

Case Report

**Unicornuate Uterus with Rudimentary Horn
and Complicated with Endometriosis*****Uterus Unikornuasi dengan Kornu Rudimenter dan Endometriosis***

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*Subdivision Urogynecology Obstetric and Gynecology**Department of Obstetrics and Gynecology/**Dr. Cipto Mangunkusumo Hospital**Jakarta***Abstract****Objective:** To report the congenital abnormalities of müllerian duct and review its literature.**Methods:** Case report.**Case:** A sixteen years old unmarried woman came to our policlinic with chief complain of cyclical abdominal pain (VAS 6). She had regular menstruation. In other hospital, she had diagnosed as suffering from ovarian tumor. Our examination showed that the uterus was unicornuate with rudimentary horn. We did explorative laparotomy. We took out the left cornu and left salpingoophorectomy due to hematosalping and endometrial cyst. The Visual analogue score after surgery was reduced until VAS 2.**Conclusion:** Congenital abnormalities of the müllerian ducts are relatively common. Comprehensive examination can diagnose this abnormality earlier, thus intervention can be given as soon as possible.

[Indones J Obstet Gynecol 2013; 1-4: 215-8]

Keywords: abnormalities müllerian ductus, endometriosis**Abstrak****Tujuan:** Untuk melaporkan kelainan kongenital pada duktus müller dan mereview beberapa literatur.**Metode:** Laporan kasus.**Kasus:** Nona 16 tahun mengeluhkan nyeri perut siklik (VAS 6). Pasien memiliki menstruasi teratur. Di Rumah sakit luar, pasien didiagnosis sebagai tumor ovarium. Pemeriksaan kami menyimpulkan bahwa pasien ini memiliki kelainan uterus dengan 1 kornu lainnya rudimenter. Kami melakukan laparotomi eksplorasi, mengangkat kornu yang rudimenter, tuba, dan ovarium yang berhubungan karena membesar akibat endometriosis. Skala nyeri setelah operasi berkurang menjadi VAS 2.**Kesimpulan:** Kelainan bawaan pada duktus müller sangatlah umum. Pemeriksaan yang menyeluruh dapat mendiagnosis kelainan ini lebih awal dan intervensi dapat dilakukan sesegera mungkin.

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Kata kunci: endometriosis, kelainan duktus müllerian**Correspondence:** Liva Wijaya, Department of Obstetrics and Gynecology Faculty of Medicine University of Indonesia/Dr. Cipto Mangunkusumo Hospital Jakarta Pusat, phone: 08122055012, email: bugmd13@yahoo.com**INTRODUCTION**

Congenital abnormalities of the müllerian ducts are relatively common, occurring in 7% to 10% of all women. It contributes to the problems of infertility, recurrent pregnancy loss, and poor pregnancy outcomes that occur in approximately 25% of women with uterine anomalies. Unicornuate uterus with a rudimentary horn is a rare Müllerian anomaly, with an incidence of 0.06%. A partial or complete lack of development of one Müllerian duct between the 7th and 8th week of gestation may result in the formation of a unicornuate uterus. The functioning unicornuate uterus may or may not be associated with a rudimentary uterine horn. Seventy-five to ninety percent of unicornuate uteri with rudimentary horns are non-communicating. The altered uterine configuration is associated with an increase

in endometriosis and in obstetrical complications (early spontaneous miscarriage, ectopic pregnancy, abnormal presentations, intrauterine growth retardation, and premature labor). Implantation in the rudimentary horn is followed by a very high rate of pregnancy wastage or tubal pregnancies. A rudimentary horn can also be the cause of chronic pain, and in this case, a surgical excision is recommended. However, most rudimentary horns are asymptomatic because they are noncommunicating, and the endometrium is not functional. Approximately 40% of patients with a unicornuate uterus will have a urinary tract anomaly (usually of the kidney). Surgical reconstructive procedures do not improve obstetrical outcomes.¹⁻³ The objective of this paper is to report our case and review its literatures.

CASE PRESENTATION

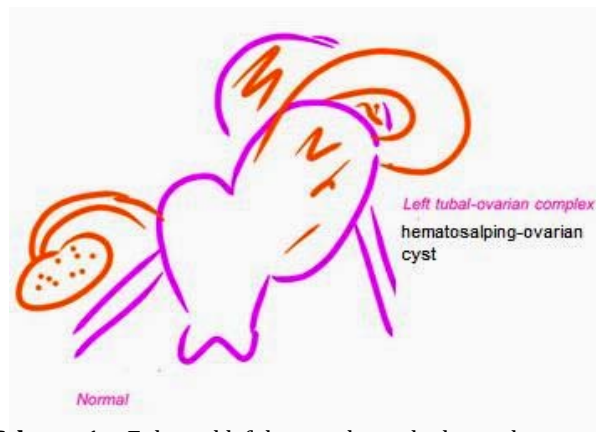
A sixteen years old unmarried woman came to our polyclinic with chief complain of cyclical abdominal pain (VAS 6) since 6 months before admission. She had regular menstruation, and each cycle has a duration of 7 to 10 days. This patient had US examination and CT scan performed out side of our hospital which revealed the presence of an ovarian cyst. She had another examination and it was revealed that she had 2 horn uterus with one cervix. Due to these two different diagnosis, she made another consultation in our hospital. On physical examination, she had normal secondary sexual sign. We found abdominal mass located on 2 finger above symphysis pubic. On inspection, we found that the vulva and urethra were normal. This patient had normal labia, normal vulva, and normal hymen. We performed vaginal ultrasound and we found that the length of the uterus was approximately 7-8 cm. On rectal examination we found normal shaped uterine. We found adnexal mass in the left side approximately 7 cm with limited mobility. No pain was felt. On US examination we found 2 fundal with 2 uterine cavum, correspond with 2 horn uterus. No abnormality was found inside of the right horn cavity, regular endometrium, 12 mm, connected with normal cervical canal. Left horn cavity has an opening of 33 x 28 mm, with eointernal material inside. This horn was not connected with normal cervical canal, thus we suspected that it was a rudimentary type. Right adnexa was normal, but on the left adnexa we found a cystic mass, round shaped, approximately 64x52 mm with echointernal material suspected as hematosalping. Normal left ovary made adhesion with the distal part of hematosalping.

Intraoperatively, after the peritoneum was opened, we found 3 cystic mass, diameter 8 cm from left adnexa corespond to ovarian cyst susp endometriosis cyst, 5 cm connected with uterus corespond to left horn, and tubuler cystic mass with diameter 3 cm corespond to left hydrosalping. On palpated we found normal right horn and ovary. There were adhesion in the posterior wall of the cyst and the colon descenden, we performed adhesiolisis. We identified the left IP ligament, we clamped, cut, and sutured. We also clamped, cut and sutured the left round ligament. After we performed left salpingoophorectomy, we can see that cervix, the right tube and ovary within normal limit. We cut the left horn, and made hemostatic suture interuptedly with vicril 1. We hang

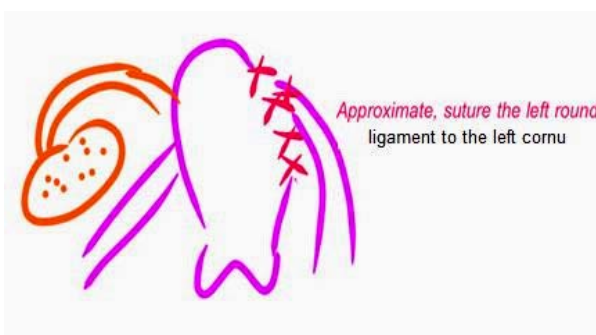
the left edge of the left cornu with left round ligament. We concluded this anomaly as Unicornuate uterus, rudimentary horn with cavity, not communicating and left endometrial cyst (confirmed by histopathology result).

DISCUSSION

A unicornuate uterus with rudimentary horn occurs as a result of failure of complete development



Scheme 1. Enlarged left horn, tube and adnexa due to non communicating rudimentary horn



Scheme 2. Excision of left horn and left salpingoophorectomy was performed. The uterus was approximated to the round ligament.

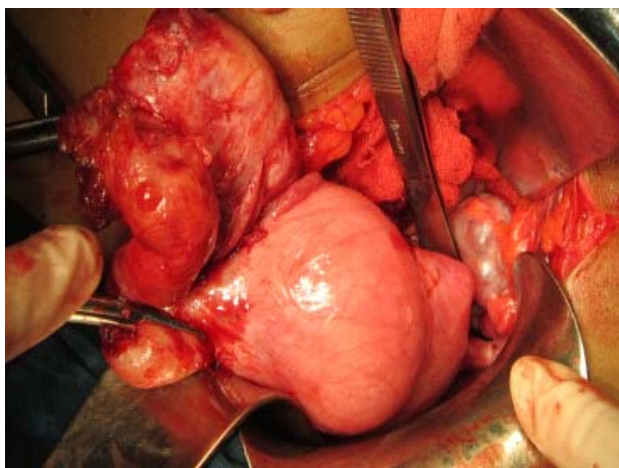


Figure 1. Normal right tube and ovary. Enlarged left horn, tube, and ovary

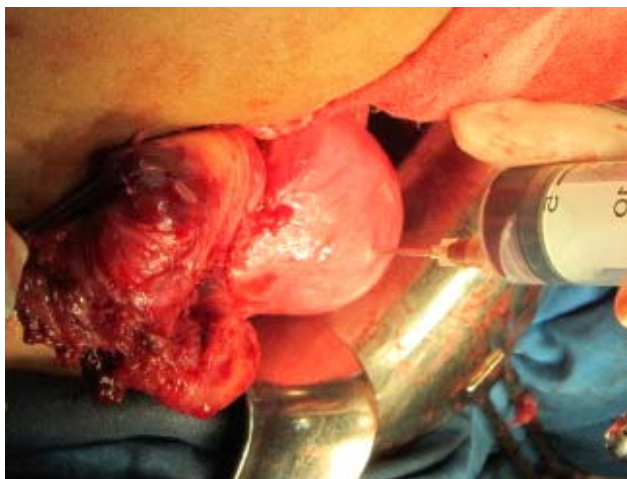


Figure 2. Aspiration of the left horn, came out brownish fluid correspond to retained blood

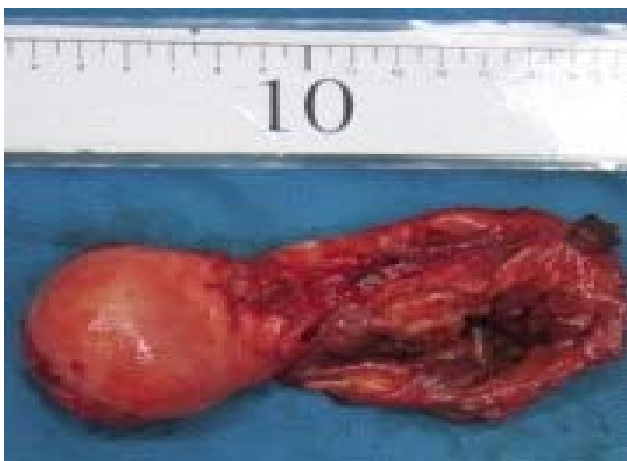


Figure 3. Enlarged left horn due to non communicating rudimentary horn



Figure 4. US examination revealed 2 uterine cavity; 1 cavity was enlarged due to retained blood

of one of the müllerian ducts and incomplete fusion with the contra lateral side. Unicornuate uterus may have either communicating or non-communi-



Figure 5. US examination revealed hematosalping and hematometra

cating rudimentary horn, absent rudimentary horn or a rudimentary horn with no cavity. A unicornuate uterus is found in 0.4% of reproductive age women and is associated with endometriosis in up to 40% of cases.^{2,4} Although there are many theories of the pathogenesis of endometriosis, a widely accepted theory is that of retrograde menstruation, postulated by Sampson. The pathophysiology of endometriosis in this case could be from retrograde menstruation. As we know, the rate of endometriosis will increase in patient with müllerian duct anomaly.² As in our case, endometriosis was found incidentally intraoperative.

The clinical manifestation is unspecified. If the horn is communicated with the normal cervical canal, there will be no symptoms. The asymptomatic communicated horn will be made complication like ectopic pregnancy, or acute abdomen due to rupture in the second trimester of pregnancy. Obstetric complication associated with unicornuate uteri are miscarriage in 37%, preterm birth in 16%, and term birth in only 45%.⁵ Cyclical pain will be felt if the horn is not communicated with the cervical. Like in this case, the patient had regular menstruation but felt cyclical pain as an effect from enlargement of the horn, tube, and adnexa.

Diagnostic modality is needed to make a diagnosis earlier. Uterine anomalies can be suspected or screened based on history taking, physical examination, hysterosalpingography and ultrasonographic evaluation. But diagnosis by these means is only succeeded in 14% of cases before these become symptomatic. In assessing a unicornuate uterus with HSG, blocked or non-communicating rudimentary horns will not be detected on film.

Reaching a diagnosis in the pregnancy will be even more difficult. In USG, gestational sac surrounded by myometrium is seen as separated from the uterus. The USG could also show a pseudo pattern of an asymmetrical bicornuate uterus, absent visual continuity in tissue surrounding the gestational sac, uterus and cervix, which all indicate rudimentary horn pregnancy. Sensitivity of USG is 26% and it decreases along with advancing pregnancy age. In most cases, diagnosis is missed by inexperienced hand. Three dimensional USG may be used to diagnose uterine abnormalities. In this case, the abnormality was found with 2D US examination. Before admission in this hospital, she had CT scan performed, which revealed a suspected ovarian cyst.

Laparoscopy is the most accurate modality to get the diagnosis. MRI is also proven to be a useful and noninvasive tool to detect uterine anomalies. Once the diagnosis is strongly suspected, these patients should be taken up for laparoscopy or laparotomy.⁶⁻¹¹

In review of literature, renal tract anomalies appeared to be found in 15-40% of the cases, including ectopic kidney, renal agenesis, double renal pelvis, horseshoe kidneys and unilateral medullary sponge kidney. MDA is also associated with skeletal anomalies like congenital fusion or absence of vertebra and it is seen in of 12-50% cases. An association of müllerian duct aplasia, renal aplasia and cervicothoracic somite dysplasia called MURCS has also been documented.^{12,13} In this case, no abnormality was found in other organs. The Visual analogue score after surgery was reduced until VAS 2.

CONCLUSION

Congenital abnormalities of the müllerian ducts are relatively common. Comprehensive examination can be diagnose this abnormality earlier and intervention can be given as soon as possible.

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