20 yrs women presented to our emergency with moderate shock due to excessive bleeding per vaginum. Her husband told that they were newly married and failed to consummate marriage. Their repeated attempts lead to profuse bleeding per vaginum. She was resuscitated and simultaneously taken for surgery with a provisional diagnosis of postcoital tear. Intraoperatively it appeared as a case of torn longitudinal vaginal septal, tears were present on

both anterior and posterior vaginal wall 2-3cm above fouchette with a mucosal tag and double cervix as findings given in table 2. Tears were sutured and both the uterine cavity explored with uterine sound. Her follow up hysterosalphingogram revealed her as a case of uterus didelphys. This finding supported our suspicion of torn longitudinal septum as a cause of her haemorrhage instead of simple postcoital tear. Finally she was diagnosed as a case of uterus didelphys with mid longitudinal vaginal septum.

Table 2: Case scenario of all the patients in tabular form.

Sr.no.	Presenting features & degree of shock	Examination findings	Management of patient	Procedure & postoperative investigations &complications	Final diagnosis
1	20 yrs old newly married female with profuse bleeding per vaginum on repeated coital efforts **Moderate shock**	Tear in anterior and posterior vaginal walls 2-3 cm above the fouchette with a mucosal tag hanging with anterior vaginal wall, two cervix also visualized.	Under GA repair of vaginal wall tear done. Both the cervix &communicating uterine cavity explored with uterine sound. Mucoal tag removed.	Repair of vaginal tears and removal of septum done Intraop 2 unit & postop 2 unit whole blood transfused. 6 wks after surgery her HSG done	Uterus didelphys with probably mid longitudinal vaginal septum that tattered during intercourse.
2	24 yr old G2P1L1A0 with previous LSCS with 30wks pregnancy **Mild shock**	P/A 30 wk size gravid uterus,FHS 60-80 irregular, on needle aspiration haemoperitoneum was present. P/V oss closed ,no bleeding per vaginum	Under GA on laparotomy premature fetus delievered by LSCS, avg 800ml haemoperitoneum drained.Bicornuate uterus with e/o prev. scar on other cornu was present.In the pregnant cornu placeta invaded myometriumand serosa and bleeding was present from serosal surface	Emergency laparotomy f/b LSCS and right side hemihysterectomy leaving right side ovary. Postoperatively 2 unit blood given .	G2P1A0 with 32 wks pregnancy with bicornuate uterus with placenta percreta with previous LSCS
3	22 yr old G4P3L2 presented with 20wk pregnancy in **moderate shock**	P/A 16-18 wk size uterus palpable, tenderness present P/V uterus was 18 wk size with tenderness and fullness in all the fornices. Haemoperitoneum present on needle aspiration	Laparotomy done under GA, haemoperitoneum of 1.5-2.0 lt. drained, well formed fetus with placenta lying in abdomen. She had a non communicating ruptured rudimentary horn connected to normal size left horn with tube & ovary	Laparotomy f/b removal of rudimentary horn. 2unit whole blood given intraoperatively and 2 unit given postoperatively with required FFP & platelets. Patient kept in ICU for 6 hrs rest postoperative period was normal.	Lt. side Non communicating rudimentary horn with unicornuate uterus
4	24 yr old G2P1L0 with 30 wks pregnancy with previous LSCS in **severe shock**	P/A 32 wk size uterus with superficially palpable fetal parts and free fluid P/V oss admitting tip of finger with bleeding per vaginum.	Laparotomy done under GA, haemoperitoneum of 2.5-3 lit drained, fetus with placenta was taken out, patient had a complete septate uterus, tear was present over previous scar site extending laterally as well as upwards to round ligament	Laparotomy f/b removal of dead fetus and repair of uterus done, 3unit whole blood rushed intraoperatively and postoperatively with required FFP& Platelets. Patient shifted to ICU on ventilator,and shifted back after 48 hrs.	Complete sepatate uterus with H/O prev LSCS one year back for retained head of a dead fetus during breech extraction.
5	30 yr old G4P3L3 with 16 wks pregnangy in moderate shock	P/A 16-18 wk size uterus palpable, tenderness present P/V uterus was 14-16 wk size with tenderness and fullness in all the fornices. Haemoperitoneum present on needle aspiration	Laparotomy done under GA, haemoperitoneum of 1.5-2.0 lt. drained, well formed fetus with placenta lying in abdomen. She had a communicating ruptured rudimentary horn connected to normal size left horn with tube & ovary	Laparotomy f/b removal of rudimentary horn and fallopian tube. 1unit whole blood given intraoperatively and 2 unit given postoperatively with required FFP & platelets. Patient postoperative period was normal.	Lt. side communicating rudimentary horn with unicornuate uterus

(O/E= on examination, LSCS= Lower segment caesarean section, P/A=per abdomen, FHS fetal heart sound, P/V per vaginum, GA=General Anaesthesia, Lt.=litre, f/b=followed by, wks=weeks

Case 2:

A 24 yrs G2P1A0 patient presented in moderate shock with a provisional diagnosis of 32 wks pregnancy with previous LSCS with fetal distress with suspicion of scar dehiscience as details given in table 2. Intraoperatively a pale live baby delivered as vertex and. Placenta was found to be morbidly adherent in right fundo corneal region of the uterus and was bleeding profusely on the serosal surface shown in figure 1. No fallopian tube and round ligament was visible on left side of the uterus. On further exploration second horn of uterus with left side tube and ovary visualized and it came out as a case of communicating bicornuate uterus with single cervix. During the previous section probably the right horn was not visualized. An emergency hemihysterectomy was done for the patient in view of placenta percreta. Clamps were applied at the communicating part of two horns, the affected horn removed. Her postoperative period was uneventful and on further investigations in followup period, she had no other anomaly.

Case 3:

A 23 years G4P3L2 patient presented to us in severe shock. After her history, clinical examination and ultrasonographic analysis she was taken for emergency laparotomy with a provisional diagnosis of ruptured ectopic pregnancy provisionally pregnancy in interstitial part of fallopian tube. Peroperatively she had left side unicornuate uterus with a ruptured

noncommunicating rudimentary horn on right side bearing the pregnancy. It was attached with the functioning uterus by a fibromuscular strand which was cut and ruptured rudimentary horn was removed. She did well in postoperative period and no associated anomaly was detected on further ultrasonography.

Case 4:

A 24 yrs old G2P1 L0 patient reported to us in severe shock with 32 weeks pregnancy with superficially palpable fetal parts and history of lower segment caesarean section one year back. She was resuscitated and taken for laparotomy with a provisional diagnosis of rupture uterus. Intraoperatively fetus was lying free in the peritoneal cavity with rupture of previous scar extending in lower segment as well as upwards to round ligament. Her history revealed that her previous section was done for removal of impacted after coming head of a dead fetus with breech. The fetal head was detached during breech extraction. She was diagnosed as a case of complete uterine septum dividing the uterus in two halves during previous LSCS. She was advised for hysteroscopic removal of septum before next conception but she did not turned up and now reported in severe shock.

Case 5:

A 30 yrs G4P3A0 patient reported to us in moderat shock pain abdomen and four months pregnancy. She was resuscitated and taken for laparotomy with a provisional diagnosis of ruptured ectopic pregnancy of cornual region, findings are given in table 2. Intraoperatively she had a ruptured right communicating horn with a functional unicornuate uterus on left side with tube and ovary. Fully formed 14-16 wk fetus with small placenta was lying in the peritoneum. About 1.5- 2 liters of haemoperitoneum was drained. The horn along with fallopian tube was excised and patient had an uneventful postoperative period.

4. DISCUSSION

Mullerian anomalies can widely affect reproductive and obstetric outcome of a patient depending upon its type. Gynecologic problems associated with mullerian anomalies include primary amenorrhea, dysmenorrhea, outflow obstruction, dyspareunia, infertility and ectopic pregnancy. Obstetric problems include recurrent pregnancy loss, preterm labor, malpresentation, foetal growth restriction, dystocia, uterine rupture and retained placenta. In the present case series all the patients presented in various degree of shock due to complications of undiagnosed mullerian anomalies and managed accordingly. According to ASRM mullerian anomalies are classified in seven groups depicted in figure 2.Failure of the fusion of the lower mullerian ducts that form the vagina can result in a longitudinal vaginal septum. The septum can be complete or partial. In this varietyyoung patients may have difficulty in using tampons but in developing countries like India where the use of tampon is unusual, it is usually diagnosed with postcoital tear, infertility, abortion or vaginal delivery. In our series also, one newly married patient presented with postcoital tear. A retrospective review of 202 patients by Handed and colleagues found complete septum in 45.6% patients, high partial in 36.1 % and medium or low partial in 18.3% cases[3]. The frequency of uterine malformation was 99.4% in cases of complete orhigh partial septum and 30.3% in partial medium or low septum, the most common malformation was complete septate uterus (59.5%), uterus didelphys (24.3%) and partial septate uterus in 45% cases. [3]

In present case series, case 1 had a mid vaginal septum which got ruptured during intercourse which was also associated with uterus didelphys. A longitudinal vaginal septum is the commonest variety of vaginal septum contributing in 75% cases. [1] Heinonen and

associates also reported case series of 21 patients of didelphic uterus with vaginal septum. Conversely a patient with a longitudinal vaginal septum has a didelphic uterus[3]. Although the literature on postcotial tear revealed that lacerated sites were mostly located in right fornix which may be due to the dextrorotation of the uterus and the less distensibility of vagina.[4] However in present series, tears were present on anterior and posterior wall in lower vagina. Despite exhaustive search of medline, no literature on septal tear following intercourse was found. Hence it is probably the first case report on vaginal septal tear following intercourse.

Uterus didelphys is a class three anomaly and accounts for 5% cases of MDA. It occurs as a result of complete failure of mullerian ducts fusion. This anomaly is associated with poor reproductive outcome with spontaneous abortion rate of 32.2%, preterm birth rate of28.3%, a term delivery rate of 36.29% and a live birth rate1 of 55.95 %. However case 1 of present series with this anomaly could not be followed up for her future reproductive outcome.

Bicornuate uterus is classified as class 4 MDA and develops due to incompelete fusion of the uterovaginal horns at the level of uterine fundus. This anomaly is associated with an overall spontaneous abortion rate of 36%, preterm birth rate of 23%, a term delivery rate of 40.6% and a live birth rate of 55.2%. Placenta percreta is a rare entity in which the chorionic villi of placenta invades full thickness of myometrium. Presentation of placenta percreta as haemoperitoneum in 32 wks pregnancy with neither previa nor previous scar in bicornuate uterus in our case 2 is again a very rare finding and yet to get reported in literature. Literature reveals high association of mullerian anomalies with placental anomalies[5]. it may be due to poorly developed musculature, scanty decidualisation and small endometrial cavity.

Unicornuate uterus results from abnormal development and fusion of the mullerian ducts. It is usually associated with various degress of rudimentary horn which may be communicating or noncommunicating with the uterine cavity. The connection with the uterus may be fibrous or fibromuscular. There is no communication between two cavities in 75% to 90% of the cases and the incidence of pregnancy in noncommunicating horn is high as6 83 %. The chances of pregnancy in the rudimentary horn are due the transperitoneal migration of sperm or ovum from the opposite side. The uterine rupture is common (90%) during second trimester as revealed by literature. In present series also, case 3 and case 5 were having this anomaly which got complicated during second trimester in accordance with literature. The abnormal shape of uterus, insufficient muscular mass, reduced uterine volume and inability to expand appears to be the possible culprits of uterine rupture through the wall of vascular rudimentary horn resulting in severe hemorrhagic shock as in present series also.

Septate uterus is class 5 MDA and most common disorder comprising 50% of MDA'S. This anomaly results due to partial or complete failure of resorption of septum after fusion of paramesonephric ducts. It is the commonest reported anomaly complicating the pregnancy. Studies on septate uterus identified a pregnancy loss rate of 44.3%, preterm delivery rate of 22.4%, term delivery rate of 33.1% and live birth rate of2 50.1%[1]. Only septate uterus as a cause of uterine rupture is still not reported but congenital uterine anomalies are considered as a precipitating factor for rupture uterus. Uterine rupture may be due to abnormal development of lower uterine segment, previous scar and the abnormal traction on the uterine scar during labour. In present series although, case 4 she was diagnosed as a case of septate uterus while undergoing LSCS for obstructed head of dead fetus in her previous

pregnancy and was advised to come after 6 weeks for hysteroscopic removal of septum, still she did not turned up for followup. Lack of education, poverty and negligence are key factors prevailing specially in developing countries like india which invites a lot of easily preventable complications. In the VBAC trial of 183 patients, 25 patients had known mullerian anomalies, out of which 14 had bicornuate uterus, 5 had septate uterus , 4 had unicornuate uterus and 2 had uterus didelphys. In this trial also, 2 uterine ruptures were reported in anomaly group. [7, 8]

5. CONCLUSION

Mullerian anomalies are not uncommon developmental entities which are very commonly missed during early reproductive period. After facing five life endangering cases due to complication of these anomalies, it is firmly suggested to all clinicians and radiologist toremain vigilant about these anamolies to prevent any future lethal event with the life of any patient. . High clinical suspicion, early diagnoses and timely intervention are the keys to reduce patient mortality and morbidity.

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