Primary Cutaneous Endometriosis of the Umbilicus: A Case Report

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Cutaneous endometriosis is a rare condition, especially in patients without a history of abdominal or pelvic surgery or known preexisting endometriosis. Most cases present with cyclic pain and bleeding at the site of an umbilical cutaneous nodule correlating with menses. We present an atypical case of primary cutaneous endometriosis of the umbilicus without a prior history of abdominal or pelvic surgery and without cyclic pain or bleeding.

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Defined simply as ectopic endometrial tissue, *endometriosis* is a relatively common condition, occurring in 8% to 15% of menstruating women.[1] The most common sites involved are the ovaries, uterine ligaments, rectovaginal septum, and peritoneum[2]; however, endometriosis has presented elsewhere, including the intestines, appendix, lungs, and kidneys.[3] Cutaneous endometriosis is a rare variant, accounting for approximately 1% of cases of endometriosis, and is most prevalent at sites of surgical scars from cesarean section, episiotomy, hysterectomy, laparoscopic surgery, and other abdominal surgeries.[1-5] Primary umbilical endometriosis, with no preexisting pelvic endometriosis or previous abdominal or pelvic surgery, is even less common.[4]

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Case Report

A 30-year-old woman presented with a soft, 1.5-cm, flesh-colored, oval-shaped nodule at the right inferiolateral aspect of the umbilicus that had steadily increased in size for 3 months prior to presentation to us (Figure 1). The patient reported an intermittent, slight aching of the lesion and it had become somewhat tender to palpation. She denied bleeding or discharge from the nodule or the presence of similar lesions elsewhere on the body. The nodule and related symptoms did not undergo any changes in correlation to menses. The patient's prior medical history was positive for oligomenorrhea, which was managed with oral contraceptives for 7 years; she discontinued the oral contraceptives 1 year prior to presentation. She denied any history of dysmenorrhea, menorrhagia, dyspareunia, or pregnancy. The patient had a 30-minute episode of severe periumbilical pain 6 months prior to presentation. Abdominal workup revealed no cause for the pain. Pelvic examination demonstrated a 2- to 3-cm soft, nontender, round mass located posterior to the urethra. Pelvic magnetic resonance imaging revealed a 1.9×1.5×1.4-cm elliptical mass just posterior to the urethra. The lesion was excised and found to be an epidermal inclusion cyst. Endocrinology workup test results, including luteinizing hormone, follicle stimulating hormone, total and free testosterone, prolactin, and thyroid stimulating hormone levels, were within reference range. Constitutional symptoms were absent. There was no history of abdominal surgery or trauma at the site of the lesion. There was no family history of gynecologic disease or malignancy. Histopathologic examination of the umbilical nodule revealed endometrial stromata and glands, extravasated red blood cells, and focal areas of necrotic tissue in the mid to lower dermis (Figure 2). The lesion was subsequently excised without sequelae.

Comment

The pathogenesis of endometriosis is not definitively known. The majority of cases of cutaneous endometriosis develop at the sites of surgical scars,

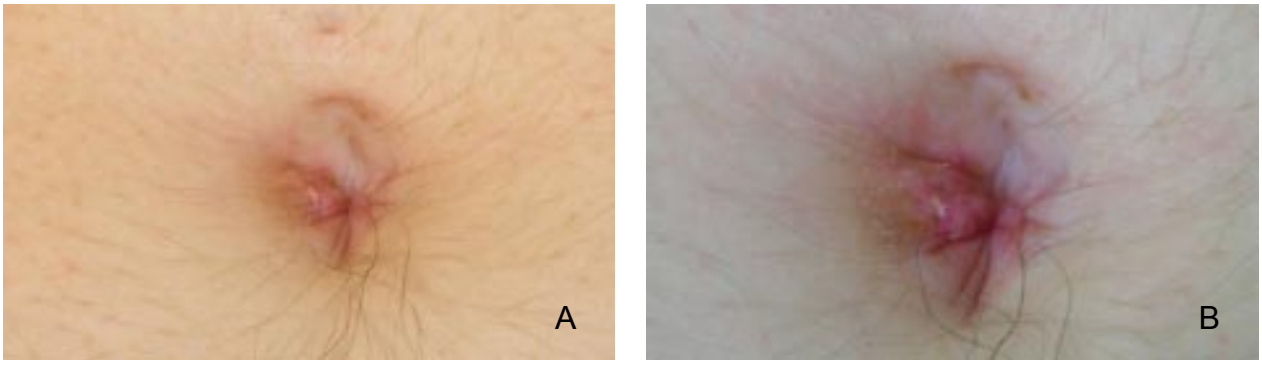

Figure 1. Slightly erythematous umbilical nodule 14 days after a biopsy from the center of the nodule (A and B). Sutures had just been removed.

particularly following procedures involving the uterus.[1,2,5,6] Physical transplantation of endometrial cells during surgery seems to be the most likely mechanism of development. However, cutaneous endometriosis can occur in women without a history of surgery, usually appearing in the umbilicus. Cutaneous endometriosis also has occurred in women without intrapelvic endometriosis and without a history of symptoms suggestive of endometriosis.[7,8] The pathogenesis of such cases is even less clear. Proposed mechanisms include spread of existing endometrial cells via lymphatics or blood, or de novo development from pluripotent cells of the coelom.[5,6]

The mean time to histologic diagnosis of cutaneous endometriosis is more than 2 years from symptom onset.[1] The differential diagnosis of a new enlarging nodule at the umbilicus is broad, including metastasis from an intra-abdominal carcinoma (eg, Sister Mary Joseph nodule), omphalomesenteric duct cyst, primary malignant mesothelioma, metastatic peritoneal malignant mesothelioma, melanoma, various granulomatous

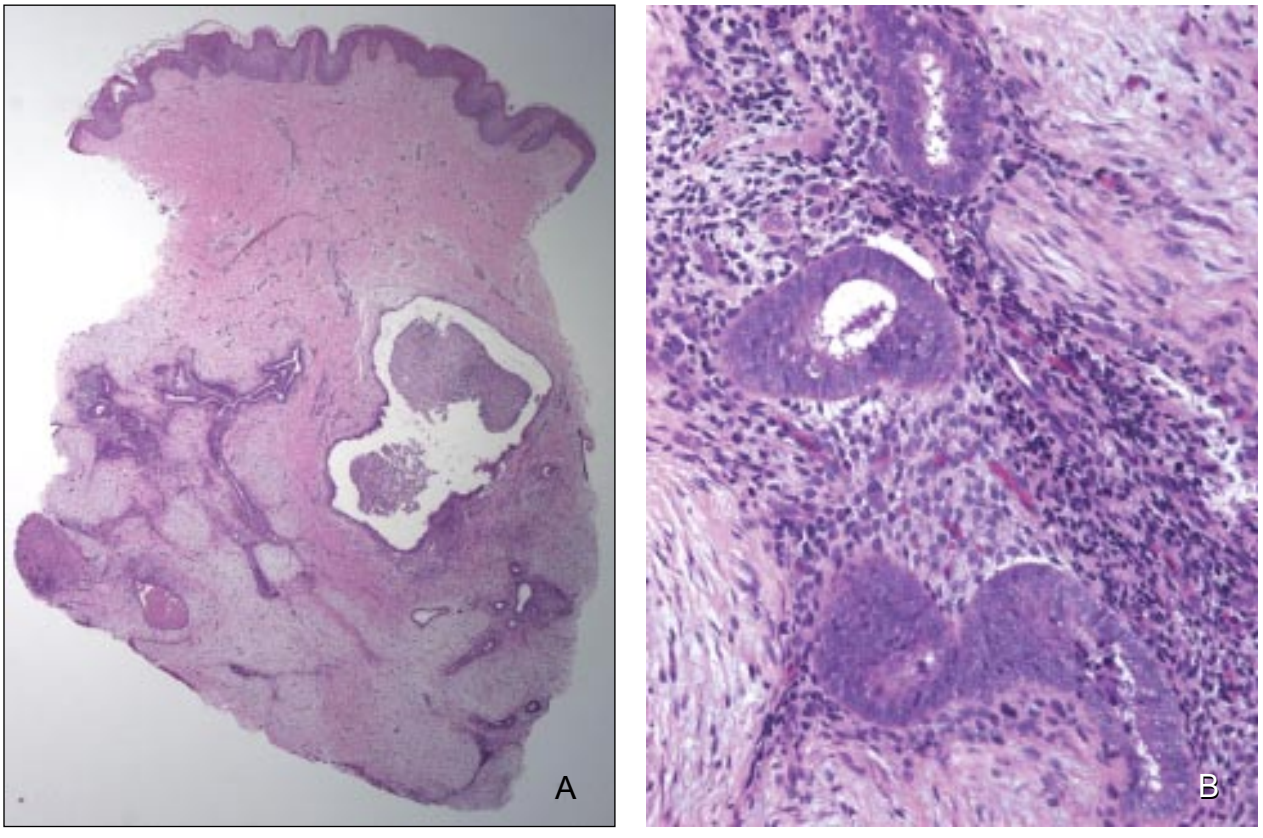

Figure 2. Endometrial stromata and glands, extravasated red blood cells, and focal areas of necrotic tissue were present in the mid to lower dermis (H&E; original magnifications ×2 and ×20, respectively)(A and B).

tumors, umbilical hernia, keloid, pyogenic granuloma, hemangioma, desmoid tumor, umbilical polyp, epidermal inclusion cyst, melanocytic nevus, seborrheic keratosis, granular cell tumor, embryologic rest, basal cell carcinoma, squamous cell carcinoma, and metastatic extra-abdominal carcinoma.[9-15] Classically, women found to have cutaneous endometriosis report a slowly enlarging nodule, with cyclic pain and bleeding at the site corresponding with menses. Even without these characteristic symptoms, as in our patient, cutaneous endometriosis should be considered in the differential diagnosis of an umbilical nodule. While the risk for malignant transformation of cutaneous endometriosis is very low, rare cases have been reported,[4] making prompt diagnosis and subsequent surgical treatment important for preventing unnecessary morbidity and mortality.

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