Primary ruptured ovarian pregnancy in a spontaneous conception cycle: a case report and review of the literature

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Summary

Ovarian pregnancy is an uncommon presentation of ectopic gestation, where the gestational sac is implanted within the ovary. Usually, it ends with rupture, which occurs before the end of the first trimester. Its presentation often is difficult to distinguish from that of tubal ectopic pregnancy and hemorrhagic ovarian cyst. We describe a case of primary ovarian pregnancy in a 31-year-old patient who presented to the emergency room with symptoms and signs of peritonism and positive urine hCG test. The gestation sac was demonstrated in the right ovary by transvaginal sonography. MSD (mean sac diameter) was 15 mm corresponding to the sixth gestational week. Free fluid was found in the Douglas pouch. Culdocentesis was positive for hemoperitoneum. Henceforth, emergency laparotomy and wedge resection of the ovary was perfomed. Aetiological, clinical and therapeutical aspects of this rare extrauterine pregnancy are described. Also, the problems of its differential diagnosis are discussed.

Key words: Ovarian pregnancy; Abdominal pregnancy; Ectopic pregnancy; Sonography.

Introduction

Primary ovarian pregnancy is the implantation of the gestational sac in the ovary and represents one of the rarest forms of extrauterine pregnancy accounting for less than 3% of all ectopic gestations [1, 2]. Since the first recorded instance by Saint Maurice in 1682 (quoted in Ricci) [3], a number of case reports and a series of literature reviews have appeared.

It is difficult to make a preoperative diagnosis of ovarian pregnancy with certainty. Clinical signs and symptoms suggestive of ovarian pregnancy, are for example abdominal pain, amenorrhea, and abnormal vaginal bleeding [4], which are similar to those found in tubal pregnancies [5, 6, 7] and in ruptured hemorrhagic corpus luteum cysts [6]. However, because of the increased vascularity of the ovarian tissue, ovarian pregnancy often causes maternal hemorrhage early in the first trimester, which disrupts the pregnancy and usually ruptures the ovary with massive hemoperitoneum that requires emergency surgery [1, 8]. The suggestive initial diagnosis is made on the operating table and the final diagnosis is confirmed only by histopathology.

In 1878 Spiegelberg [9] established four pathological criteria for the diagnosis of ovarian pregnancy: 1) The ipsilateral tube is intact and separate from the ovary. 2) The gestational sac is located in the position of the ovary. 3) Ovary and sac are connected to the uterus by the utero-ovarian ligament. 4) Definite ovarian tissue is identified in the wall of gestational sac.

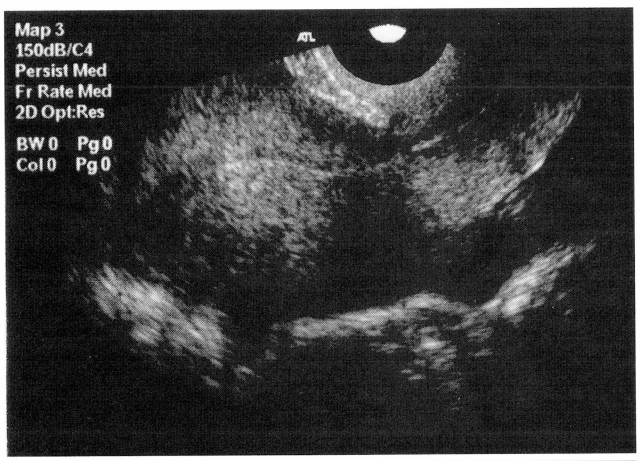
Here, we report a very rare case of primary ovarian pregnancy in a natural conception cycle, in which the diagnosis was made only when signs and symptoms of massive hemoperitoneum occurred and emergency laparotomy was mandatory.

Revised manuscript accepted for publication March 25, 2002

Case report

A 31-year-old woman presented with acute abdominal symptoms on arrival at the emergency room of the "George Gennimatas" General State Hospital of Athens. She had an obstetrical history of cesarian section for a twin pregnancy after IVF four years earlier. Her medical history included a laparoscopic cholocystectomy one year earlier. There was no history of possible chlamydial infection or present use of an intrauterine device (IUD). She had menstruation every 28 days. The last menstrual cycle was 13 days before. Five hours before admission to our hospital she had visited a private diagnostic center because of low abdominal pain for six hours. Abdominal sonography showed an anteverted uterus measuring 63 x 32 x 52 mm with an endometrial thickness of 3.8 mm and regular right and left adnexa, each measuring 25 x 22 mm. No intraperitoneal fluid was reported. The hematological examination revealed: hematocrit 35.4%, hemoglobin 11.9g/dl, platelet count 198 x 10^9/l, leukocyte count 8.5 x 10^9/l.

In our emergency room abdominal examination revealed a diffused lower abdominal tenderness and rebound mainly in the right lower quadrant; her abdomen showed muscular resistance; the urine hCG was positive, but she was not aware of her pregnancy. Pelvic examination revealed an anteverted, slightly enlarged tender uterus; the right adnexa was tender and palpable; the palpation of the posterior pouch of Douglas was very painful. Cervical movement during the bimanual examination was painful; no cervical bleeding was present. Culdocentesis was performed and hemoperitoneum with clots was found. The presence of ruptured ectopic pregnancy was considered. However, blood pressure and heart rate of the patient were still stable. Therefore, a transvaginal sonography was done, which revealed the absence of an intrauterine gestational sac and endometrium of normal thickness. The posterior border of the uterus was ill defined, because of the presence of echogenic fluid in the cul-de-sac, suggesting hemorrhage (Figure 1). Fluid with fine echoes was also depicted in the vesicouterine pouch (Figure 2). A thick-walled ring-like cystic formation with a peripheral solid mass was found in the anatomical site of the right ovary. Color Doppler ultrasound revealed a remarkable



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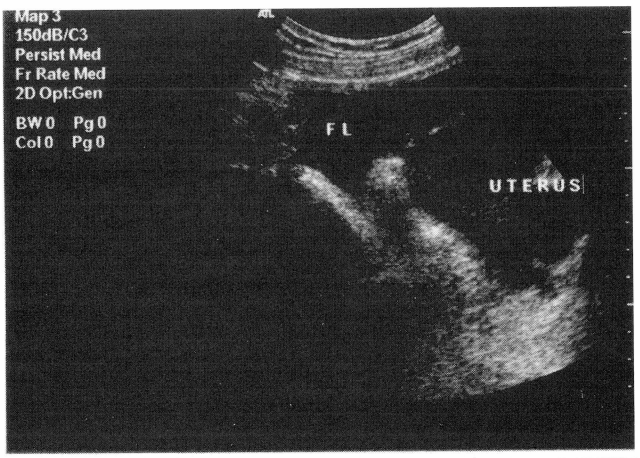

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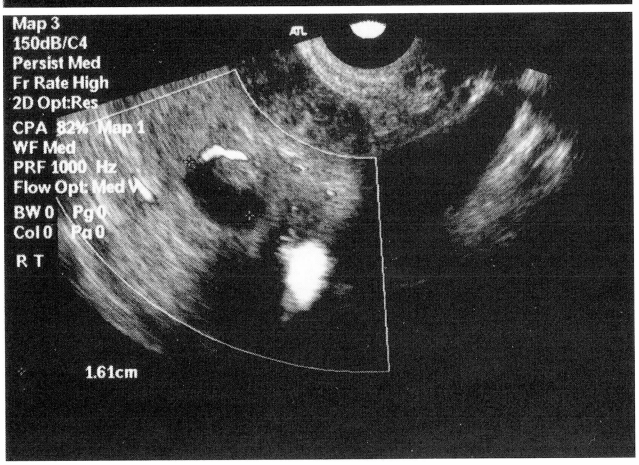

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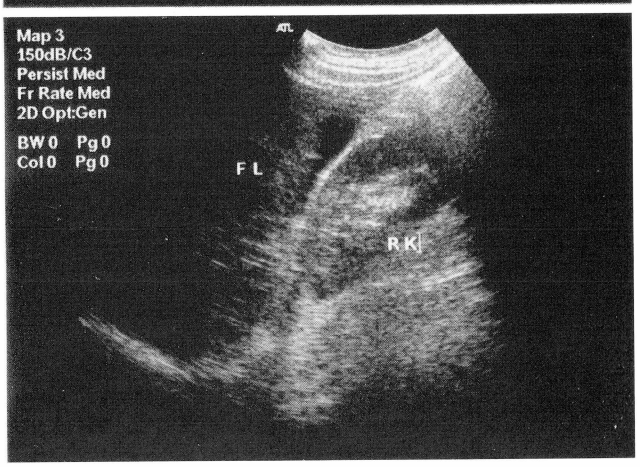

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Figure 1. — No intrauterine gestation sac was revealed by transvaginal ultrasonography (longitudinal section). Echogenic fluid fills the posterior pouch, suggesting hemorrhage.
Figure 2. — Uterus surrounded by fluid with fine echoes.
Figure 3. — Demonstration of the gestational sac in the right ovary; MSD (mean sac diameter) is 15 mm corresponding to the 6th gestational week. Peripheral, ring-like, flow was depicted.
Figure 4. — Free fluid in Morisson's space demonstrated by abdominal ultrasonography.

peritrophoblastic type of flow and low resistance arterial flow (Figure 3). Free fluid in Morisson's space was demonstrated by abdominal ultrasound scan (Figure 4). These findings, as well as the clinical history of the patient were highly indicative of a ruptured ectopic pregnancy of the ovary. No pulses were depicted within the solid mass and the longitudinal diameter of the cystic formation representing MSD (16 mm) corresponded to the sixth gestational week. The preoperative hematological data showed: hematocrit 33.3%, hemoglobin 11.0g/dl, platelet count 186 x 10^9/l, leukocyte count 8.4 x 10^9/l. The preoperative biochemical tests were within normal rates.

An emergency exploratory laparotomy was processed by the gynecological team of our hospital, with a midline subumbilical incision, which revealed 1.8 of free blood in the abdominal cavity. The volume of the right ovary was increased compared with this of the left ovary. The cortex of the right ovary was disrupted by a hemorrhagic mass, which was bleeding. The left ovary and both tubes were normal in appearance. The picture of the right ovary resembled a ruptured hemorrhagic corpus luteum. However, the macroscopic appearance of the right ovary, the normal uterine tubes, the positive pregnancy urine test, and the ultrasound picture in our hospital, were suggestive of primary ovarian pregnancy. A wedge resection was done in the right ovary. In addition, a bilateral salpingectomy was performed for sterilization, because the couple gave preoperative consent for this. The upper and lower abdomen were washed with normal saline.

The specimen was 3.2 x 1.5 x 1cm in size and on cross-section the hemorrhagic mass was on the periphery of the corpus luteum. The excision of the corpus luteum reassured us because in case of an unfounded abdominal pregnancy, it should regress by removal of the corpus luteum.

Postoperatively, the patient recovered uneventfully except for a transfusion of two units of packed red blood cells on the day of operation. Her serial serum beta-hCG titers were 1460 m IU/ml, 1013 m IU/ml, 572 m IU/ml, 216 m IU/ml the 1st, 2nd, 3rd and 6th postoperative days respectively. The fall of serum beta-hCG postoperatively confirmed the regression of pregnancy. The removal of the corpus luteum caused menstruation 36 hours after surgery.

The patient was discharged on the seventh postoperative day with hematocrit 31.7%, hemoglobin 10.6g/dl, platelet count 187 x 10^9/l, leukocyte count 4,6 x 10^9/l. The histological findings confirmed the presence of primary ovarian ectopic pregnancy. On follow-up three months later the patient was in good condition; a repeat serum beta-hCG showed undetectable titers. She menstruated every 28 days after surgery and a transvaginal ultrasound demonstrated both ovaries normal.

Discussion

The incidence of primary ovarian pregnancy seems to be increasing [10] from a rate of one in 40,000 pregnancies in 1950 [11] to a rate of one in 7,000 pregnancies in 1983 [7]. However, the chance of ovarian pregnancy recurring is not likely, as there are no repeat ovarian pregnancies in the world literature [8].

Some authors have suggested that the increased incidence of ovarian pregnancy is caused by the widespread use of the IUD [6, 12-16]. According to Lehfeldt's study [11], the IUD is effective in preventing intrauterine pregnancy in 99.5% and tubal pregnancy in 95%, but it has little effect on ovarian pregnancy. Among reported cases of ovarian pregnancy, the rate of concurrent use of an IUD was 17 to 25% [7].

It is important to consider whether pre-existing pathology in the reproductive tract produces environmental conditions that interfere with normal zygote transport and favors an ovarian pregnancy [7]. Indeed, it has been found that the incidence of ovarian pregnancy is increased in women with a history of pelvic inflammatory disease (PID) [7]. Matseoane [17] and Grimes et al [7] noted prior history of pelvic inflammatory disease in 42 and 46% of ectopic pregnancies, respectively. It may be due to the fact that PID can induce either a reduction of tubal motility, or a thickening of the ovarian albuginea secondary to an inflammatory response, thus causing an increased risk of intrafollicular pregnancy due to a hampered follicular dehiscence [18]. In our case, the patient had an obstetrical history of cesarian section for a twin pregnancy after IVF four years earlier. It is possible that the prior surgical procedure on the pelvis may also apply for the development of ovarian pregnancy.

Also, it is important to consider if primary ovarian pregnancy after the use of assisted reproductive technologies occurs with greater frequency. Spontaneous ectopic pregnancy is estimated to occur in about 0.08% of all pregnancies [19], while it is a relatively common complication of assisted reproductive technology cycles. Approximately 5% of pregnancies achieved after IVF are ectopics [20]. Marcus and Brinsden [2] reported a 6% incidence of primary ovarian pregnancies of all extrauterine pregnancies after IVF and embryo trasfer (ET) or 0.3% of all pregnancies achieved after IVF and ET. Based on this data, the occurrence of ovarian pregnancy after the use of assisted reproductive technology seems to be increased.

Several mechanisms have been suggested to explain ovarian implantation. One theory holds that fertilization occurs normally and implantation of the ovary follows reflux of the conceptus from the tube along with blood from the uterus [7]. Another theory provokes defective ovum release at ovulation, with fertilization occurring within the follicle [8]. The most likely aetiological mechanism of ovarian pregnancy after IVF is the reverse migration of an embryo towards the fallopian tube and implantation in the ovary due to the volume of culture medium injected at embryo trasfer or to the position of the patient. Finally, it is possible that ovarian pregnancy may result from in vivo fertilization of unrecovered oocytes if coitus occurs near the time of oocyte recovery. For this reason, patients are instructed to avoid intercourse at that time [2].

It is difficult to make a preoperative diagnosis of ovarian pregnancy with certainty. In the present study the positive urine hCG test and the symptoms and signs of peritonism raised our suspicion of a ruptured ectopic pregnancy, which was subsequently proved by a positive culdocentesis. Also, the transvaginal ultrasound was suggestive of a ruptured ovarian pregnancy, while the abdominal ultrasound scan showed free fluid in Morisson's space suggestive of hemoperitoneum. It has been found that when hemoperitoneum is present in a woman with a positive pregnancy test, ectopic gestations will exist in 85 to 97% of instances [21, 22]. In the present study the patient on admission was stable regarding blood pressure and heart rate, and although the culdocentesis was positive for hemoperitoneum, we additionally performed a transvaginal ultrasound scan to exclude the rare possibility of heterotopic pregnancy. In such cases of simultaneous intrauterine and extrauterine pregnancy, the surgical treatment with minimal manipulation of the uterus is mandatory in order for the intrauterine pregnancy to proceed without complications. Our transvaginal ultrasound demonstrated the gestational sac in the right ovary. The MSD was 15 mm corresponding to the sixth gestational week. Sonographically, the appearance of ovarian pregnancies varies as widely as that seen in tubal pregnancies. Reported cases include a cystic mass containing a partially solid area, a complex adnexal mass associated with free fluid, a definite gestational ring in the adnexa and a cystic adnexal mass containing echoes [4].

In regard to the mortality of ectopic pregnancy, according to Barder [23], it is the fifth leading cause of maternal death, where as, Gerbie [24] noted that it accounts for 6% of maternal deaths in the United States. Moreover, an ovarian pregnancy is by far more dangerous than a tubal pregnancy [25]. In our case, the patient presented at the emergency department of our hospital with massive hemoperitoneum, although her blood pressure and heart rate were stable. Five hours before her admission she had visited a private diagnostic center because of low abdominal pain lasting for six hours but the report of the abdominal ultrasound scan was negative. A pregnancy test was not performed and the patient went home. We suggest that all the patients presenting with low abdominal pain, amenorrhea or vaginal spotting should initially receive a pregnancy test. It seems that an abdominal ultrasound scan is not very helpful for early detection of an ovarian pregnancy. We should keep in mind, in cases of ovarian pregnancy, that the vascularity of the ovaries results in a more massive hemoperitoneum at an earlier stage and can be very threatening for the patient's life. In a series of studies by Raziel *et al.* [1], 30% of cases had circulatory collapse, while 35% needed blood transfusions. In this report 90% of the ovarian pregnancies occurred in users of intrauterine contraceptive devices (IUD) and this high rate of circulatory collapse might be associated with late diagnosis due to the use of IUDS,

where a delay in the diagnosis is possible. Ovarian pregnancies after IVF and ET are usually diagnosed when unruptured because of the close follow-up of the patients. In a series by Marcus and Brinsden [2], of the ovarian pregnancies after IVF and ET 50% presented with abdominal pain with or without vaginal discharge or bleeding. In the remaining cases, the ovarian pregnancies were asymptomatic at the time of diagnosis. The diagnosis of ectopic pregnancy with transvaginal ultrasound scan was correctly made on all the described cases and in five out of seven the diagnosis of ovarian gestation was suspected.

The treatment of an ovarian pregnancy includes ipsilateral oophorectomy, cystectomy or wedge ovarian resection, at both laparotomy and laparoscopy [26]. Other alternatives for the treatment of unruptured ovarian pregnancies include the use of prostaglandin or methotrexate. Koite *et al.* in 1990 [27] reported the first successful case of an unruptured ovarian pregnancy treated medically by local injection of 0.5 mg prostaglandin F_{2a} with an additional dose of 1.5 mg of prostaglandin E_2 (dinoprostone) administered orally for 14 postoperative days. Also, two years later, Shamma and Schwartz [28] reported for the first time the successful treatment of an unruptured ovarian pregnancy by intramuscular injection of 72 mg (50mg/m^2) methotrexate in two divided doses. These conservative approaches to treatment should be important for patients who desire future childbearing because possible surgical complications such as hemorrhage, ovariectomy or subsequent pelvic adhesive disease are prevented. However, the conservative treatment of ovarian pregnancy is questionable because histologic proof is mandatory for the diagnosis of primary ovarian pregnancy according to the old but very relevant Spiegelberg criteria [25]. Theoretically, persistent trophoblastic tissue may be present after a wedge resection, as occasionally seen in patients with salpingostomy after conservative tubal ectopic surgical treatment [26]. In these cases adjuvant therapy with methotrexate may be useful. In our case, the ovarian pregnancy surgically resembled a ruptured hemorrhagic corpus luteum. We performed a wedge resection in the right ovary and subsequent serial measurement of beta-hCG titers demonstrated a gradual decline to 0, indicating the absence of persistent trophoblastic tissue. In addition, in our case, the ovarian pregnancy was found on the periphery of the corpus luteum and as is known, excision of the corpus luteum in early pregnancy results in regression of any persistent trophoblastic tissue. Gross and microscopic criteria for diagnosis of an ovarian pregnancy as formulated by Spiegelberg in 1878 were confirmed in the present case report.

In conclusion, we presented a very rare case of primary ovarian pregnancy in a natural conception cycle, where the patient presented to our emergency department with symptoms and signs of peritonism. Due to the highly vascular structure of the ovary, the potential for massive hemoperitoneum in cases of ovarian pregnancy is very high. Obstetrician-gynecologists should keep in mind this case of unusual pathology of pregnancy and intervene early surgically, before it ruptures and becomes threatening for the patient's life. Conservative surgical treatment is feasible and should be attempted in young patients and those desiring future childbearing.

Acknowledgement

The authors would like to thank colleagues in the Pathology Department for the microscopic examination of the surgical specimen.

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