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Unicornuate uterus with non-communicating rudimentary horn – different clinical presentations

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OBJECTIVE(S): To analyse gynecological and reproductive morbidities associated with unicornuate uterus with non-communicating rudimentary horn.

METHOD(S): The clinical details of 18 cases of unicornuate uterus with non-communicating rudimentary horn found on laparotomy in a duration of 7 years (April 1997 - March 2004) were reviewed.

RESULTS: Out of 18 patients three teenagers presented with dysmenorrhea and pain in abdomen and had hematometra in the non-communicating rudimentary horn. Seven had ectopic pregnancy, which occurred in non-communicating rudimentary horn of which five presented in a state of shock. Out of these seven ectopic pregnancies, five were seen in 1st trimester and two in 2nd trimester. Preoperative diagnosis of non-communicating rudimentary horn pregnancy was possible in two cases. Eight patients had intrauterine pregnancy and at cesarean delivery, they were diagnosed to be having unicornuate uterus with non-communicating rudimentary horn. Pregnancies associated with this condition had high incidence of abortion, preterm labor, malpresentations and cesarean delivery.

CONCLUSION(S): Unicornuate uterus with non-communicating rudimentary horn is a rare condition but is associated with many gynecological and reproductive morbidities. It should be diagnosed before pregnancy occurs or at the latest before rupture occurs and should be treated by immediate surgery.

Key words: unicornuate uterus, rudimentary horn, pregnancy

Introduction

Unicornuate uterus with a rudimentary horn is a rare type of mullerian duct malformation and results from the defective fusion of the malformed duct with the contra-lateral duct ¹. A fibrous or fibro-muscular band usually connects the horns of the ducts but in 80 - 90% of cases there is no communication. The rudimentary horn may consist of a functional endometrial cavity or it may be a small solid lump of uterine muscle with no functional endometrium.

This malformation is rare and it can be associated with many

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House No. 79, Sector 24-A, Chandigarh - 160 023 Tel. 2727219, 2727220 Email : poonam1302@yahoo.com complications throughout a woman's reproductive life beginning from menarche when hormonal stimulation may gradually activate the endometrium of the rudimentary horn. The resulting obstruction of the menstrual flow may cause hematometra, leading to endometriosis and infertility.

Pregnancy in non-communicating rudimentary horn is a rare form of ectopic gestation and its incidence is between 1/100,000 to 1/140,000 pregnancies ^{2, 3}. It occurs following transperitoneal migration of sperm or zygote.

Variable thickness of rudimentary horn musculature, dysfunctional endometrium and poor distensibility of the myometrium lead to rupture of the rudimentary horn. This complication is usually seen in the 2nd trimester and can be a life threatening condition for the mother resulting from hemoperitoneum and hemorrhagic shock. It is difficult to

diagnose preoperatively and in the literature, only 5% of rudimentary horn pregnancies were diagnosed preoperatively and the remaining were found unexpectedly at laparotomy ³.

If the pregnancy occurs in the semi-uterus of this malformation, it is associated with increased incidence of abortion, preterm labor and malpresentations. These patients also have high incidence of cesarean deliveries ^{4,5}.

With this background, a study was conducted where all cases of unicornuate uterus with rudimentary horn found on laparotomy were analysed to know their gynecological and obstetric implications.

Material and Methods

This study was conducted in a tertiary care teaching hospital. Eighteen cases, which were diagnosed to be having unicornuate uterus with non-communicating rudimentary horn at laparotomy (April 1997 to March 2004), were retrospectively analysed. These patients were divided into three groups. Group I (n=3) consisted of patients who presented with gynecological complaints and in whom the condition was diagnosed at laparotomy. Group II (n=7) consisted of patients with pregnancy in the rudimentary horn. Group III (n=8) consisted of patients who had pregnancy in the semi-uterus; all of them underwent cesarean section during which this malformation was diagnosed.

Age, presenting complaints, previous reproductive history, investigations, preoperative diagnosis and intraoperative findings were studied.

Group I

The age of the three patients in this group was 16, 17 and 19 years. One patient with complaints of severe dysmenorrhea and two presented with severe acute pain in the abdomen. These two patients also gave history of progressive dysmenorrhea and were operated with a diagnosis of twisted ovarian cyst. The patient whose complaint was only dysmenorrhea was operated with a preoperative diagnosis of hematometra in the rudimentary horn that was suspected clinically and on ultrasonography. Intra-operatively, all patients had hematometra in the non-communicating rudimentary horn and underwent excision of the rudimentary horn with ipsilateral salpingectomy. None of the patients had evidence of endometriosis.

Group II

Seven patients had pregnancy in the non-communicating rudimentary horn. Detailed data about these patients is given in Table 1. The average age of these patients was 27 years (range 22 to 39 years). Two patients presented in the first

trimester and five in the 2nd trimester. Pain was the chief complaint in all these women and five women were in a state of shock. Preoperative diagnosis of rudimentary horn pregnancy was suspected in two cases only and in the remaining, the diagnosis was made at laparotomy. The malformation was on the right side in four cases and on the left side in three. All women underwent excision of the rudimentary horn with ipsilateral salpingectomy.

Group III

The average age of presentation in the eight patients of this group was 25 years (range 20 to 32 years). All cases were diagnosed at lower segment cesarean section (LSCS). The average gestational age at LSCS was 36 weeks. LSCS was done for breech presentation in six cases, for transverse lie in one and for previous two LSCS in one. Table 2 gives the outcome of pregnancies that occured in Group II and III patients (previous and present pregnancies). There were 31 pregnancies in all. Eight ended in abortion 7 were in the rudimentary horn (Group II), and 16 pregnancies continued beyond 20 weeks of which two required cerclage and six ended in preterm labor or preterm premature rupture of membrances. The incidence of breech presentation was 56% (9/16) and cesarean section 75% (12/16).

Uterine malformation is usually associated with urinary tract malformation. Sonography for urinary tract or intravenous pylography could be done only in six cases. One had absent kidney, one had pelvic kidney, and one had duplication of the renal system on one side. Three had no abnormality.

Discussion

Unicornuate uterus with non-communicating rudimentary horn is susceptible to many gynecological and obstetric complications which can occur at any stage of reproductive life. It is difficult to truly estimate the incidence of these complications as the data available are in the form of case reports and surveys collected from the literature that usually has only the severe cases requiring surgery ⁵. To decrease the serious complications in future, early diagnosis is of utmost importance. The patient should be treated by excision of the rudimentary horn. High index of suspicion should be kept in teenagers presenting with dysmenorrhea and every effort should be done to exclude the condition by conducting relevant investigations in suspected cases. If in patients presenting with infertility, hysterosalpingography shows that the uterus is deviated to one side and there is unilateral tubal block, this condition should be strongly suspected. In our analysis, hematometra was seen in three cases only. This shows that rudimentary horn rarely has a cavity with functioning endometrium. Endometriosis is another complication that is seen in these patients. In one study the

Table 1. Pregnancy in the noncommunicating rudimentary horn.

Age Year	Obsteric History	Duration of gestation (weeks)	Clinical diagnosis	Preoperative diagnosis	Rudimentary horn location
27	$G_3P_1A_1L_1$	7	History of secondary infertility Unilateral tubal block on hystero- salphingography Pain in abdomen	Rudimentary horn pregnancy	Right side
23	G_{1}	9	History of primary infertility Pain, bleeding	Missed abortion in a bicornuate uterus	Right side
28	G_{1}	18	Pain in abdomen, shock	Tubal pregnancy	Right side
27	$G_2P_0A_1$	12	Pain in abdomen, vomiting, shock	Ruptured tubal pregnancy	Left side
24	$G_5P_2A_2L_1$	12	Vaginal bleeding, pain in abdomen	Rudimentary horn pregnancy	Left side
22	$G_2P_1L_1$	12	Pain in abdomen, shock	Ruptured tubal pregnancy	Left side
39	G ₃ P ₂ L ₂ with two cesarean deliveries	17	Pain in abdomen, shock	Ruptured uterus Ectopic pregnancy	Right side

In all cases the rudimentary horn was excised along with its tube. In the last case the left tube was ligated in addition.

Table 2. Outcome of current and previous pregnancies in Group II and Group III patients.

	Observation		Number
	Pregnancy in the rudimentary horn	07	(Group II)
	Pregnancy in the semi-uterus	08	(Group III)
	Total pregnancies (current and past)		
	a) Abortionsb) Rudimentary horn pregnancy		08 (25.8%)
			(22.6%)
	c) Deliveries		16 (51.6%)
	Cerclage required	02	(12.5%, 2/16)
	Preterm labor and preterm premature rupture of membrances	06	(37%, 6/16)
	Breech	09	(56%, 9/16)
	Transverse lie	01	(1.6%, 1/16)
	Cesarean section	12	(75%, 12/16)

incidence was as high as 20% ⁵. None of our cases had evidence of endometriosis.

Pregnancy in a rudimentary horn is very rare but it can be life threatening. Five of our patients presented in a state of shock due to rudimentary horn rupture. The duration of the pregnancy at the occurrence of rupture are dependent on the thickness of the myometrium. In some cases the pregnancy ends up in missed abortion or intrauterine fetal death due to decreased blood supply and defective endometrium as was seen in two of our cases who had missed abortion, one at 7 weeks and the other at 9 weeks. If the pregnancy grows, it usually overcomes the first trimester period uneventfully as the rudimentary horn is thicker than the fallopian tube and 80 - 90% of the ruptures occur in the second trimester 5,7. As it is associated with such catastrophy every effort should be made to diagnose it in early pregnancy but according to the literature, less than 5% of the reported cases were diagnosed preoperatively and mostly the diagnosis was made at laparotomy or laparoscopy 3. A careful pelvic examination in the 1st trimester showing deviated uterus with palpable contra-lateral pelvic adnexa should arouse suspicion of uterine anomaly 6. Ultrasonogaphy can also pick up this anomaly with reasonable accuracy. A gestational sac surrounded by myometrium by the side of a normal empty uterus and non-communication of gestational sac with the endometrial cavity and the cervix differentiates it from pregnancy in one of the horns of a bicornuate uterus 4,7,8. MRI and CT scan are also gaining popularity for diagnosing uterine malformations. Both clinically and radiologically the diagnosis is more accurate in the early first trimester when the two horns are separate in the pelvis. Once the diagnosis is strongly suspected these patients should be taken up for laparoscopy or laparotomy, depending upon the general condition of the patient, and the rudimentary horn should be excised along with its tube, so as to prevent future tubal ectopic pregnancy in that tube. In our study, preoperative diagnosis was possible on clinical and sonography findings in only two out of the seven patients.

There have been case reports of fetal survival in late rupture of rudimentary horn pregnancies. Thirteen survivals have been reported from such unusual and life threatening condition till 1999.

When the pregnancy is located in the semi-uterus of the anomaly, it is associated with high incidence of abortion, preterm labor, cervical incompetence, malpresentation and cesarean section rate as was also seen in our analysis (Table 2).

In all such cases, evaluation of renal system is indicated because of the high incidence of associated urological anomalies. In our study, only six patients underwent such evaluation and three showed normality, one had absent kidney, one had pelvic kidney and one had duplication of renal system on one side.

Every effort should be made to diagnose this condition before pregnancy occurs or at least before rupture occurs. The rudimentary horn should be excised whenever diagnosed.

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