

Case 15

A 46-year-old Thai female from Bangkok

Chief complaint: A flesh-colored nodule on umbilicus for two weeks



Present illness:

The patient accidentally found a flesh-colored painless nodule on her umbilicus for two weeks. There were not any ulcers or bleeding on her umbilicus formerly. This nodule did not get larger. There was no cyclic pain on this nodule during periods of menstruation

Past history:

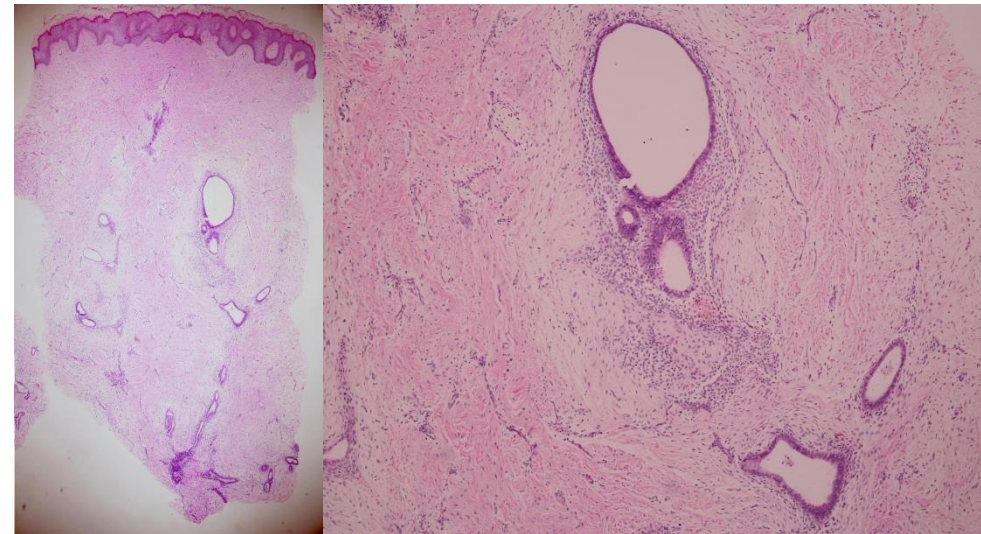
She had been done caesarean section twice for 13 years ago on bikini's line. She denied any histories of dysmenorrhea, oligomenorrhea and hormonal uses.

Skin examination:

- A firm flesh-colored painless nodule about 1.5 cm. in diameter on umbilicus.
- There was no discharge or bleeding on the nodule.

Histopathology: (S14-029208A)

- Multiple cysts and dilated ducts lined by columnar epithelial cells, some of which containing cell debris.
- Surrounding stroma composed of scattered fibroblasts, delicate collagen bundles and inflammatory cells infiltrate of lymphocytes admixed with some hemosiderin.
- Perivascular and nodular inflammatory cell infiltrate of lymphocytes admixed with a few histiocytes, some of which tend to form small tuberculoid granuloma.
- **Microscopic diagnosis:** Cutaneous endometriosis



Gynecological examination:

- Per vaginal examination : normal
- Ultrasonography of pelvis: multiple small uterine myomas about 1.3-1.7 cm. in diameter.

Diagnosis: Cutaneous endometriosis**Treatment:**

- Deep surgical excision of endometriosis

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Discussion:

Endometriosis is defined as the presence of functional endometrial tissue (glands and stroma) outside the normal uterine cavity. It affects 10–15% of all women of reproductive age and 6% of the perimenopausal women.¹ The main location is the ovary, but it has been described in many other extragenital locations, mainly the intestinal wall and the epiploon, but also the lung and the skin. Of the reported cases of cutaneous endometriosis, over 70% are phenomena secondary to prior surgery and occur at the site of surgical scars.² The most widely accepted theory as to the cause of scar endometriosis is the iatrogenic transplantation of endometrial implants to the wound site during pelvic surgery.³ but only if its onset occurs within 2 years after the procedure.^{4, 5} The case of primary cutaneous endometriosis described here remains a rare entity with a reported incidence of 0.5–1% of all extragenital endometrial ectopia. The pathogenesis is still unclear. There are

theories suggesting the spreading of endometrial tissue through lymphatic system and implantation on skin.⁶ The most common area is umbilicus. Other areas are inguinal region and abdominal wall.⁷

Clinically it presents as a reddish-brown nodule, commonly painful, with cyclic variations in size, with or without bleeding, that may coincide or not with the patient's menstrual cycle.⁸

The differential diagnosis of umbilical endometriotic lesions include pyogenic granuloma, umbilical polyps, melanocytic nevus, seborrhoeic keratosis, hemangioma, desmoids, and granular cell tumor. Malignant lesions like melanoma, umbilical metastasis (Mary Sister Joseph nodule), adenocarcinoma, squamous and basal cell carcinoma should be ruled out. Omphalitis, keloid, umbilical hernia, and foreign body granuloma are other differential diagnosis.¹

Diagnosis is confirmed histologically,⁹ cutaneous endometriosis is characterized by the presence of endometrial glands and stroma in the mid- or deep dermis. The endometrial glands are made of tall columnar epithelium with basophilic cytoplasm and basally located nuclei, forming irregular glandular lumina, sometimes with a marked mitotic activity, depending on the phase of the menstrual cycle. The high cellular and vascular stroma is composed of spindle cells and is usually edematous. Menstrual bleeding into the dermis leads to hemosiderin deposition (which is seen with the Perls stain), scarring and chronic inflammation [32, 33]. The pathologic appearance varies somewhat according to the phase of the cycle (proliferative, secretion, decidualization) [33]. with the presence of endometrial glands and stroma in the mid to deep dermis, as well as possible haemosiderin deposition.⁷ Immunohistochemistry can serve as a helpful tool

in supporting the diagnosis of cutaneous endometriosis. CD10 is diffusely and strongly expressed in the cytoplasm of benign endometrial stroma. CD10 can be particularly helpful in the evaluation of examples of cutaneous endometriosis, in which there is a paucity of glands and abundant stroma, in which there is merely a thin periglandular cuff of stroma, in which the stroma is obscured by histiocytes or in which there is uncertainty about the origin of the stroma. Estrogen receptor and Progesterone receptor immunostains show strong nuclear positivity in the glands and stroma, except in the case of decidualized endometriosis, in which the expression of these antigens can be lost because of the downregulation of hormones.¹⁰

The treatment of choice is radical excision of umbilical endometriotic lesions to prevent recurrences and yields good aesthetic results at an extensive follow-up. Local relapsing is quite rare and would appear on the same location during the first year follow-up.¹¹ The systematic use of laparoscopy aids in the adequate excision of the umbilical lesion and the accurate reconstruction of the fascial and subfascial planes, and also allows the surgeon to identify and eventually treat concomitant pelvic disease.¹² Preoperative hormonal therapy, although sometimes associated with such side effects as amenorrhea, may be used in cases of large endometriotic masses to reduce the size of the surgical defect.¹³

In spite of the good prognosis for this disease, but there were some reports of Cutaneous endometrial cancer arising from heterotopic endometriosis in an abdominal caesarean section scar¹⁴⁻¹⁶ only 0.3-1% in secondary endometriosis.¹⁷ The most common reported malignant arising on cutaneous endometriosis is endometrioid carcinoma followed by clear-cell carcinoma, sarcomas and serous carcinoma.⁷

In conclusion, this patient was diagnosed as cutaneous endometriosis which is most likely to be primary cutaneous endometriosis. Although she had ever been done previous caesarean sections, the lesion did not located on the surgical scar, so she was undergone wide excision of the nodule and found that the mass extended downward to the rectus sheath intraoperatively. It was completely excised with rectus sheath closure. There was no recurrence at 3 month postoperatively but long term follow up is needed.

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