Heterotopic pregnancy in the absence of risk factors – diagnostics difficulties

Ciąża heterotopowa u pacjentki bez czynników ryzyka – trudności diagnostyczne

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Abstract

Heterotopic pregnancy (HP) is a rare entity occurring in 1:30000 pregnancies but its incidence raises as a result of assisted reproductive techniques (ART) to 1:100-1:500.

The aim of this report was to present a rare case of simultaneous intrauterine and extraterine tubal pregnancy in a natural conception cycle. The ectopic pregnancy was not diagnosed in the first trimester scan and at 14 and 16 weeks of gestation led to severe hemoperitoneum and two exploratory laparotomies.

Diagnostic problems were discussed and literature was reviewed.

Key words: ectopic pregnancy / heterotopic pregnancy / diagnostic difficulties /

Streszczenie

Ciąża heterotopowa występuje z częstością 1:30000 ciąży, ale częstość jej wzrasta, w konsekwencji stosowania technik wspomaganych rozrodu, do 1:100-1:500.

Celem tej pracy jest opis rzadkiego przypadku ciąży jajowodowej współistniejącej z ciązą wewnątrzmaciczną. Ciąża ektopowa nie była zdiagnozowana w pierwszym trymestrze ciąży a w 14 i 16 tygodniu ciąży dwukrotnie doprowadziła do laparotomii z powodu obfitego krwawienia do jamy brzusznej.

W pracy przedstawiono problemy diagnostyczne oraz przegląd aktualnej literatury.

Słowa kluczowe: ciąża ektopowa / ciąża heterotopowa / trudności diagnostyczne /

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Introduction

Heterotopic pregnancy is defined as the presence of multiple gestations, one in the uterine cavity and the other outside the uterus, commonly in the fallopian tube (95-97% of all ectopic pregnancies), especially in the ampulla portion of the tube, where over 55-80% of the cases are located, the isthmus (20-25%), and seldom, the infundibulum and fimbra (17%). Intertstitial implantation accounts for 2-4% of all tubal pregnancies (1, 2). The most common site is tubal pregnancy and the most uncommon one is the cervix or the ovary (3). Heterotopic pregnancies can pose a diagnostic dilemma because an early transvaginal ultrasound may not diagnose an ex-utero gestation in all cases. The diagnosis of a pseudosac should be made with caution, as even in the presence of a pseudo sac there can be a high false positive diagnosis of an ectopic pregnancy (4). Sometimes the presence of a hemorrhagic corpus luteum can confuse and delay the diagnosis of a heterotopic pregnancy (5).

The detection rate of heterotopic pregnancy can vary from 41 to 84% with transvaginal ultrasound scans (5, 6). It is influenced by factors such as routine and easy access to transvaginal ultrasound scans for high-risk patients with a history of previous ectopic pregnancy and those who received fertility treatment.

With the increase in assisted conception techniques (ART), the likelihood of detecting heterotopic pregnancy will increase but misdiagnosis or delayed diagnosis of spontaneous heterotopic pregnancy remain a diagnostic dilemma and a challenge for gynecologists.

Case report

A 34-year-old multigravida at 14 weeks gestation was admitted to the surgical emergency department due to abdominal pain, emesis, diarrhea and sudden loss of consciousness. There was no history of abortion, infertility, pelvic inflammatory disease or abdominal surgery. At the age of 17 the patient underwent cardiosurgery of atrial septal defect. Upon admission her heart rate was 95/min, blood pressure was (RR) 110/60. Physical examination was suggestive of acute abdomen, tenderness in the lower abdomen and signs of guarding. Laboratory data on admission revealed white blood cells 6,32 K/µl, hematocrit 28,8%, hemoglobin 9,6g/dl, platelets 232 K/µl. Transvaginal ultrasound examination (TVS) revealed in uterus a fetus with detectable heartbeat and CRL 80 mm (14 weeks of pregnancy), placenta on the posterior wall of the uterus. Adnexa were poorly visualized. The abdominal ultrasound revealed free peritoneal fluid surrounding the spleen (5-9 cm), and the right hypogastrium (4-9cm). The patient was moved to the operating room for an emergency exploratory laparotomy to control the source of bleeding under general anesthesia through a subumbilical incision. Laparotomy revealed 1 liter of blood with clots, which was evacuated from the free peritoneal cavity. Fourteen weeks gravid uterus, ovaries and fallopian tubes were normal. No source of bleeding was found. Removal of hemoperitoneum and peritoneal lavage was performed. After five days of observation the patient was discharged home in a good condition. At 16 weeks gestation the patient was again admitted to the Gynecology and Obstetrics Department of the same hospital, because of sudden pain in the lower abdomen. In gynecological examination a brownish discharge from vagina was present. Patient was admitted for a close follow-up. The initial management was conservative. The patient’s status was stable (RR 130/80, hearth rate was 72/min). Laboratory results revealed signs of anemia on three consecutive days: hemoglobin 9,1 to 6,5g/dl, hematocrit 27,6 to 20,5%. Abdominal ultrasound examination showed presence of free peritoneal fluid and a viable intrauterine pregnancy, adnexa were hard to visualize. The patient was qualified for the second exploratory laparotomy to control the source of bleeding under general anesthesia. One liter of blood with clots was evacuated from peritoneum. The size of the gravid uterus corresponded to 16 weeks gestation and the ovaries looked normal. The right fallopian tube was wide, cyanotic and bleeding. A partial right salpingectomy was performed. Two units of blood after surgery were transfused, peritoneal lavage and drainage were performed. Patient recovered uneventfully and was discharged from the hospital within 4 days with viable intrauterine pregnancy which proceeded without complications and was delivered spontaneously at term (fetal weight 3990, Apgar score 10). Histopathology of the salpingectomy specimen confirmed chorionic villi suggestive of an ectopic pregnancy.

Discussion

Heterotopic pregnancies (HP) are diagnosed in the presence of one or more intrauterine pregnancies coexisting with an ectopic one i.e. tubal, ovarian, cervical, cornual or abdominal. It is a very rare condition in natural conception cycles and can be easily overlooked (7, 8, 9, 1, 4, 5). The risk factors for HP are the same as for ectopic pregnancy i.e. tubal damage after pelvic inflammatory disease, endometriosis, tubal sterilization, and tubal infertility or tubal reconstructive surgery, uterus malformation, use of intrauterine devices, progestrone only contraceptive pills, and assisted reproductive techniques ART (10, 11). The possible explanation for this complication is that the transferred embryos which migrate into the damaged tubes are not expelled by peristaltic movements (12, 13, 14, 15). Early diagnosis of HP is difficult due to lack of symptoms. There are four most common symptoms defined by Reece et al.: abdominal pain, adnexal mass, peritoneal irritation and enlarged uterus (16). In some reports in HP the abdominal pain was present in 83% of cases, 13% had hypovolemic shock and abdominal tenderness, half of them experienced vaginal bleeding (14).

Measurement of serum beta human chorionic gonadotropin (beta hCG) is the most helpful in diagnosing ectopic pregnancy or pregnancy of unknown location. In our case there was no reason to measure beta hCG due to an advanced age of confirmed intrauterine pregnancy and severity of patient status upon hospitalization. In case of HP, the intrauterine placenta’s beta hCG production can mask the ectopic one and its use in such cases is debatable, because it can lead to false assurance (3). Although there was a case of heterotopic pregnancy misdiagnosed as only ectopic described by Ludwig et al (7). They noticed an increase in beta hCG level on the first postoperative day after salpingectomy. Pregnancy was confirmed histopathologically, what gives a clue to measure beta hCG after procedure, even if ultrasound does not confirm the presence of an intrauterine gestational sac.

HP presents a diagnostic challenge because TVS procedure in early pregnancy may not diagnose an ex-utero gestation in all cases (4). Sometimes the presence of hemorrhagic corpus luteum can confuse and delay the diagnosis of HP (17). The detection rate of HP with TVS can vary from 41 to 84% (18).
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Literature review from 1971 to 1993 revealed 112 cases of HP, 46 diagnosed by an ultrasound and 66 diagnosed only by laparoscopy or laparotomy (6). Similar review from 1994 to 2004 showed that out of 80 HP cases, 21 were diagnosed by ultrasound and 59 at surgery. This leads to a conclusion that USG did not change its diagnostic ability over a period of time. One of the reasons for this unexpected observation is that HP is a rare condition and most patients with HP present to the emergency department with symptoms of a rupture of ectopic component. Thus, a preoperative diagnosis of HP remains a challenge (10).

Women who experienced ectopic pregnancy, pelvis inflammatory disease or abdominal surgery may be at higher risk and should be scanned in early pregnancy to confirm its location. Also, caution is necessary in case of low-risk, symptomatic women with abdominal or pelvic pain in which ultrasound findings present intrauterine gestation sac while free fluid is noted in the pelvis with or without adnexal excessive mass, because they also might be suspected of ectopic pregnancy (3).

There are numerous reasons why ectopic pregnancy may fail to be visualized on TVS, including poor quality of the ultrasound equipment or technique, an inexperienced ultrasound operator, increased maternal body mass index or the presence of uterine fibroids or ovarian pathology making visualization of the adnexa difficult (11).

After an ectopic pregnancy a woman should be informed that there is a 7 to 13 fold increase in the risk of subsequent ectopic pregnancy (19). The chance that the subsequent pregnancy will be intrauterine is 50% to 80%, and the chance of a subsequent tubal pregnancy is 10% to 25%, and the remaining patients (2-5%) may become infertile (11). The first-line option treatment for HP is surgery by laparoscopy or laparotomy. In our case we preferred laparotomy because of severe internal bleeding. Another possible way of treatment of ectopic pregnancy is injection of potassium chloride into the ovum, but in HP pregnancy this way of treatment as well as Methotrexate are not recommended, because of the second fetus. Survival rate of intrauterine pregnancy is 60,9% for surgery and 50% for potassium chloride injections (20, 22), although after this kind of treatment some cases will also require a surgery. The risk for surgery is 13% to 50%, respectively (20, 22).

Factors such as maternal hemodynamic status, fetal congenital abnormality, fetal viability, gestational age at presentation, and the availability of neonatal facilities should be considered when managing a heterotopic pregnancy (21).

Conclusion

HP can occur in the absence of any predisposing risk factors. The presence of intrauterine pregnancy does not exclude the possibility of simultaneous existence of an ectopic pregnancy. All pregnant women with intrauterine pregnancy should have a complete ultrasonographic examination of the pelvis, especially the adnexa, to exclude the presence of an ectopic pregnancy because even now a heterotopic pregnancy remains to be a life-threatening diagnostic challenge due to its rarity and atypical presentation.

References