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Case report

A large post-caesarean Niche (Isthmocele) with amenorrhea, a symptom that was not reported in the medical literature: A rare case report

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ABSTRACT

Introduction and importance: Isthmocele is identified as an iatrogenic defect in the myometrium of the anterior uterine wall at the site of a previous cesarean scar due to defective tissue healing. Patients may have varied symptoms including abnormal uterine bleeding (AUB) and pelvic pain.

Herein, we report a rare case of a large isthmocele that manifested with secondary amenorrhea; which was not reported in the medical literature previously.

Case presentation: A 30-year-old Syrian woman, G5P5, came to our clinic with a complaint of secondary amenorrhea that began two years ago. She was treated symptomatically with progesterone with no response. She has had five cesarean sections. Ultrasonography findings suggested a large uterine niche. Trans-Abdominal niche repair was the obtained technique, depending on the drainage of the isthmocele, excising the fibrotic tissue from the edges and re-approximating them. On follow-up, menstruation returned to normal.

Clinical discussion: Isthmocele can be, radiologically, defined as a hypoechoic or anechoic, triangular area at the scar site. Its pathophysiology is still unknown. Although, an isthmocele can be diagnosed using a variety of imaging techniques like ultrasonography (US), magnetic resonance imaging (MRI), sonohysterography, and hysteroscopy; transvaginal ultrasound (TVUS) is the first method described for assessing it.

The goal of isthmocele treatment is to alleviate symptoms.

Conclusion: We recommend that health awareness campaigns alert people to the need to see a specialist doctor in the context of a serious complaint. For the uterine niche, many risk factors can be avoided to reduce its probability.

1. Introduction

Cesarean section (CS) is one of the most common surgical operations worldwide and its rate is growing dramatically to form one-third of all deliveries (1). With this uncontrolled increase in CS deliveries, the early and late complications of this procedure also increase. One of these complications is the isthmocele. Isthmocele or uterine niche, or cesarean scar defect (CSD) is identified as an iatrogenic defect in the myometrium of the anterior uterine wall at the site of previous cesarean scar due to defective tissue healing. Sometimes it is described as myometrial thinning (2). The prevalence of isthmocele varies due to diagnostic techniques. Tulandi et al. have reported that in transvaginal ultrasound (TVUS) examination, the prevalence ranges between 24 % to 70 %,

while in sonohysterography (SHG) examination the prevalence ranges from 56 % to 84 % (3,4). The etiology is still unclear but many risk factors were determined such as multiple CS (3), long duration of labor, cervical dilatation, stage of the presenting part, and low uterine incision (1). Patients may have varied symptoms including abnormal uterine bleeding (AUB), pelvic pain, post-menstrual spotting, and infertility, though many women may be asymptomatic and diagnosed incidentally (2,5).

The diagnosis is made by sonography (transvaginal and trans-abdominal), saline instillation SHG, or magnetic resonance imaging (MRI) (4,5). The optimal treatment differs from one patient to another. Treatment options range widely from medical treatment to hysterectomy (5).

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This manuscript has been reported in line with SCARE's 2020 Criteria (6).

Herein, we report a rare case of a large isthmocele that manifested with secondary amenorrhea; which was not reported in the medical literature previously.

2. Presentation of case

A 30-year-old Syrian woman, G5P5, came to our private clinic with a complaint of secondary amenorrhea and chronic pelvic pain. Her complaint began after the last cesarean section which was two years ago. Pelvic pain was moderate, colic, and non-responsive to analgesics. For secondary amenorrhea, she was treated symptomatically with progesterone to perform withdrawal bleeding; without undergoing any diagnostic tests; but with no response. She is a smoker with no medical history. She has no family history of a similar problem. She has had five cesarean sections. Physical examination and laboratory tests were unremarkable.

TVUS of the pelvis showed a cystic structure with a thick wall and turbid content on the front face of the uterus at the site of the CS scar, behind the bladder. The structure measured (57 × 38 × 32) mm and connected with the uterine cavity by a 10 mm duct. The residual myometrium (RMT) was <3 mm. These findings suggested a large uterine niche at the site of the CS. No additional investigations were obtained.

As the patient underwent failed medical treatment previously, surgery was the treatment of choice. The hysteroscopic and laparoscopic approaches were excluded due to three factors: the expected adhesions due to five past CS, high costs, and low resources in our country after the war. On the other hand, laparotomy was preferred due to the large size of the defect and RMT < 3 mm.

Trans-Abdominal niche repair was the obtained technique. First, Hegar's dilator and intracervical Foley catheter were applied by US guidance to achieve better identification of the defect.

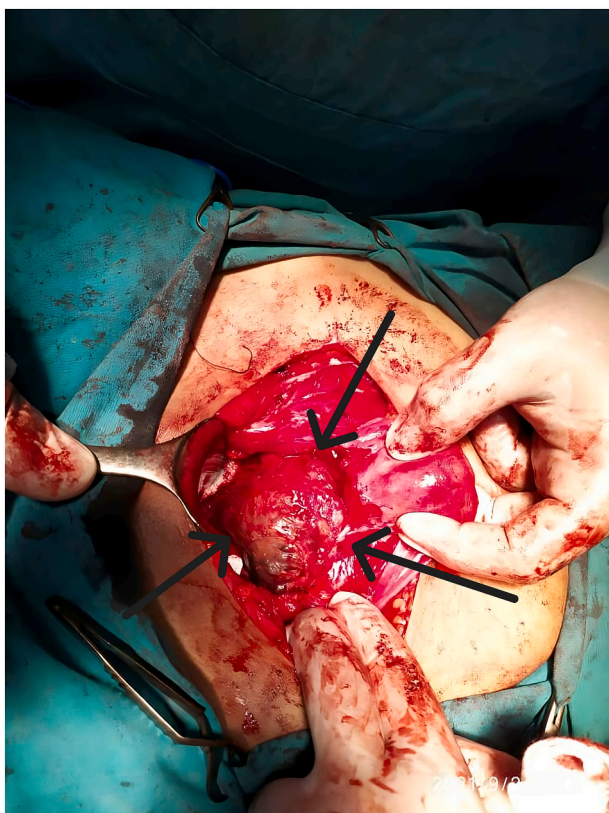


Fig. 1. A large isthmocele that measures 5 cm in diameter, before drainage.

After the abdomen was opened, the bladder was dissected from the anterior face of the uterus where the isthmocele was located (Fig. 1). The isthmocele was drained (Figs. 2 and 3) by a longitudinal incision. Fibrotic tissue from the edges was excised and reapproximated in 2 Layers.

On follow-up, pelvic pain was resolved and menstruation returned to normal two months after the operation. The RMT significantly increased.

3. Discussion

While the optimal CS rate recommended by The World Health Organization (WHO) is around 15 % (7), the current rate is increasing rapidly with prevalence ranging between 6 and 27.2 % according to a study including data from 150 countries (8,9).

Although Cesarean incisions recover well, it has complication as any other surgical procedure (10).

CSD is one of the complications which is also called niche, isthmocele, diverticulum, or pouch. Poidevin in 1961, first described isthmocele (11).

Radiologically, isthmocele can be defined as a hypoechoic or anechoic, triangular area at the scar site (4,12).

The pathophysiology of the uterine niche is still unknown, although several risk factors were reported (1,7,13).

Vervoort et al. have reported four hypotheses about the etiology. The first hypothesis focuses on the site of the hysterotomy. This hypothesis proposes that the lower uterine incision on its cervical part is associated



Fig. 2. The discharge of the isthmocele content after a longitudinal incision on it.



Fig. 3. The green arrow indicates the isthmocele margin. The black arrow demonstrates a Foley catheter inside the isthmocele. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

with a higher incidence of isthmocele. This is explained by the presence of mucous glands in this part where the incision was made. These glands produce mucus during the healing time, which could dilate the sutured rims of the myometrium. This can be seen in cases of longer active labor prior to emergency cesarean (7).

The second hypothesis concerns the surgical technique. The invalid closure of the deeper muscular layer or an incomplete closure could be responsible for CSD formation and development. The third hypothesis proposes that impaired wound healing, and attracting the rims of the wound due to early adhesion formation between the hysterotomy scar and the anterior abdominal wall are the essential causes of isthmocele (7,14).

The fourth hypothesis relates to patient factors, such as individual/genetic predisposition, and post-operative infection (7). The details for the past five cesarean sections of our patient; the indication, location of the incision, and uterine closure technique; were not available, so we could not favor one hypothesis over another.

The principal risk factor is multiple CS (3), **as in our case**. There are several other risk factors such as duration of labor, cervical dilatation, stage of the presenting part, and low uterine incision that accompany lesser vascularized myometrium resulting in inadequate healing (1,15). Retroflexed Uterus is also an important risk factor, as gravity increases the counteracting forces on the uterus (1,7). **The uterus in our case was anteфлекed.**

In some cases, the symptoms could be present because of the size of the defect (3).

Abnormal uterine bleeding (AUB) is the most common symptom which usually presents as postmenstrual bleeding (3). **But, in our case, the main symptom was secondary amenorrhea which was not reported in the medical literature.**

Accumulation of blood and menstrual debris within the defect may be predisposed by the presence of an isthmocele, which is associated with decreased uterine contractility due to fibrotic tissue around the scar. This accumulation slows menstrual flow leading to AUB (4,16).

Morris (17) suggests, based on the pathology findings of free erythrocytes in the scar tissue, that there was a recent hemorrhage and that the blood could also have been produced in situ, causing intermittent spotting. No matter what the source, the presence of blood in the isthmocele is likewise connected with a higher mucus emission, which could add to postmenstrual AUB (18).

Patients with CSD may also suffer from a significant problem in the form of infertility. The lower ripeness rate may be connected with the constancy of menstrual blood in the pocket, which influences the cervical mucus, as well as sperm motility and implantation (1,19).

Several symptoms were recorded in the literature, such as dysmenorrhea and pelvic pain (13).

Usually, many obstetric complications occur during pregnancy, associated with the presence of an isthmocele. These obstetric complications include uterine rupture, placenta previa, and scar dehiscence (20).

A cesarean scar ectopic pregnancy, which occurs in almost 1 in 1,886 to 2,216 pregnancies, is another complication that has been reported (19).

The walls of the isthmocele may rupture during the development of the fetus and the gestational sac, resulting in the known severe complications of an ectopic pregnancy (13,19).

Until now, there are no clear criteria for the diagnosis of isthmocele (3,13,21).

The anterior uterine wall can be evaluated and an isthmocele can be diagnosed using a variety of imaging techniques like US, MRI, SHG, hystero-graphy, and hysteroscopy (22).

The first and most common method described for assessing the integrity of the uterine wall in non-pregnant patients is TVUS (13,22), **as in our case.**

Because postmenstrual bleeding is the primary symptom, the early proliferative phase best demonstrates the blood inside the isthmocele, making its identification possible even without saline infusion (3).

On TVUS, the defect has been identified as either a deformity (wedge, shape, concavity, or sacculation) on the anterior isthmus or an anechoic triangle defect in the myometrium with the base communicating with the uterine cavity (23,24).

Using six shapes to describe the defect, Bij de Vaate et al. (4) proposed a more systematic classification: droplet, cyst, semicircle, rectangle, circle, and triangle.

In the evaluation of isthmocele, the most useful discriminating measurement is the residual myometrium thickness (25).

In addition, patients who have had two or more previous CSs have a scar that is thinner. The scar is thicker in those who had their last CS more than two years ago (26).

The goal of isthmocele treatment is to alleviate symptoms. As a result, cases without symptoms should not be treated (27).

Medical treatment is an option, though surgery is the most common choice. Surgical options include laparotomy, vaginal repair, hysteroscopy, and laparoscopy (including robotic laparoscopy) (28).

For symptomatic ladies who would rather not get pregnant and favor a conservative treatment, oral contraceptive pills could be the chance of choice (29).

The leftover myometrial thickness is the principal parameter to carry out hysteroscopy. Furthermore, if the myometrium thickness at the site of the defect is <3 mm, the hysteroscopic approach may result in bladder injury and uterine perforation (30).

If there are symptoms and a desire to preserve fertility, a trans-

abdominal approach (laparotomy, laparoscopic, robotic) has been recommended for large defects (RM <3 mm) (5).

In trans-abdominal isthmocele repair, the edges of the isthmocele are cut off to remove the scar tissue, and two-layer sutures are used to close the defect (3).

The trans-abdominal approach makes it easier to see where the problem is, which makes it possible to repair it and makes the myometrium thicker (14).

In a retrospective study, Zhang found that the transvaginal repair and the laparoscopic approach produced comparable outcomes. Moreover, It was reported that the transvaginal isthmocele repair was less expensive, required less time to perform, and was comparable to laparoscopy in terms of effectiveness (31). **For our patient**, laparoscopy could not be conducted due to high costs and low resources. Also, the transvaginal approach was not obtained due to the large size of the isthmocele and our available surgical abilities on the trans-abdominal approach are more qualified than the transvaginal ones.

Hysterectomy is the definitive treatment for symptomatic patients who have finished their reproductive life (22).

4. Conclusion

Symptomatic treatment is a common act in developing countries. Maybe, it is acceptable due to expensive diagnostic tests in these countries, but to a specific limit. Long symptomatic management with no response can aggravate small problems which can be resolved with lesser interventions. To limit this problem, we recommend that health awareness campaigns alert people to the need to see a specialist doctor in the context of a serious or long complaint.

In addition, for the uterine niche, some risk factors can be avoided by the patient and the doctor. For the patient, cesarean planning is effective. For doctors; electing the appropriate indication for cesarean, the best site for uterine incision, and the best technique for uterine closure; all these points can reduce uterine niche probability.

Abbreviations

CSD	Cesarean scar defect
CS	Cesarean section
TVUS	Transvaginal ultrasound
SHG	Sonohysterography
AUB	Abnormal uterine bleeding
MRI	Magnetic resonance imaging
US	Ultrasound
RMT	Residual myometrium
WHO	World Health Organization

CRedit authorship contribution statement

Basel Al-Ghotani: contributed to drafting, reviewing, and editing.
Nafiza Martini: contributed to drafting, reviewing, and editing.
Ebaa Alabdallah: contributed to drafting, reviewing, and editing.
Ieman Alawad: contributed to data collecting, reviewing and editing.
Khaled Hussien: is the mentor. Contributed to reviewing and supervising.

All authors read and approved the final manuscript.

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Ethics approval

Ethical approval for this study was provided by the Ethical Committee of Damascus University (ID number: 4369), Damascus, Syria on 30 June 2022.

Declaration of competing interest

All the authors declared that they have no conflicts of interest.

Data availability

All the relevant patient data and clinical history is provided within this article.

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Guarantor

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