Clinical history: 34-year-old G2P0010 asymptomatic female with positive pregnancy test presents for routine early obstetric ultrasound examination. According to her last menstrual period, her gestational age is 12 weeks 2 days.

Figure 1. Transvaginal sagittal ultrasound image of the uterus demonstrates no intrauterine gestational sac.
Figure 2. Transvaginal ultrasound image demonstrates a gestational sac between the right ovary (OV) and the uterus (UT).
Figure 3. Transvaginal M-mode demonstrates fetal cardiac activity at 190 beats per minute.

Figure 4. Transvaginal ultrasound image of the fetus. Crown rump length measures 44 mm, corresponding to a gestational age of 11 weeks 1 day.

Figure 5. Transvaginal ultrasound image of the right adnexa demonstrating a second gestational sac (calipers) with internal echoes adjacent to the gestational sac with
the 11 week size fetus (figure 3). There is a thick membrane between the sacs corresponding to a diamniotic dichorionic twin pregnancy. Note the twin peak sign.

Figure 6. Pathology gross specimen demonstrates two fetuses with size discordance. The placenta and umbilical cord for fetus A are visible.

Imaging diagnosis: Twin ectopic pregnancy. Live fetus A measures 1 week size less than dates. Fetus B not visualized. Twin peak sign suggests dichorionic diamniotic gestation.

Operative diagnosis: Right-sided 5-cm intact cornual ectopic pregnancy. The patient underwent right salpingectomy and oversewing of the right cornu.
Pathology diagnosis: Twin ectopic pregnancy. Morphologically unremarkable male fetus A with crown rump length of 4.9 cm. "Macerated" fetus B with crown rump length of 2 cm.

Final diagnosis: Cornual ectopic pregnancy of diamniotic dichorionic twins, with one live fetus and one co-twin demise.

Discussion:

Ectopic pregnancy occurs in 1.4% of pregnancies. 95% of ectopic pregnancies implant in the ampulla or isthmus of the fallopian tube. The second most common site of implantation, the intramural portion of the fallopian tube (interstitial or cornual pregnancy), accounts for 2-3% of ectopic pregnancies. The small remainder of ectopic pregnancies implant in the cervix, ovary, or abdominal cavity [1]. The mortality rate for ectopic pregnancy is currently estimated to be less than 1/1,000 pregnancies. The mortality rate for interstitial pregnancies is twice that of tubal pregnancies, secondary to later rupture, usually in the second trimester. Adjacent myometrium allows for prolonged development and increased myometrial vascular proliferation, resulting in more severe hemorrhage after rupture [1]. Risk factors for ectopic pregnancy include prior ectopic pregnancy, tubal or uterine abnormality, history of tubal surgery or cesarean section, pelvic inflammatory disease, intrauterine contraceptive device, age, and ovulation induction/in vitro fertilization [1,2,3].

Twin ectopic pregnancy is rare in spontaneous pregnancies. As of 2006, just over 100 cases total and fewer than 10 preoperatively diagnosed cases had been reported. Risk factors for ectopic pregnancy also increase risk for twinning, including delayed transit time through the fallopian tube and in vitro fertilization [6].

Clinical presentation of ectopic pregnancy, including interstitial pregnancy, is non-specific and overlaps with other pelvic pathology. Symptoms may include pelvic pain, adnexal or cervical motion tenderness, vaginal bleeding, and palpable adnexal mass [1]. Patients may also be asymptomatic prior to rupture.

Transvaginal sonography is more sensitive for diagnosis of ectopic pregnancy than transabdominal sonography, partly because transvaginal sonography is more sensitive for the presence of an intrauterine gestational sac [1]. Classic sonographic findings of ectopic pregnancy are non-specific and include absence of an intrauterine gestational sac or pseudosac within the endometrial cavity, complex adnexal mass, and free fluid in the pelvis. The only sonographic finding which is 100% specific for ectopic pregnancy is cardiac activity within an adnexal mass [1]. In cases of uncertainty, if the patient is clinically stable, serial evaluation of serum beta-hCG can be helpful.
The imaging criteria for interstitial ectopic pregnancy include eccentrically located gestational sac, cornual mass, absence of the double decidual sac sign, thinning of the myometrial mantle (less than 5 mm), and the interstitial line sign [3,4]. The interstitial line sign is an echogenic line extending toward a sac or mass in the cornual region. This line is thought to represent either endometrium or the interstitial portion of the fallopian tube. The interstitial line sign is the most sensitive and specific ultrasound finding for interstitial pregnancy. One study estimates the sensitivity and specificity to be 80% and 98%, respectively [4]. In comparison, the sensitivity and specificity of adjacent myometrial thinning are reportedly 40% and 93%, respectively, and the sensitivity and specificity of an eccentrically located gestational sac are 40% and 88%, respectively [4]. Three-dimensional ultrasonography may provide increased sensitivity due to better visualization of the structure of the endometrial cavity [5]; however, this is not currently the standard of care and has not been definitively studied. Early in gestation, a normal intrauterine gestational sac located superolaterally within the endometrial cavity may be mistaken for an interstitial pregnancy. In cases of uncertainty, close sonographic follow-up will demonstrate eventual extension of a normal sac into the endometrial cavity, whereas an interstitial pregnancy will remain eccentric [1].

Treatment options for interstitial ectopic pregnancy include surgical resection, methotrexate, and uterine artery embolization [2]. Since interstitial ectopic pregnancies tend to present later than tubal pregnancies, surgical management is most common.

Patients with a history of interstitial pregnancy are at increased risk for uterine rupture with future pregnancies due to myometrial thinning in the region of the previous pregnancy. [2]

References: