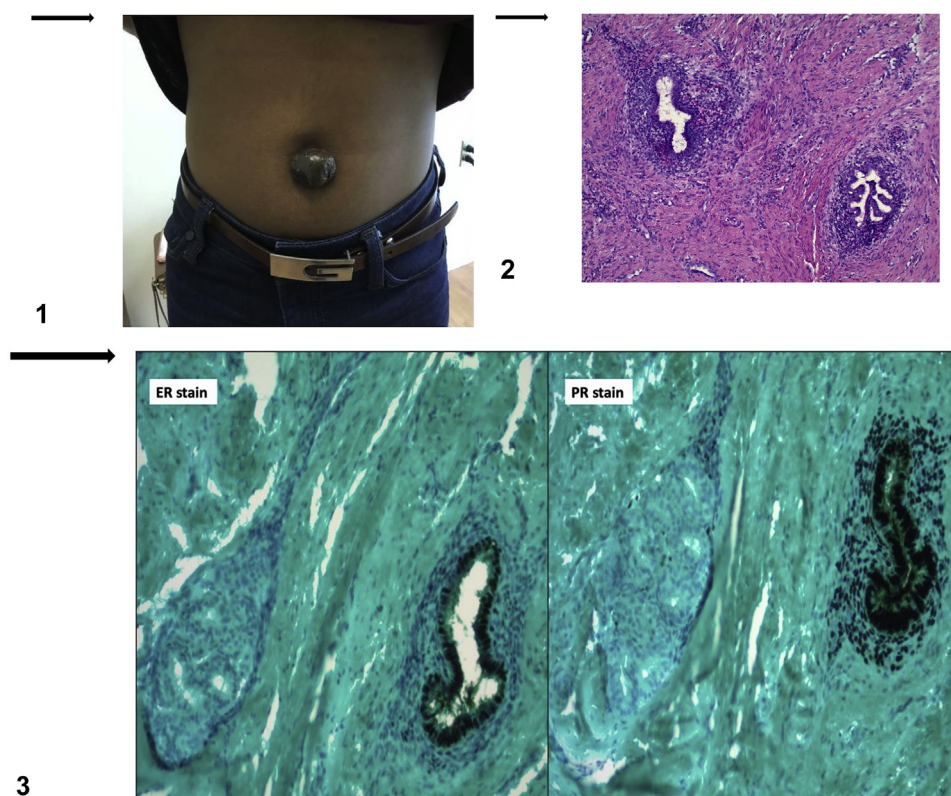


## A solitary brown nodule on the umbilicus



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**Key words:** keloid; melanoma; nodule; pyogenic granuloma; umbilical endometriosis.



A 29-year-old black woman presented with complaints of a firm, moderately tender mass on her umbilicus. She stated that the lesion appeared 2 years ago, slowly increased in size, and intermittently bled with monthly periods. She denied previous trauma, piercings, or surgeries. Physical examination found a solitary well-circumscribed 1.5-cm brown, firm, immobile, nonhemorrhagic nodule on the superior aspect of the umbilicus (Fig 1). A 4-mm punch biopsy found multiple foci of glands lined by ciliated columnar and cuboidal epithelium, surrounding stroma-contained hemosiderin-laden macrophages (Fig 2). Doppler ultrasound scan showed minimal vascular flow. Magnetic resonance imaging without contrast of abdomen/pelvis showed no visceral involvement.

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**Question 1: What is the most likely diagnosis?**

- A. Keloid
- B. Primary cutaneous endometriosis
- C. Lobular capillary hemangioma
- D. Nodular melanoma
- E. Dermatofibrosarcoma

**Answers:**

**A.** Keloid — Incorrect. Because the patient is of African ancestry, she is at a higher risk for spontaneous keloid formation. Histology finds whorls and nodules of thick, smooth glassy, “bubble gum” collagen fibers haphazardly arranged throughout dermis.

**B.** Primary cutaneous endometriosis — Correct. Cutaneous endometriosis makes up 0.5% to 1% of all ectopic endometriosis; less than 10 cases of primary cutaneous endometriosis of the umbilicus have been described in literature.<sup>1</sup> Cyclical bleeding/pain should clue clinicians to this diagnosis.<sup>1</sup> Histology finds multiple foci of glands lined by ciliated columnar and cuboidal epithelium recapitulating endometrial glands, surrounded by hemosiderin-laden macrophages.<sup>2</sup> Doppler ultrasound scan typically finds scarce vascularity with low flow; computed tomography/magnetic resonance imaging typically find no visceral connection.<sup>3</sup> This was the case for our patient’s lesion.

**C.** Lobular capillary hemangioma — Incorrect. Lobular capillary hemangiomas may bleed with trauma. Histology shows an exophytic nodule with an ulcerated epidermis and multiple lobules of capillaries separated by fibrous chords in the dermis.

**D.** Nodular melanoma — Incorrect. Nodular melanoma should be high on the clinician’s differential diagnosis. Histology shows a relatively well-circumscribed, mostly dermal tumor with multiple aggregates of atypical melanocytes and mitoses throughout dermis and superficial subcutaneous fat.

**E.** Dermatofibrosarcoma — Incorrect. Dermatofibrosarcoma usually presents as a slow-growing, skin-colored, indurated plaque that develops violaceous to red-brown nodules measuring from one to several centimeters in diameter. Dermatofibrosarcoma has a predilection for proximal extremities and pelvis. Histology shows multiple bland spindle cell infiltrates in a storiform pattern throughout the dermis, with infiltration of the fat in characteristic honeycomb pattern.

**Question 2: What would be the histologic staining pattern of this biopsy?**

- A. Factor XIII<sup>A</sup><sup>-</sup>/CD34<sup>-</sup>
- B. CK7<sup>+</sup>/CD10<sup>+</sup>/ estrogen receptor (ER)<sup>+</sup>/progesterone receptor (PR)<sup>+</sup>
- C. CD31<sup>+</sup>/CD34<sup>+</sup>/vimentin<sup>+</sup>
- D. S100<sup>+</sup>/Melan-A<sup>+</sup>/HMB45<sup>+</sup>/SOX10<sup>+</sup>
- E. Factor XIII<sup>A</sup><sup>-</sup>/ CD34<sup>+</sup>

**Answers:**

**A.** Factor XIII<sup>A</sup><sup>-</sup>/CD34<sup>-</sup> — Incorrect. This answer is incorrect, as it is the staining pattern of a keloid. No keloidal collagen is seen on hematoxylin-eosin staining.

**B.** CK7<sup>+</sup>/CD10<sup>+</sup>/ER<sup>+</sup>/PR<sup>+</sup> — Correct. In primary cutaneous endometriosis the stroma stains shows CD10<sup>+</sup> while endometrial glands stain CK7<sup>+</sup>. CD10 is particularly helpful in the evaluation 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 the origin of stroma is questionable. CK7 stains positively more than 95% of the time in primary endometrial adenoid tumors and helps distinguish primary from metastatic tumors.<sup>2</sup> The endometrial glands demonstrate ER<sup>+</sup>, PR<sup>+</sup> staining.<sup>2</sup> Our patient’s tumor stained positive for ER and PR (Fig 3).

**C.** CD31<sup>+</sup>/CD34<sup>+</sup>/vimentin<sup>+</sup> — Incorrect. This answer is incorrect, as this is the staining pattern of a lobular capillary hemangioma.

**D.** S100<sup>+</sup>/Melan-A<sup>+</sup>/HMB45<sup>+</sup>/SOX10<sup>+</sup> — Incorrect. This is the staining pattern for a malignant melanoma. No atypical nevus cells seen on biopsy.

**E.** Factor XIII<sup>A</sup><sup>-</sup>/ CD34<sup>+</sup> — Incorrect. Although this is a scar-like process, this would be the staining pattern for a dermatofibrosarcoma. The histologic pattern of biopsy does not suggest this.

**Question 3: What is the best treatment modality for this patient?**

- A. Hormonal treatment including gonadotropin-releasing hormone, oral contraceptives, or danazol
- B. Early local excision of the lesion with 3- to 4-mm margins with layered closure and sparing of the umbilicus

- C.** Conservative wound care with chlorhexidine 4% wash every other day
- D.** Surgical excision with complete resection of the umbilicus
- E.** Treatment of vascular lesion with pulsed dye laser

**Answers:**

- A.** Hormonal treatment including gonadotropin-releasing hormone, oral contraceptives, or danazol — Incorrect. Although these lesions can be treated with hormone therapy, this is an adjunct therapy used for debulking presurgically and to reduce recurrence postsurgically.
- B.** Early local excision of the lesion with 3- to 4-mm margins with layered closure and sparing of the umbilicus — Incorrect. Although this may be a less invasive, this treatment increases the chance of recurrences.<sup>4</sup>
- C.** Conservative wound care with chlorhexidine 4% wash every other day — Incorrect. Although this treatment may prevent secondary infection of the lesion, it will not provide a definitive cure or treatment for this lesion.
- D.** Surgical excision with complete resection of the umbilicus — Correct. There is no standard of

treatment for umbilical endometriosis, the consensus for treatment involved complete excision of lesion and total umbilical resection down to fascia and reconstruction. This treatment provides the lowest chance for recurrence and provides good cosmesis depending on flap used.<sup>5</sup>

**E.** Treatment of vascular lesion with pulsed dye laser — Incorrect. This is not the standard of care of this lesion, as it is not a primarily vascular lesion.

**Abbreviations used:**

ER: estrogen receptor

PR: progesterone receptor

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