Clinical Dilemmas and Risks of Misdiagnosis and Mismanagement Associated with Endogenous Caesarean Scar Pregnancy: A Case Series and Literature Review

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Abstract

A caesarean scar pregnancy (CSP) is a pregnancy that implants into the myometrium at the site of a previous uterine incision. In this paper, we present three cases of women affected by caesarean scar implantations. Each case presented differently and was managed in separate ways. We describe their differing presentations and care as well as reflecting on how such women may be better cared for. Endogenic CSPs grow into the uterine cavity and can easily be mistaken for a normal pregnancy implantation. Differentiation can often only be seen at early gestations. All women with a previous uterine scar should potentially be offered early trans-vaginal ultrasound to correctly diagnose the implantation site. However, in many women, the diagnosis is not suspected until a complication arises. We advocate for there to be a higher index of suspicion for CSPs during early pregnancy ultrasound and that all practitioners performing such scans should be trained to allow a confident diagnosis of CSP.

Keywords: Caesarean scar pregnancy; Caesarean sections; Intravenous; Ultrasound scan; Human chorionic gonadotrophin

Abbreviations: CSP: Caesarean Scar Pregnancy; TVS: Trans Vaginal Ultrasound; CS: Caesarean Sections; TOP: Termination of Pregnancy; MVA: Manual Vacuum Aspiration; IV: Intravenous; A&E: Accident & Emergency; TAS: Trans Abdominal Scan; UV: Ultrasound Scan; HCG: Human Chorionic Gonadotrophin

Introduction

Caesarean scar pregnancy is defined as implantation into the myometrium defect occurring at the site of the previous uterine incision. The prevalence of CSP is estimated to be approximately 1 in 2000 pregnancies and these pregnancies may be ongoing potentially viable pregnancies or miscarriages within the scar. 37 The diagnostic criteria described for diagnosing caesarean scar implantation on transvaginal ultrasound include:

I. Empty uterine cavity.

II. Gestational sac or solid mass of trophoblast located interiorly at the level of the internal as embedded at the site of the previous lower uterine segment caesarean section scar.

III. Thin or absent layer of myometrium between the gestational sac and the bladder.

IV. Evidence of prominent trophoblastic/placental circulation on Doppler examination.

V. Empty endocervical canal.

Thirteen percent of reported cases of CSP were misdiagnosed as intrauterine or cervical pregnancies at presentation. The true prevalence of caesarean scar pregnancies is likely to be somewhat higher than estimated in the literature as some cases will end in the first trimester, either by miscarriage or termination, and go unreported and undiagnosed. There is a spectrum of severity CSP is defined as implantation into the myometrial defect occurring at the site of the previous uterine incision. The prevalence of CSP is estimated to be approximately 1 in 2000 pregnancies and these pregnancies may be ongoing potentially viable pregnancies or miscarriages within the scar.
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The natural history of such pregnancies has not been fully elucidated. The morbidity and early indicators of outcomes are yet to be fully described. It is suggested placenta accreta is the end point of an ongoing caesarean scar ectopic pregnancy [3]. Management options are variable and the best way to prevent recurrence unclear [4]. As with other non-tubal pregnancies misdiagnosis is common resulting in mismanagement and poorer outcomes. In addition to a comprehensive history, transvaginal ultrasound (TVS) is the primary modality for diagnosis. A recent RCOG guideline has emphasized ultrasound diagnostic criteria, however, these have not been validated [4]. In this paper, we present three cases of women affected by caesarean scar implantations, describing their presentation and management as well as reflecting on how such women may be better cared for.

Case Study 1

A 30-year-old Afro-Caribbean, gave a history of two previous caesarean sections (CS) and a first trimester termination of pregnancy (TOP). The surgeon performing her last CS had advised her against future pregnancies due to a very thin lower segment. As a result, when she inadvertently fell pregnant she opted to terminate the pregnancy. TVS at the local TOP service suggested that the pregnancy was ‘outside the womb’ and referral was made to a neighbouring early pregnancy unit where she was scanned by a sonographer. TVS here showed a viable seven-week intrauterine pregnancy with an irregularly shaped gestational sac. Consequently, she returned to the termination service for her surgical procedure at 9 weeks and 3 days. She underwent a manual vacuum aspiration (MVA) under sedation. This was complicated by heavy vaginal bleeding and the operator was concerned about a possible uterine perforation. The patient was given uterotonics (syntocinon 5+5IU IV, misoprostol 1mg PR) as well as intravenous (IV) tranexamic acid (1g). A vaginal pack was inserted and ambulance transfer to our accident and emergency (A&E) resuscitation room was arranged.

On arrival, her blood pressure was 96/58mmHG with a heart rate of 89bpm, her haemoglobin estimation was 70g/dL. A transabdominal ultrasound scan (TAS) showed a bulky uterus with a clot in the cavity and a second mass, presumed clot, above the fundus. She was consented and transferred to theatre. She underwent laparoscopy with Palmers point entry. There were dense adhesions from her previous CS scar resulting in the uterus being attached to the anterior abdominal wall. These adhesions were divided and the bladder reflected, revealing a thinned out lower segment scar and a ballooning above the cervix (Figure 1). A diagnosis of a CSP was made. Using a harmonic scalpel the lower segment of the uterus was opened and the placental bed excised. A 400ml blood clot was removed from inside the
She gave a history of one previous CS for foetal distress, pain, vaginal bleeding and a positive urinary pregnancy test. Case Study 3

A 38-year-old Caucasian lady, presented with abdominal pain, vaginal bleeding and a positive urinary pregnancy test. She gave a history of one previous CS for foetal distress five years previously. TVS showed a gestational sac implanted low in the uterine cavity with early foetal demise. Subsequent TVS performed by a consultant, immediately prior to a scheduled MVA procedure, showed a very vascular and irregular gestational sac measuring 42x32x45mm lying low in the uterus near the CS scar with only a thin layer of covering myometrium. A CSP was suspected and a decision was made to perform an evacuation of retained products of conception under general anaesthetic in anticipation of potential complications. A suction evacuation was performed with ultrasound guidance. The theatre team were prepared for large blood loss and appropriate blood products had been made available. Following removal of the pregnancy there was brisk bleeding. TAS confirmed an empty cavity and there was no suspicion of perforation. She received misoprostol (1mg PR), syntocinon 5+5iu IV and 40IU infusion, carboprost IM 250mg twice and IV tranexamic acid (1g) alongside bimanual compression for 10-15 minutes compression. Her bleeding settled with a measured blood loss of 2250ml. Starting haemoglobin was 138g/dl, she received two units of red blood cells and haemoglobin following these was 113g/dl. She remained on the ward overnight for observation and was given three, four hourly doses of oral misoprostol. There was no further blood loss she discharged the following day.

Discussion

CSPs are a relatively modern phenomenon. The increasing incidence is thought to be either due to increased reporting or increasing global caesarean section rates. The symptoms of CSP do not differ from those of other non-tubal pregnancies but they occur exclusively in women who have had previous CS, occurring in about 6.1% of women with an ectopic pregnancy and at least one CS [5]. The pathophysiology of caesarean scar pregnancies remains unclear. It is likely that a CSP forms following invasion of the implanting blastocyst through a microscopic niche in the healed uterine scar. Additionally, there may be a more global impact on the endometrium due to the very presence of scar tissue which allows aberrant implantation [6]. CSPs form a clinical spectrum from a partial implantation over a thick scar, which grows into the uterine cavity, to a pregnancy fully located outside the uterine cavity, only connected to the uterine cavity via a thin tract. Two main types have been described [7].

The first type, also known as type 1 or endogenic, implants over the scar but proceeds to develop within the uterine cavity. It may appear as a viable intrauterine pregnancy, complete with yolk sac, embryo and cardiac activity, and progress to a viable gestational age with possible problems with placentation [8]. Type 2 or exogenic scar ectopics are pregnancies that implant more deeply within the scar and grow in the direction of the serosal surface of the uterus towards the broad ligament or bladder. A thin layer of myometrium may be seen at early gestations between the gestational sac and the serosa. In two thirds of cases this will measure less than 5mm [9]. As the pregnancy grows this layer thins and disappears. Consequently,
the pregnancy bulges through the scar defect with a much greater rate of early uterine rupture and subsequent haemorrhage.

Ultrasound remains the most used modality for diagnosis. A recent guideline from the RCOG [4] describes criteria for diagnosis which include:

a. Empty uterine cavity.

b. Gestational sac or solid mass of trophoblast located anteriorly at the level of the internal os embedded at the site of the previous lower uterine segment caesarean section scar.

c. Thin or absent layer of myometrium between the gestational sac and the bladder.

d. Evidence of prominent trophoblastic/placental circulation on Doppler examination.

a. Empty endocervical canal: Ultrasound can also be used to differentiate between type 1 and type 2 CSP. In endogenic (type 1) CSP, the deformity seen on ultrasound is less apparent as the implantation grows towards the uterus. The ultrasound image consequently can appear as a viable intrauterine pregnancy [10]. In these types of low implantation endogenic CSP, differentiation from a normal pregnancy can often only be made by ultrasound at earlier gestations [3]. For optimal management of CSP a high index of suspicion and early diagnosis is the key to successful outcomes [11]. This also allows women to make informed choices based on potential morbidity [4]. As a rule of thumb, taking the woman’s wishes into consideration, CSP diagnosed at early gestation should be terminated. The decision is easier for non-viable or unwanted pregnancies as opposed to wanted pregnancies where a heartbeat is identified on scan. Though there have been reports of successful deliveries an expectant approach increases the risk of a morbidly adherent placenta with likely caesarean hysterectomy and significant haemorrhage [12-15].

An additional question raised by 2 of our 3 cases is whether it is safe for women who want to have a TOP and have a history of CS to have this performed in community based TOP centres or whether they should be performed in centres with options for laparoscopic surgery or intensive care facilities should the need arise.

There are very few randomised studies of the management of CSP. Most evidence comes from case series and reviews [4], there is one systematic review of outcomes [16]. Current evidence regarding differing management options has recently been reviewed [6].

Conclusion

Literature suggests that we are not over diagnosing caesarean scar pregnancies. Most case reports, including our own, describe situations where the diagnosis was only suspected following significant maternal bleeding after instrumentation of the uterus for management of a miscarriage or termination. The key to the diagnosis is a high index of suspicion and a TVS performed by an appropriately experienced and skilled operator. Up to 13.6% of CSPs are misdiagnosed as an inevitable miscarriage or a cervical ectopic [6]. Interventions in such cases without the necessary preparations for major haemorrhage can lead to increased morbidity and hysterectomy. Strong Doppler colour flow and the presence of the ‘sliding sign’ differentiates a scar ectopic from a low lying inevitable miscarriage. Ballooning of and a sac within the cervix are the hallmarks of a cervical ectopic pregnancy.

We recommend that all women with a history of caesarean section are referred for an early viability TVS with an appropriately skilled sonographer or gynaecologist to rule out a diagnosis of caesarean scar ectopic. This should be performed before routine dating scans, to allow time for counselling and management should a CSP be diagnosed. To facilitate this; sonographers, nurse specialists and doctors performing these scans in early pregnancy units should be encouraged to consider the diagnosis and be appropriately trained in the ultrasound diagnosis of CSP. We recommend that all women undergoing TOP with a caesarean section scar site implantation or suspected Caesarean ectopic pregnancy have their procedure performed in units with facilities not only to cope with major haemorrhage but also to perform laparoscopic surgery should this be needed.

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