What is your diagnosis?

A 42-year-old woman presented to the outpatient department with a skin-colored, non-tender, firm, immobile subcutaneous lump on the left corner of the Pfannenstiel scar. The patient noticed the mass eight years back. Initially it was pea sized but gradually increased to approximately 15x15 cm. It was associated with dull aching pain that started two days before menses and lasted five days after completion of menses. This period was also associated with cyclical dyspareunia and swelling around the lump.

Her obstetric history was notable for a full term, normal vaginal delivery 15 years earlier followed by medical termination of pregnancy 12 years earlier because of a malformed fetus. She underwent full term lower segment cesarean section (LSCS) for antepartum hemorrhage 10 years back. Her previous menstrual cycles were regular with average flow. The patient used homeopathic medication for 6-8 months, but did not experience any relief. She had multiple consultations and hospital visits for the same complaint for the last six years. Fine needle aspiration cytology done six years earlier at another center was suggestive of inflammatory cells, while another performed four years before presentation to our department reported degenerated cells.

On abdominal examination, an immobile, non-tender, hard mass of about 15x12 cm was felt above the pubic symphysis with no local rise of temperature. The mass was adherent to the anterior abdominal wall (Figure 1). On per vaginal and per rectal examination, the cervix was firm, regular and pulled anteriorly, the uterus was posterior and adherent to the mass, although bilateral fornices were free.

Given the clinical presentation, the differential diagnosis may include hematoma, stitch granuloma, lymphadenopathy, dermatofibroma, keloid mass, neuroma, abscess, desmoid tumor, or scar endometrioma and imaging will provide additional insight for diagnosing the lesion.

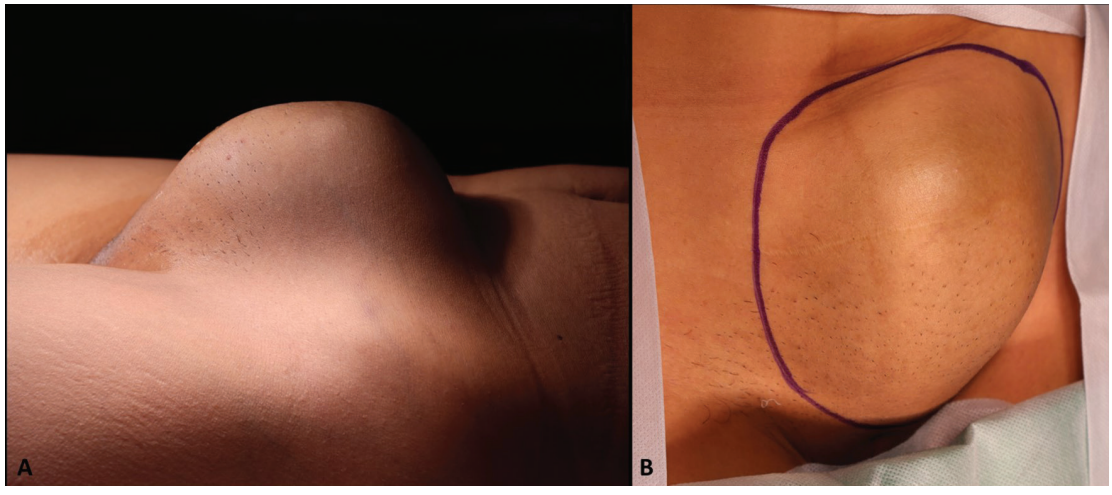

Figure 1. Large, immobile, non-tender mass with restricted mobility at the Pfannenstiel scar site [(A) lateral view, (B) anterior view]

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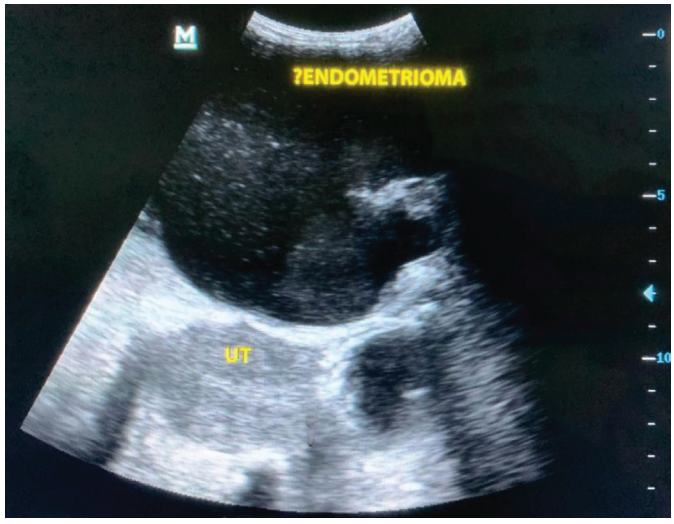

Figure 2. Ultrasonography showing hyperechoic large cystic mass with echogenic contents within with no vascularity

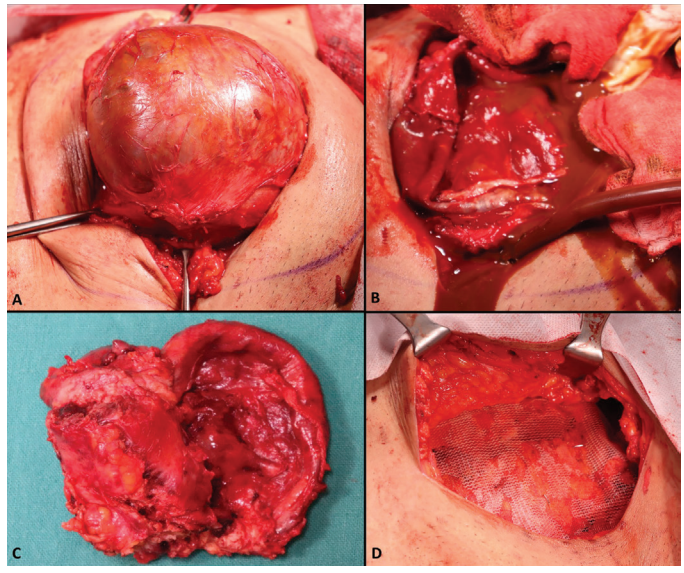

Figure 3. (A) 15x12cm scar site endometrioma adhered to rectus sheath. (B) Drainage of chocolate colored fluid from the endometriotic cyst. (C) Cut section of the specimen. (D) Onlay prolene mesh placed after primary closure of anterior rectus sheath

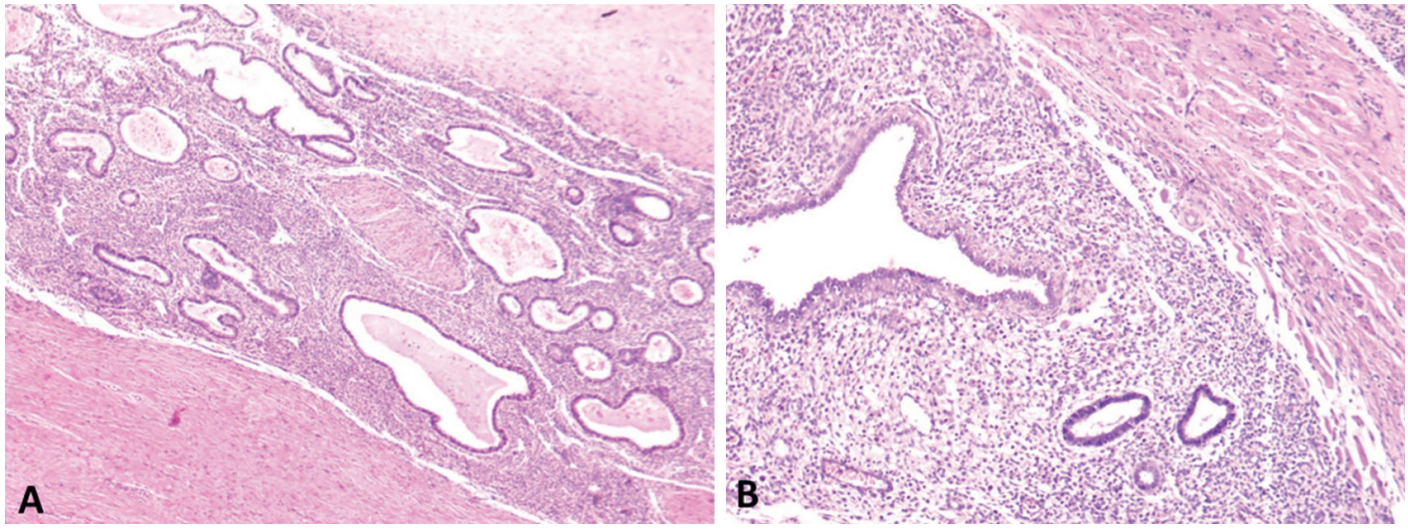

Figure 4. (A) Endometriotic glands and stroma in the subcutaneous tissue consistent with scar endometriosis. (B) Benign endometrial glands and stroma surrounded by scar tissue

Answer

Ultrasonography found a hyperechoic, large, cystic mass with echogenic contents with no vascularity and was suggestive of hemorrhagic cyst (Figure 2). Semisolid components and acoustic shadows were present. The uterus was adenomyotic, bulky and thick. Magnetic resonance imaging (MRI) revealed anterior abdominal wall or scar endometriosis of size 19.3x10.1x9.4 cm.

Based on the characteristic history, examination and radiological findings, the diagnosis of scar endometrioma was made. The patient underwent wide local endometriotic cyst excision followed by onlay prolene mesh repair (Figure 3). The lump was excised in total and final histopathology confirmed scar endometriosis (Figure 4). The patient tolerated the procedure well with an uneventful postoperative course. Currently, the patient is disease free, three years following surgery.

Endometriosis is one of the common gynecological conditions affecting reproductive age women where the non-neoplastic, functional endometrial layer is found outside the uterine cavity. It afflicts at least 11% women in the reproductive age group (1). Endometriosis generally involves pelvic sites, like ovaries, fallopian tubes, pouch of Douglas, uterine ligaments, rectovaginal septum or the pelvic peritoneum (2). Extra pelvic endometriosis is rare and found in unusual sites, such as the bladder, central nervous system, gastrointestinal tract, thorax or cutaneous tissues, including LSCS scar or episiotomy scar, especially after obstetric or gynecological surgical interventions (3).

Scar endometriosis is an extraordinary type of extrapelvic endometriosis with a prevalence reported between 0.03-2% (4). Probable differential diagnoses, including hematoma, stitch granuloma, lymphadenopathy, dermatofibroma, keloid mass, neuroma, abscess or desmoid tumor may cause delayed diagnosis. Depending on the surgical history, cutaneous endometriosis is further divided into primary and secondary cutaneous endometriosis. Primary cutaneous endometriosis occurs when endometriosis develops without any prior surgical intervention whereas secondary cutaneous endometriosis, also known as scar endometriosis, is associated with prior pelvic or abdominal surgery (5). Primary cutaneous endometriosis is less common than secondary cutaneous endometriosis and is thus less likely.

In terms of the pathogenesis of primary and secondary cutaneous endometriosis, the latter is easier to conceptualize. The prevailing hypothesis for secondary cutaneous endometriosis is direct implantation of stromal endometrial cells during surgery, within and adjacent to the incision site, which proliferate under hormonal stimulus; the “cellular transport theory”. However, for primary cutaneous endometriosis, some have proposed that seeding occurs hematogenously or via lymphatics. A third theory, the “coelomic metaplasia” theory proposes that cutaneous endometriosis is the result of metaplasia of pluripotent mesenchymal cells into endometrial tissue (6). The endometrial implant typically appears as a deep-lying or subcutaneous nodule infiltrating the fascia and the muscle, as seen in the present case. The implant was confirmed to be a scar endometrioma rather than an ovarian endometrioma adherent to the anterior abdominal wall, as both ovaries appeared normal and were distinctly separate from the mass. The classical triad is helpful in the diagnosis of subcutaneous endometriosis, which includes menstrual pain, presence of an abdominal wall mass, and history of surgery. However, this triad is only present in 60% of affected women. The frequency of scar endometriosis has increased due to the increased incidence of cesarean sections and laparoscopies performed in recent years. Certain studies have suggested a potentially increased risk of endometriosis associated with a Pfannenstiel incision compared to a midline vertical incision. However, the available evidence is insufficient to draw definite conclusions (7). Scar endometriosis may be noticed after procedures such as amniocentesis or laparoscopy (8). The endometrial implant is commonly observed as a deep-seated or subcutaneous nodule that infiltrates both the fascia and the muscle and during menstruation, there is bleeding into the tissue, leading to cyclic local pain, tenderness, and discoloration. If the nodules are superficial, noticeable signs include cyclic discoloration, bleeding, and ulceration (9). Careful and thorough history taking, physical examination and appropriate imaging modalities like ultrasonography, computed tomography or MRI are key for diagnosis. Ideally, all patients warrant gynaecological workup to rule out concomitant pelvic endometriosis (8). Histopathological examination suggestive of hemosiderin pigment, endometrial glands and stroma in the excised tissue is the diagnostic proof. Local wide excision, with at least 1 cm of margin, is the treatment of choice (9). Large lesions might require placement of synthetic mesh (10). Various protective surgical measures, such as thorough flushing of the wound cavity, eliminating dead space, employing an intro-flexed suture for the uterine incision, and closing both the visceral and parietal peritoneum, have been recommended as strategies to reduce the incidence of cesarean scar endometriosis (8). Postoperative strategies, including the use of combined oral contraceptives or hormonal suppression with gonadotropin-releasing hormone analogs or dienogest, can help mitigate the risk of recurrence and prevent new growth. While these agents are primarily used in the management of pelvic endometriosis, their use in cases of scar endometriosis may also be beneficial, particularly in patients with extensive disease or those who are not candidates for further surgery. However, the supporting evidence for these measures remains limited (11).

A cesarean scar is the most common site for extra pelvic endometriosis. Therefore, it is important to focus on prompt and accurate diagnosis, effective treatment, and preventive measures for scar endometriosis.

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