

JAMA Dermatology Clinicopathological Challenge

A Solitary Umbilical Nodule

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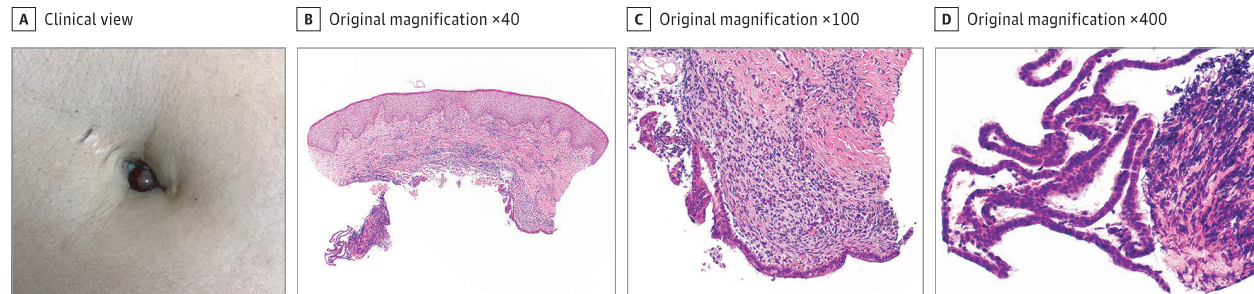


Figure. Clinical image and punch biopsy specimen. A, Smooth reddish-brown nodule within the umbilicus. B, Low-power view showing glandular and stromal tissue within the dermis (hematoxylin-eosin). C, Higher-power view demonstrating a spindle-cell stroma with hemorrhage and hemosiderin-laden macrophages (hematoxylin-eosin). D, Glandular epithelium showing prominent decapitation secretion (hematoxylin-eosin).

A woman in her 40s with no significant medical history presented with a 1-year history of an umbilical nodule. She described occasional pain and intermittent bleeding from the site. Prior treatment with intralesional triamcinolone injections for a presumed keloid yielded no improvement. Physical examination showed a discrete, reddish-brown, noncompressible, smooth nodule within the umbilicus (Figure, A). There was no change in size with the Valsalva maneuver.

Histopathologic examination revealed stromal tissue and glandular epithelium within the dermis (Figure, B). The stroma tissue was characterized by small spindle cells, hemorrhage, and numerous hemosiderin-laden macrophages (Figure, C). Glandular epithelium composed of basophilic cuboidal cells and showing decapitation secretion was also present (Figure, D). Computed tomography scan of the abdomen showed a well-circumscribed nodule within the umbilicus without intrabdominal extension or herniation. Surgical excision followed by umbilical reconstruction was performed.

WHAT IS YOUR DIAGNOSIS?

- A. Metastatic adenocarcinoma
- B. Primary umbilical endometriosis
- C. Urachal duct cyst
- D. Omphalomesenteric duct remnant

Diagnosis

B. Primary umbilical endometriosis

Discussion

Endometriosis is a condition characterized by the presence of functional endometrial tissue outside of the uterine cavity. It occurs in 10% of women of reproductive age and most commonly occurs in the pelvic organs.¹ While extrapelvic endometriosis is rare, numerous sites of involvement have been reported, including the eyes, brain, lungs, bowel, and skin.²

Umbilical endometriosis comprises 0.5% to 1% of all extrapelvic cases.³ In secondary umbilical endometriosis, there is iatrogenic implantation of endometrial tissue into the umbilicus following surgery, most often a laparoscopic procedure.⁴ Primary umbilical endometriosis, which occurs in the absence of preceding surgery, is much less common. While the pathogenesis of the primary form is unknown, dissemination of endometrial cells via vascular or lymphatic channels has been suggested.^{5,6} Scar tissue may be particularly susceptible to this endometrial cell migration. In the present case, the endometrioma occurred within an umbilical piercing scar.

Although it is unclear if cosmetic piercings increase the risk of umbilical endometriosis, one similar case has been reported.⁷

Umbilical endometriosis is characterized by a reddish-brown nodule and may clinically resemble a keloid, urachal duct cyst, omphalomesenteric duct remnant, metastatic adenocarcinoma, abdominal hernia, or nodular melanoma.⁸ A key feature that distinguishes umbilical endometriosis from these other entities is pain and bleeding that coincides with menstrual periods. Although these symptoms are considered pathognomonic for endometriosis when they occur cyclically with menses, histopathologic analysis is mandatory for diagnosis. If any suspicion for abdominal hernia exists after thorough physical examination, ultrasound or computed tomography scan should be obtained prior to biopsy to prevent perforation of underlying bowel, which may be complicated by severe infection or even death.⁹ Examination findings suggestive of an uncomplicated hernia include a reducible umbilical mass that expands with the Valsalva maneuver.

The general histopathologic features of cutaneous endometriosis are the presence of both stroma and glandular tissue.¹⁰ The stroma is characterized by small spindle cells, edema, and many

hemosiderin-laden macrophages as a result of prior hemorrhage. The glandular epithelium is composed of basophilic cuboidal or tall columnar cells. Importantly, the histopathologic features of endometriosis vary depending on the menstrual phase during which biopsy is obtained. Endometrial glands may show marked mitotic activity during the proliferative phase, while they demonstrate decapitation secretion with little mitotic activity during the secretory phase. A CD10 immunohistochemical stain may be used to aid in diagnosis because it is a highly sensitive marker for endometrial stroma. Estrogen receptor and progesterone receptor immunostains show strong nuclear positivity, and a cytokeratin 7–positive/cytokeratin 20–negative profile is also observed. Importantly, this cytokeratin staining pattern may also occur in gastric adenocarcinoma, the most common cause of umbilical metastasis in men. A CDX2 immunostain may be used to reliably exclude metastases of intestinal origin when histopathologic results show atypia.

Collectively, the histopathologic findings of endometriosis readily distinguish it from other conditions that may affect the umbilicus. An umbilical metastatic adenocarcinoma, also called Sister Mary Joseph nodule, would show atypical cells of the viscera of origin. A urachal duct cyst, which is an embryonic remnant between the ura-

chus and bladder, would show a cystic structure lined by flattened urothelium with the surrounding dermis exhibiting fibrosis and calcification. An omphalomesenteric duct consists of remnants of the vitelline duct, which connects the small intestine to the yolk sac in early fetal development and normally involutes by birth. Histopathologic results would show a cystic structure lined by enteric mucosa. Nodular melanoma may also present as a reddish-brown umbilical nodule but is characterized microscopically by dermally based atypical melanocytes.

The treatment of choice for umbilical endometriosis is surgical excision.³ Danazol has also been successfully used to alleviate pain and bleeding.⁵ Because long-term use of antigonadotropin therapy typically leads to antiestrogenic adverse effects, this treatment is more commonly used as a short-term adjunct to decrease endometrioma size preoperatively.

Primary umbilical endometriosis is a rare but well-described clinical entity with unique histopathologic features. While the diagnosis should be suspected in an umbilical nodule that cyclically bleeds during menses, histopathologic evaluation is mandatory to exclude nodular melanoma, metastatic adenocarcinoma, and other malignant tumors.

ARTICLE INFORMATION

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