

HEMORRHAGIC SHOCK DUE TO INTRACOITAL VAGINAL RUPTURE IN CASES OF GONADAL DYSGENESIS (46 XX)

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We report on hemorrhagic shock due to intracoital vaginal rupture in a sporadic case of gonadal dysgenesis with uterine and bilateral oviduct aplasia (46 XX) verified during laparoscopy. Resuscitation with primary wound sutures was performed with full recovery.

Key words: vaginal rupture, hemorrhagic shock, gonadal dysgenesis, uterine and oviduct aplasia, ovarian hypoplasia

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INTRODUCTION

Life-threatening conditions in urgent adolescent gynecology, such as hemorrhagic shock, may be due to acute intra-abdominal hemorrhage with acute abdomen syndrome (ectopic pregnancy, corpus luteum hemorrhage) or spontaneous or violent external hemorrhage (metrorrhagia, intracoital rupture), and require surgical therapy with resuscitation (1).

Gonadal dysgenesis (GD) is one of the main causes of primary amenorrhea and is most commonly caused by Turner's or Swyer syndrome. Ovarian, gonadal dysgenesis is a disorder of sexual development. GD is characterized by incomplete or defective formation of the gonads due to either structural or numerical anomalies of sex chromosomes or mutations in the genes involved in the development of the gonad with complex clinical findings (2). müllerian duct anomalies are very rare in GD, such as partial or total genital aplasia or complex Rokitansky-Hauser syndrome, sporadically described in the literature (3,4).

We report on hemorrhagic shock due to intracoital vaginal rupture in a sporadic case of gonadal dysgenesis with uterine and oviduct aplasia (46 XX).

CASE REPORT

A 21-year-old amenorrheic patient was admitted to the emergency gynecologic department because of severe vaginal bleeding. Her medical history was unremarkable. Upon arrival, her clothes were covered with blood, as were her legs and abdomen. The patient was alert, with somnolence, hypotension (BP 90/50), tachycardia (140 /min), with massive arterial colporrhagia. Initial approach included intravenous fluids (1000 mL Ringer lactate and 500 mL colloid solution containing 10% hydroxyethyl starch) and tranexamic acid 2 g intravenously. She reported coitus two hours before and bleeding since then. She had never menstruated and had only one gynecologic examination in her life at the age of 20 years, which was suspicious of uterine agenesis. Gynecologic pelvic examination was impossible due to pelvic pain and patient distress. Emergency ultrasound examination did not show adnexa and uterus but showed some fluid in the pelvis. Physical examination showed no pubic and armpit hair and stage I mammary development on Tanner scale. She was 170 cm tall weighing 49 kg without any typical features of Turner's syndrome. Laboratory findings revealed acute severe secondary posthemorrhagic anemia (erythrocytes $1.2 \times 10^{12}/L$, hemoglobin 64 g/L, hematocrit 0.16 L/L) and normal coagulation parameters.

Considering heavy vaginal bleeding and hemorrhagic shock, we suggested urgent operation, which was accepted by the patient. Our initial diagnosis was intracoital vaginal rupture due to rudimentary vagina as part of Mayer-Küstner-Hauser-Rokitansky syndrome according to patient history and primary findings. After endotracheal anesthesia, we performed vaginal examination, which revealed rupture of the right vaginal wall 6x2 cm with arterial hemorrhage (Fig. 1).

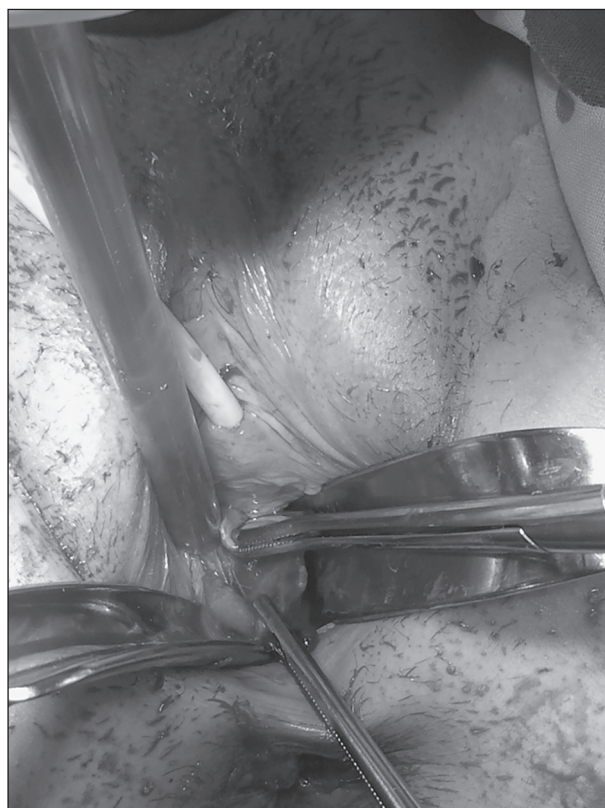


Fig. 1. Intracoital vaginal rupture.

Contrary to our expectation, vagina was normal in size (9 cm) and two transverse fingers in diameter. External genitals were normal. We placed sutures on vaginal wall, which stopped bleeding. After optimal hemostasis was achieved, laparoscopy revealed some blood in the pelvis, uterine aplasia, fallopian tube aplasia and hypoplastic ovaries (U5b C4 V0 by ESHRE classification) (5) (Fig. 2). During the operation, the patient received 2500 mL of crystalloid solution and 750 mL of erythrocyte concentrate.

After initial postoperative period, we obtained detailed patient medical history. She had never menstruated, was generally of good health. She had only one gynecologic examination but she had no follow up or recommended testing after her first visit. Her family history was inconspicuous. Her mother did not take any diethylstilbestrol during pregnancy nor received chemo- or radiotherapy. The patient did not suffer any

hearing loss or other body anomalies. Considering uterine agenesis with hypoplastic ovaries and normal vagina, we opted to do laboratory tests and peripheral blood karyotyping (46 XX). Hormonal profile showed hypergonadotropic hypogonadism (Table 1). We recommended hormone replacement therapy, supplementation with vitamin D and higher Ca intake for prevention of osteoporosis. We also recommended diagnostic biopsy of streak gonads, but the patient refused any further investigation and treatment. On the third postoperative day, she left the hospital healthy.

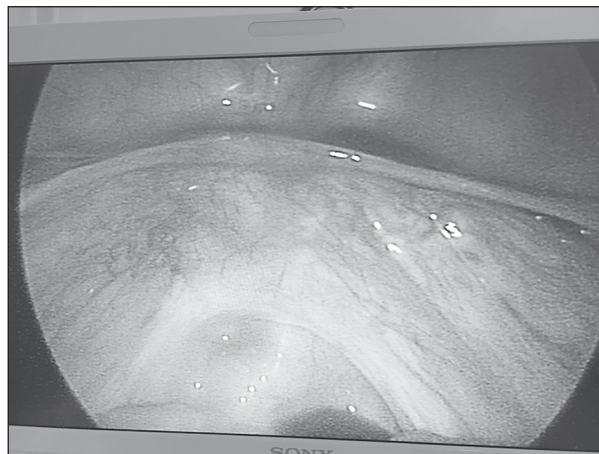


Fig. 2. Laparoscopic findings of uterine and bilateral oviduct aplasia.

Table 1. Postoperative laboratory findings

Estrogen	102 pmol/L	Low
Progesterone	1.31 nmol/L	Low
Prolactin	310 mIU/L	Normal
Testosterone	2.18 nmol/L	Normal
DHEA-S	11.1 μ mol	Elevated
FSH	46.42 U/L	Elevated
LH	13.06 U/L	Elevated

DHEA-S = dehydroepiandrosterone sulfate; FSH = follicle-stimulating hormone; LH = luteinizing hormone

DISCUSSION

Verified normal karyotype (46 XX) with hypergonadotropic hypogonadism and U5b C4 V0 classified anomaly indicated diagnosis of sporadic ovarian dysgenesis, also known as pure ovarian dysgenesis with primary amenorrhea and müllerian duct anomalies in our case (3,5). Anecdotally, intraoperative (for vital indications) findings verified uterine and fallopian tube aplasia with hypoplastic ovaries. The association of gonadal dysgenesis with aplastic or hypoplastic müllerian duct anomalies is extremely rare (3,4).

There are very few case reports of XX 46 gonadal dysgenesis and most of them are related to familial gonadal dysgenesis. According to Pertusa and Palacios, the most important issues are long-term lack of estrogen and sterility (6). Sterility in most cases remains unsolved issue. Lack of estrogen is preventable with hormone therapy, however, early osteoporosis remains an important issue. According to Ropke *et al.*, important part of further management is ovarian biopsy because ovarian tissue can have different karyotype than peripheral blood karyotype, thus carrying an increased risk of dysgerminoma (7). The highest risk of ovarian dysgerminoma is in 46 XY ovarian dysgenesis because of Y region. The risk of ovarian dysgerminoma in XX 46 ovarian dysgenesis is unknown and rarely described due to very few cases reported. To conclude, there is no evidence-based recommendation for early gonadectomy in 46 XX ovarian dysgenesis, however, a reasonable approach would be to perform ovarian biopsy and ultrasound follow up once a year (8,9).

Possible concomitant hypoplasia of other female genitalia can be the cause of intracoital rupture of vagina and hemorrhagic shock development, although in this case the vagina was of normal dimensions. A rare case of concomitant genital tract anomalies with ovarian dysgenesis is presented.

REFERENCES

1. Habek D, Karša M, Kalajžić T. Clinical evaluation of paediatric and adolescent gynecology in Clinical Hospital Osijek. *Gynaecol Perinatol* 2005; 14: 145-8.

2. McCann-Crosby B, Mansouri R, Dietrich JE *et al.* State of the art review in gonadal dysgenesis: challenges in diagnosis and management. *Int J Pediatr Endocrinol* 2014; 2014 (1): 4. doi: 10.1186/1687-9856-2014-4. Epub 2014 Apr 14.

3. Oppelt PG, Lermann J, Strick R *et al.* Malformations in a cohort of 284 women with Mayer-Rokitansky-Küster-Hauser syndrome (MRKH). *Reprod Biol Endocrinol* 2012; 57: 4-7.

4. Bousfiha N, Errarhay S, Saadi H, Ouldin K, Bouchikhi C, Banani C. Gonadal dysgenesis 46, XX associated with Mayer-Rokitansky-Küster-Hauser syndrome: one case report. *Obstet Gynecol Intern* 2010; 1-3. doi:10.1155/2010/847370

5. Di Spiezio Sardo A, Campo R, Gordts S *et al.* The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies. *Hum Reprod* 2015; 30(5): 1046-58.

6. Pertusa S, Palacios A. 46 XX pure gonadal dysgenesis: an infrequent cause of primary amenorrhoea. *BMJ Case Reports* 2009; doi: 10.1136/bcr.07.2008.0485

7. Ropke A, Pelz A, Volleth M *et al.* Sex chromosomal mosaicism in the gonads of patients with gonadal dysgenesis, but normal female or male karyotypes in lymphocytes. *Am J Obstet Gynecol* 2004; 190: 1059-62.

8. Zangen D, Kaufman Y, Zeligson S *et al.* XX ovarian dysgenesis is caused by a *PSMC3IP/HOP2* mutation that abolishes coactivation of estrogen-driven transcription. *Am J Hum Genet* 2011; 89(4): 572-9. doi: 10.1016/j.ajhg.2011.09.006

9. Kanagal DV, Prasad K, Rajesh A *et al.* Ovarian gonadoblastoma with dysgerminoma in a young girl with 46, XX karyotype: a case report. *J Clin Diagn Res* 2013; 7(9): 2021-2. doi: 10.7860/JCDR/2013/6412.3393

SAŽETAK

HEMORAGIČNI ŠOK ZBOG RAZDORA RODNICE U BOLESNICE S GONADALNOM DISGENEZOM
(46 XX)

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Prikazana je bolesnica s hemoragičnim šokom zbog intrakoitalnog razdora rodnice u rijetkom slučaju gonadalne disgeneze i maternične i obostrane aplazije jajovoda (46 XX), što je pronađeno tijekom laparoskopije. Učinjena je reanimacija i primarne šavi ozljede s potpunim oporavkom.

Ključne riječi: razdor rodnice, hemoragični šok, gonadalna disgeneza, aplazija maternice i jajovoda, hipoplazija jajnika