

Tender Nodule on the Umbilicus

Alison C. Lam, MD; Peter Ng, MD; Willis H. Lyford, MD; Curtis L. Hardy, DO; Shaun P. Ostrofe, DO

Eligible for 1 MOC SA Credit From the ABD

This Photo Challenge in our print edition is eligible for 1 self-assessment credit for Maintenance of Certification from the American Board of Dermatology (ABD). After completing this activity, diplomates can visit the ABD website (<http://www.abderm.org>) to self-report the credits under the activity title "Cutis Photo Challenge." You may report the credit after each activity is completed or after accumulating multiple credits.



A 25-year-old woman was referred to the dermatology clinic by her primary care provider for evaluation of a tender nodule on the inferior umbilicus of 2 years' duration at the site of a preexisting keloid scar. The patient reported that the lesion caused occasional pain and tenderness. A few weeks prior to the current presentation, a dark-red bloody discharge developed at the superior aspect of the lesion that subsequently crusted over. The patient denied any use of oral contraceptives or history of abdominal surgery.

The original keloid scar had been treated successfully by an outside physician with intralesional steroid injections, and the patient was interested in a similar procedure for the current nodule. She also had a history of a hyperpigmented hypertrophic scar on the superior periumbilical area from a previous piercing that had resolved several years prior to presentation.

Physical examination of the lesion revealed a 1.2-cm, soft, tender, violaceous nodule with scant yellow crust along the superior surface of the umbilicus. There was no palpable abdominal wall defect, and the nodule was not reducible into the abdominal cavity. An interval history revealed bleeding of the lesion during the patient's menstrual cycle with persistent pain and tenderness. A punch biopsy was performed.

WHAT'S YOUR DIAGNOSIS?

- a. dermatofibrosarcoma protuberans
- b. lobular capillary hemangioma
- c. Sister Mary Joseph nodule
- d. umbilical hernia
- e. Villar nodule

PLEASE TURN TO **PAGE 129** FOR THE DIAGNOSIS

Dr. Lam is from the I Marine Expeditionary Force Information Group, Camp Pendleton, California. Dr. Ng is from the Family Medicine Department, Navy Medical Readiness and Training Command, Camp Pendleton. Drs. Lyford, Hardy, and Ostrofe are from the Dermatology Department, Naval Medical Center, San Diego, California.

The authors have no relevant financial disclosures to report.

The views expressed in this article are those of the authors and do not necessarily reflect the official policy or position of the Department of the Navy, Department of Defense, or the United States Government.

Correspondence: Alison C. Lam, MD, 9th Communication Battalion, I Marine Expeditionary Force Information Group, I Marine Expeditionary Force, Box 555351, Camp Pendleton, CA (alison.lam@comcast.net).

Cutis. 2025 October;116(4):123, 129-130, 137. doi:10.12788/cutis.1273

THE DIAGNOSIS:

Villar Nodule

The biopsy revealed features consistent with cutaneous endometriosis in the setting of a painful, tender, multilobulated nodule with a cyclical bleeding pattern (Figure 1). The bleeding pattern of the nodule during menses and lack of surgical history supported the diagnosis of primary cutaneous endometriosis in our patient. She was diagnosed with endometriosis by gynecology, and her primary care physician started her on an oral contraceptive based on this diagnosis. She also was referred to gynecology and plastic surgery for a joint surgical consultation to remove the nodule. She initially decided to do a trial of the oral contraceptive but subsequently underwent umbilical endometrioma excision with neo-umbilicus creation with no evidence of recurrence.

Primary cutaneous endometriosis should be considered in young females who present with tender umbilical nodules. Endometriosis refers to the presence of an endometriumlike epithelium outside the endometrium and myometrium.¹ The condition affects 10% to 15% of reproductive-aged (ie, 18-49 years) women in the United States and typically involves tissues within the pelvis, such as the ovaries, pouch of Douglas, or pelvic ligaments.² Cutaneous endometriosis is the growth of endometrial tissue in the skin and is rare, accounting for less than 5.5% of cases of

extrapelvic endometriosis worldwide, affecting primarily the umbilicus, abdominal wall, and vulva.^{3,4}

The 2 main types of cutaneous endometriosis are primary (spontaneous) and secondary. Primary lesions develop in patients without prior surgical history, and secondary lesions occur within previous surgical incision sites, often scars from cesarean delivery.⁵ Less than 30% of cases of cutaneous endometriosis are primary disease.⁶ Primary cutaneous endometriosis of the umbilicus, known as Villar nodule, was first described in 1886.^{3,7} Up to 40% of patients with extrapelvic endometriosis worldwide presented with Villar nodules in a systematic literature review.⁶ The prevalence of these nodules is unknown, but the incidence is less than 1% of cases of extragenital endometriosis.⁴

There are 2 leading theories of primary cutaneous endometriosis pathogenesis. The first is the transportation theory, in which endometrial cells are transported outside the uterus via the lymphatic system.⁸ The second is the metaplasia theory, which proposed that endometrial cells develop in the coelomic mesothelium in the presence of high estrogen levels.^{8,9}

Secondary cutaneous endometriosis, also known as scar endometriosis, is suspected to be caused by an iatrogenic implantation of endometrial cells at the scar of a prior surgical site.⁹ Although our patient had an existing umbilicus scar from a piercing, it was improbable for that to have been the nidus, as the keloid scar was superficial and did not have contact with the abdominal cavity for iatrogenic implantation. Clinical diagnosis for secondary cutaneous endometriosis often is made based on a triad of features: a nonmalignant abdominal mass, recurring pain and bleeding of the lesion with menses, and prior history of abdominal surgery.^{9,10} On clinical examination, these features typically manifest as a palpable subcutaneous mass that is black, blue, brown, or red. Often, the lesions enlarge and bleed during the menstrual cycle, causing pain, tenderness, or pruritus.³ Dermoscopic features of secondary cutaneous endometriosis are erythematous umbilical nodules with a homogeneous vascular pattern that appears red with a brownish hue (Figure 2).^{9,11} Dermoscopic features may vary with the hormone cycle; for example, the follicular phase (correlating with day 7 of menses) demonstrates polypoid projections, erythematous violaceous color, dark-brown spots, and active bleeding of the lesion.¹² Clinical and dermoscopic examination are useful tools in this diagnosis.

Imaging such as ultrasonography, computed tomography, or magnetic resonance imaging may be useful in identifying abdominal endometriomas.^{8,13,14} Pelvic involvement of endometriosis was found in approximately 15% of patients in a case series,⁴ with concurrent

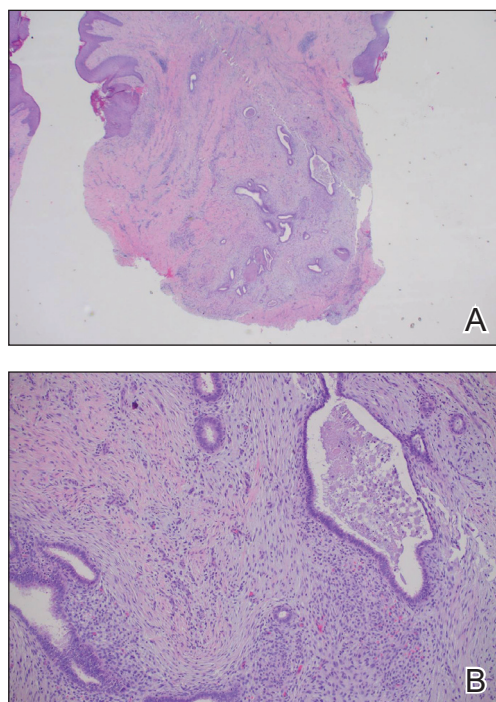


FIGURE 1. A and B, Histopathology revealed glandular spaces and a loose concentric fibromyxoid stroma that contained red blood cells (H&E, original magnifications $\times 2$ and $\times 10$).



FIGURE 2. Dermoscopic examination showed a 1.2-cm violaceous nodule within the umbilicus (original magnification $\times 10$).

primary umbilical endometriosis. Imaging studies may assist evaluation for fistula formation, presence of malignancies, and the extent of endometriosis within the abdominal cavity.

Histopathology is key to confirming cutaneous endometriosis and shows multiple bland-appearing glands of varying sizes with loose, concentric, edematous, or fibromyxoid stroma (Figure 1).³ Red blood cells sometimes are found with hemosiderin within the stroma. Immunohistochemical staining with estrogen receptors may aid in identifying the endometriumlike epithelial cells.¹³

Standard treatment involves surgical excision with 1-cm margins and umbilical preservation, which results in a recurrence rate of less than 10%.^{4,10} Medical therapy, such as aromatase inhibitors, progestogens, antiprogesterones, combined oral contraceptives, or gonadotropin-releasing hormone agonists or antagonists may help manage pain or reduce the size of the nodule.^{4,15} Simple observation also is a potential course for patients who decline treatment options.

Differential diagnoses include lobular capillary hemangioma, also known as pyogenic granuloma; Sister Mary Joseph nodule; umbilical hernia; and dermatofibrosarcoma protuberans. Lobular capillary hemangiomas commonly are acquired benign vascular proliferations of the skin that are friable and tend to ulcerate.¹⁶ These lesions typically grow rapidly and often are located on the face, lips, mucosae, and fingers. Histopathologic examination may show an exophytic lesion with lobules of proliferating capillaries within an edematous matrix, superficial ulceration, and an epithelial collarette.¹⁷ Treatment includes surgical excision, cauterization, laser treatments, sclerotherapy, injectable medications, and topical medications, but recurrence is possible with any of these interventions.¹⁸

Cutaneous metastasis of an internal solid organ cancer, commonly known as a Sister Mary Joseph nodule, typically manifests as an erythematous, irregularly shaped nodule that may protrude from the umbilicus.¹⁴ Gastrointestinal symptoms such as change in bowel habits or obstructive symptoms in the setting of a progressive malignancy are common.¹⁴ Clinical features include a firm fixed lesion, oozing, and ulceration.¹⁹ On dermoscopy, polymorphous vascular patterns, milky red structureless areas, and white lines typically are present.¹¹ Although dermoscopic features may differentiate this entity from cutaneous endometriosis, tissue sampling and histologic examination are crucial diagnostic tools to identify malignant vs benign lesions.

An umbilical hernia is a protrusion of omentum, bowel, or other intra-abdominal organs in an abdominal wall defect. Clinical presentation includes a soft protrusion that may be reduced on palpation if nonstrangulated.²⁰ Treatment includes watchful waiting or surgical repair. The reducibility and presence of an abdominal wall defect may point to this diagnosis. Imaging also may aid in the diagnosis if the history and physical examination are unclear.

Dermatofibrosarcoma protuberans is a slow-developing, low- to intermediate-grade, soft-tissue sarcoma that occurs in less than 0.1% of all cancers in the United States.²¹ Lesions often manifest as small, firm, slow-growing, painless, flesh-colored dermal plaques; subcutaneous thickening; or atrophic nonprotuberant lesions typically involving the trunk.²¹ Histopathologically, they are composed of uniform spindle-cell proliferation growing in a storiform pattern and subcutaneous fat trapping that has strong and diffuse CD34 immunoreactivity.^{21,22} Pathologic examination typically distinguishes this diagnosis from cutaneous endometriosis. Treatment includes tumor resection that may or may not involve radiotherapy and targeted therapy, as recurrence and metastases are possible.

Primary cutaneous endometriosis is a rare but important diagnosis for dermatologists to consider when evaluating umbilical nodules. Clinical features may include bleeding masses during menses in females of reproductive age. Dermoscopic examination aids in workup, and histopathologic testing can confirm the diagnosis and rule out malignancies. Surgical excision is the treatment of choice with a low rate of recurrence.

REFERENCES

1. International Working Group of AAGL, ESGE, ESHRE and WES; Tomassetti C, Johnson NP, et al. An international terminology for endometriosis, 2021. *Hum Reprod Open*. 2021;2021:hoab029. doi:10.1093/hropen/hoab029
2. Batista M, Alves F, Cardoso J, et al. Cutaneous endometriosis: a differential diagnosis of umbilical nodule. *Acta Med Port*. 2020; 33:282-284. doi:10.20344/amp.10966
3. Brown ME, Osswald S, Biediger T. Cutaneous endometriosis of the umbilicus (Villar's nodule). *Int J Womens Dermatol*. 2020;6:214-215. doi:10.1016/j.ijwd.2020.01.001

CONTINUED ON PAGE 137

CONTINUED FROM PAGE 130

4. Bindra V, Sampurna S, Kade S, et al. Primary umbilical endometriosis - case series and review of clinical presentation, diagnosis and management. *Int J Surg Case Rep.* 2022;94:107134. doi:10.1016/j.ijscr.2022.107134
5. Loh SH, Lew BL, Sim WY. Primary cutaneous endometriosis of umbilicus. *Ann Dermatol.* 2017;29:621-625. doi:10.5021/ad.2017.29.5.621
6. Victory R, Diamond MP, Johns DA. Villar's nodule: a case report and systematic literature review of endometriosis externa of the umbilicus. *J Minim Invasive Gynecol.* 2007;14:23-32. doi:10.1016/j.jmig.2006.07.014
7. Van den Noulant D, Kaur M. Primary umbilical endometriosis: a case report. *Facts Views Vis Obgyn.* 2017;9:115-119.
8. Machairiotis N, Stylianaki A, Dryllis G, et al. Extrapelvic endometriosis: a rare entity or an under diagnosed condition? *Diagn Pathol.* 2013;8:194. doi:10.1186/1746-1596-8-194
9. Huang QF, Jiang B, Yang X, et al. Primary versus secondary cutaneous endometriosis: literature review and case study. *Heliyon.* 2023;9:E20094. doi:10.1016/j.heliyon.2023.e20094
10. Gonzalez RH, Singh MS, Hamza SA. Cutaneous endometriosis: a case report and review of the literature. *Am J Case Rep.* 2021;22:E932493. doi:10.12659/AJCR.932493
11. Buljan M, Arzberger E, Šitum M, et al. The use of dermoscopy in differentiating Sister Mary Joseph nodule and cutaneous endometriosis. *Australas J Dermatol.* 2019;60:E233-E235. doi:10.1111/ajd.12980
12. Costa IM, Gomes CM, Morais OO, et al. Cutaneous endometriosis: dermoscopic findings related to phases of the female hormonal cycle. *Int J Dermatol.* 2014;53:E130-E132. doi:10.1111/j.1365-4632.2012.05854.x
13. Mohaghegh F, Hatami P, Rajabi P, et al. Coexistence of cutaneous endometriosis and ovarian endometrioma: a case report. *J Med Case Rep.* 2022;16:256. doi:10.1186/s13256-022-03483-8
14. Raffi L, Suresh R, McCalmont TH, et al. Cutaneous endometriosis. *Int J Womens Dermatol.* 2019;5:384-386. doi:10.1016/j.ijwd.2019.06.025
15. Saunders PTK, Horne AW. Endometriosis: etiology, pathobiology, and therapeutic prospects. *Cell.* 2021;184:2807-2824. doi:10.1016/j.cell.2021.04.041
16. Habif TP. *Clinical Dermatology a Color Guide to Diagnosis and Therapy.* St. Louis, Mo. Elsevier; 2016.
17. Patrice SJ, Wiss K, Mulliken JB. Pyogenic granuloma (lobular capillary hemangioma): a clinicopathologic study of 178 cases. *Pediatr Dermatol.* 1991;8:267-276. doi:10.1111/j.15251470.1991.tb00931.x
18. Kaleeny JD, Janis JE. Pyogenic granuloma diagnosis and management: a practical review. *Plast Reconstr Surg Glob Open.* 2024;12:E6160. doi:10.1097/GOX.00000000000006160
19. Ha DL, Yang MY, Shin JO, et al. Benign umbilical tumors resembling Sister Mary Joseph nodule. *Clin Med Insights Oncol.* 2021;15:1179554921995022. doi:10.1177/1179554921995022
20. Lawrence PF, Smeds M, Jessica Beth O'connell. *Essentials of General Surgery and Surgical Specialties.* Wolters Kluwer Health; 2019.
21. Hao X, Billings SD, Wu F, et al. Dermatofibrosarcoma protuberans: update on the diagnosis and treatment. *J Clin Med.* 2020;9:1752. doi:10.3390/jcm9061752
22. Allen A, Ahn C, Sangüeza OP. Dermatofibrosarcoma protuberans. *Dermatol Clin.* 2019;37:483-488. doi:10.1016/j.det.2019.05.006