Pregnancy at an unusual location: A case report with review of literature

Manjula Reddy H1*, Germeen Joseph2
1Department of Obstetrics and Gynaecology, 2Deparment of Radiology, Gulf Medical College Hospital and Research Centre, Ajman, UAE

*Presenting author

ABSTRACT
Implantation of the embryo at the site of a previous Caesarean scar is the rarest form of ectopic pregnancy, with a high risk of maternal complications. The incidence of CSP (caesarean scar pregnancy) is estimated in a recent series as 1:2226 of all pregnancies. A delay in establishing a diagnosis and in starting treatment can result in uterine rupture, massive hemorrhage and serious maternal morbidity, and may require hysterectomy. Several options are available to treat CSP if diagnosed early, although there are no evidence-based guidelines recommended due to its rarity. The management should be tailored to the individual situations. Little is known about the future pregnancies, outcomes and recurrences after fertility-preserving treatments following CSP.

We report a case of suspected CSP in a 28 year old Gravida 2, Para one, who was referred to us for the management of incomplete miscarriage. Her previous delivery was six years back and was by Cesarean section. Ultrasound examination revealed that the patient had a large anterior lower uterine segment vascular mass of 9.3x8.2x9cms, suspected to be a persistent trophoblastic tissue invading the anterior uterine wall, though a degenerating fibroid could not be ruled out. The patient underwent dilatation and curettage as she had been bleeding for more than a month and still βHCG being positive. The procedure was also used to establish a histopathological diagnosis. A follow up MRI and Ultrasound revealed a heterogenous mass. With a strong clinical suspicion based on history and early ultrasound reports, the diagnosis of an anterior uterine wall mass probably due to penetrating trophoblastic tissue on previous caesarean scar was made. The patient has been referred for either uterine artery embolisation or a laparoscopic removal in order to preserve her fertility.

Key words: Caesarean scar pregnancy(CSP), cervicoisthmic pregnancy, myoma, persistent trophoblastic tissue, uterine artery embolisation, ectopic pregnancy, magnetic resonance imaging, transvaginal ultrasound.

INTRODUCTION
Implantation of the embryo on a previous caesarean scar is a type of ectopic pregnancy though it is inside the uterus. It is one of the aftermaths of a globally-rising incidence of caesarean section resulting in significant maternal morbidity, the need for blood transfusions, and even hysterectomy unless identified and treated early.

Review of English medical literature revealed the first case of caesarean scar pregnancy had been reported in 19781. Though not many cases have been reported since then, during the past five years there have been a substantial number of publications regarding this clinical entity. This could reflect either an apparent increase due to the better diagnostic modalities being available now or could be a real increase because of increasing caesarean rates worldwide.

A recent case series2 estimates the incidence of CSP at1:2226 of all pregnancies, with a rate of 0.15% in women with previous caesarean scar and 6.1% of all ectopics in women who had at least one caesarean section. No much predilection with maternal age or
parity has been noticed. The time interval since the last caesarean has ranged from anywhere between six months to 12 years. It has been reported even following IVF with embryo transfers.

CASE REPORT
A 28 year old Gravida 2 Para 1 was referred from a private clinic to GMCHRC on 12-05-2012 with a diagnosis of incomplete miscarriage, for evacuation of retained products of conception. By dates (LMP 20-01-12) she was about 15-16 wks of gestation and had been bleeding per vaginum for the past 1 month. The patient had been having a live intrauterine fetus in early pregnancy, low-lying in the intrauterine cavity, and was expecting a spontaneous miscarriage (with a probable diagnosis of inevitable miscarriage).

The patient’s last delivery was six years back and by caesarean section. She had subfertility following her last childbirth and was on treatment for the same. She had been diagnosed as PCOS and was on Metformin.

The patient’s hemoglobin was 10gm% and coagulation profile was normal. βHCG was 150mIU/ml.

USG at GMCHRC showed retroverted uterus of normal size, with endometrial thickness of 12mm. A heterogeneous highly vascular mass of 9.3x8.2x9cm with cystic changes was noted in the anterior lower uterine segment, intramuraly, extending towards the submucous region and lying just above the cervix. Cervix, adnexa and Pouch of Douglas were normal. The probable diagnosis was degenerating fibroid (Figures 1, 2 & 3).

In view of the prolonged bleeding and a positive HCG titer, the patient was posted for D&C under Ultrasound guidance. Elective evacuation of retained products of conception was done. Plenty of friable yellowish necrotic material was evacuated from the anterior uterine wall. As expected, there was heavy bleeding and the procedure was abandoned midway and bleeding was controlled with uterotonins. Though there was a mild drop in hemoglobin levels, the patient made an uneventful recovery and was discharged the next day under antibiotics cover.

Histopathology report suggested a degenerated decidual tissue with occasional degenerated/ghost-like chorionic villi with a few tiny fragments of endometrial tissue showing a few inactive glands.

Two weeks postoperatively, the patient underwent MRI. The results indicated a heterogeneous anterior lower uterine segment mass of 9x7x5cm, relatively hypovascular compared to the uterus, suggestive probably
very few of them progress beyond the first trimester. Literature review shows first and second trimester rupture of uterus in Caesarean scar pregnancies. If they progress to the second and third trimesters they are likely to result in catastrophic hemorrhage, uterine rupture, need for hysterectomy, bladder invasion and loss of future fertility. If the pregnancy continues within the uterus it is likely to end up with a 3-5 fold increase in placenta accreta. Cases that have progressed up to 35 weeks ending up with massive hemorrhage and disseminated intravascular coagulation requiring hysterectomy have been reported.

The exact reason why this implantation occurs is not well understood. Implantation on scar is different from placenta accreta. But it would appear to have same pathogenesis and pathophysiology. Placenta accreta is characterized by the absence of decidua basalis and invasion of trophoblast into uterine myometrium. The pregnancy is however in the uterine cavity, whereas in CSP, the gestation is surrounded entirely by the myometrium and fibrous scar tissue and separate from the uterine cavity. This could suggest that the invasion into myometrium occurs through microtubular tract between the caesarean section scar and endometrial cavity. CSP is more aggressive than placenta accreta because of the early invasion. CSP could grow either towards cervicoisthmic space or towards the uterine cavity.

It is not clear whether the risk of CSP is related to the number of caesarean sections or the time interval since the previous surgery. Some believe that multiple caesarean section increases the chances because of increased scar surface area. Caesarean section pregnancies have been reported even as late as 12 years following previous caesarean sections. Thus, it is difficult to say that incomplete healing might have contributed to scar pregnancy. Little is known whether single layer closure of the uterus increases the risk of CSP.

The early pregnancy ultrasound pictures and the reports which were done around 7 weeks showed a low positioned live intrauterine pregnancy of 6+ weeks of gestational age, with no other uterine masses. After evaluating them, a probable diagnosis of caesarean scar pregnancy with persistent trophoblastic activity invading the anterior uterine wall was made.

The other probable differential diagnoses were degenerating fibroid or a cervicoisthmic pregnancy. In view of the need for preserving the patient’s fertility, she was referred to undergo uterine artery embolisation or laparoscopic removal of this persistent uterine mass.

**DISCUSSION**

Little is known about the natural history of caesarean scar pregnancy (CSP) as of a degenerating fibroid. Repeat HCG level was 37mIU/ml. Repeat ultrasound findings were not much different from those of MRI.

Magnetic resonance image showing a well defined lesion in relation to the antero-inferior aspect of the uterus extending to the area of the anterior cervix and measuring 9x5x7 cm, which is mostly submucous splaying the endometrium both superiorly and inferiorly, extending more anteriorly to the anterior subserosal surface (Figure 4).
Jurkovic et al\textsuperscript{5} have described a negative "sliding organ sign", defined as the inability to displace the gestational sac from its position at the level of internal os by gentle pressure applied by the transabdominal probe, as diagnostic.

![Figure 6. Ultrasound image of a sagittal section of uterus showing protrusion of the gestations sac with fetus anteriorly through the scar, with empty uterine cavity at fundus](image)

Doppler shows a distinct circular peritrophoblastic perfusion surrounding the sac 3D power. Doppler ultrasound has been used to enhance the diagnostic accuracy. MRI is used as an adjunct, with sagittal and transverse sequences showing gestational sac embedded in the anterior lower uterus. MRI also helps in planning management with methotrexate or surgery, by improving intraoperative orientation.

According to many authors, a TVS combined with a colour Doppler is highly reliable in diagnosing a caesarean scar pregnancy, and MRI may be reserved for cases in which they are inconclusive. Diagnostic hysteroscopy and laparoscopy have also been used in the diagnosis of CSP.

Histology of an excised CSP or in a hysterectomy specimen reveals interstitial trophoblasts within fibromuscular tissue. These features with the absence of the surrounding endocervical gland confirms CSP and rules out cervical pregnancy (Figure 7).
DIFFERENTIAL DIAGNOSIS

CSP could be easily misdiagnosed as miscarriage in progress or as cervicisthmic pregnancy.

In case of a spontaneous miscarriage in progress, the gestation sac should be seen in the cervical canal on TVS, and on colour flow Doppler, the sac should appear avascular, indicating that the sac has been detached from its implantation site, in contrast to the well-perfused CSP. Unlike a CSP, in cervicisthmic pregnancy there would be a layer of healthy myometrium visible between the bladder and the gestation sac, and heavier bleeding as the presenting symptom.

On a transabdominal ultrasound with full bladder, a gestational sac present dominantly within the cervix, giving an hour-glass shaped empty uterus with a ballooned cervical canal establishes the diagnosis of cervical pregnancy.

Rarely CSP could be misdiagnosed as trophoblastic tumor infiltrating myometrium. Establishing the diagnosis is easier in early pregnancy because as the pregnancy advances, differentiating it from other differential diagnosis becomes more difficult.

TREATMENT

It is surprising that as many as 31 different and combination treatment modalities have been described in the literature. They range from expectant management, D&C, ultrasound-guided local methotrexate injections, laparoscopic excision, hysteroscopic excisions, uterine artery embolisations to hysterectomy. Sporadically, the cases and individualised treatment approaches could be the reason why so many treatment modalities have been suggested. According to Seow et al, it has been found that the caesarean section mass may take two months to as long as 1 year to regress after treatment. The review of literature shows expectant management of viable CSPs either fail or carry a high risk of rupture, compromising fertility. The absence of evidence-based guidelines has made the obstetricians and gynecologists uncertain about the best probable method of approach in these cases. However, an early diagnosis and treatment are certain to improve the outcome as life-threatening complications could reduce.

Is it possible to prevent CSP? And what are the future prospects?

Several articles in the literature have suggested a surgical repair of a caesarean scar defect seen as a niche (non-pregnant state) during transvaginal ultrasonography either hysteroscopically or laparoscopically. However, the outcomes of the procedure have not been reported. Further research is indeed needed to recommend these methods in preventing CSP.

Due to the insufficient data, it is difficult to advise on the recurrence risk of CSP. Some authors recommend future pregnancies should be delayed for more than three months and probably for 1-2 years. It is unclear whether any specific method of closure of caesarean section, either single or double layer, could have an impact on the occurrence CSP or placenta accreta.

CONCLUSION

According to an extensive review of the literature done by Ilan Timor et al and reported in AJOG July 2012, there were a few impressive clinical conclusions drawn.

1. CSP and placenta accreta were often misdiagnosed as low intrauterine...
pregnancy, cervical pregnancy or miscarriage in progress.
2. The best diagnostic tool is high frequency transvaginal ultrasound. MRI, which could be considered as an additional method to arrive at a diagnosis.
3. The earlier the diagnosis the better the outcome, and it starts with patient education. The patient after caesarean delivery should be advised on an early visit to the obstetrician in future pregnancy.
4. If possible D&C should be avoided. Even systemic methotrexate as a single treatment of choice should be avoided.
5. Uterine artery embolisation as a primary treatment in an uncomplicated case should be used sparingly. However its use as a rescue procedure is undisputed.
6. Transvaginal or transabdominal guided local methotrexate injections with or without additional intramuscular methotrexate as well as surgical excision by hysteroscopic guidance carried the lowest complication rate.
7. There is an urgent need for a standardized guideline in managing these cases as its occurrence is increasing.

Owing to the lack of evidence-based guidelines it is essential for every clinician to watch out for CSP in women who conceive after caesarean, and actively involve the patient in decision making after discussing the available treatment modalities. It is the need of the hour to have more extensive research and evidence-based guidelines in the management of this catastrophic and dangerous clinical entity.

REFERENCES