Solitary Lesion on the Umbilicus

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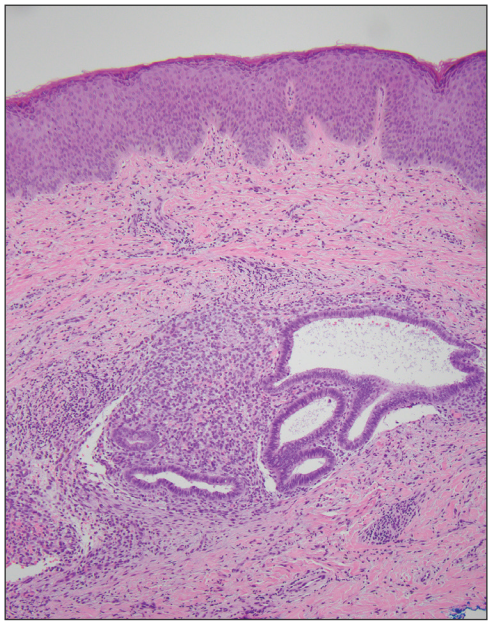

H&E, original magnification ×100.

A 33-year-old woman with no notable medical or surgical history presented to our clinic with a solitary indurated nodule on the umbilicus that had been progressively enlarging for 1 year. The patient reported that she had undergone piercing of the umbilicus more than 5 years prior. She noted that the lesion was uncomfortable and pruritic and occasionally bled spontaneously. Physical examination revealed no other mucosal or cutaneous findings. A shave biopsy of the nodule was performed.

THE BEST **DIAGNOSIS IS:**

a. cutaneous endometriosis
b. metastatic carcinoma of the breast
c. nodular hidradenoma
d. Sister Mary Joseph nodule
e. urachal duct cyst

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THE **DIAGNOSIS:** Cutaneous Endometriosis

Endometriosis is the ectopic presence of endometrial tissue and occurs in approximately 13% of women of childbearing age.[1] This non-neoplastic lesion can manifest on the skin in less than 5.5% of endometriosis cases worldwide. Historically, secondary cutaneous endometriosis (CE) most frequently has been associated with prior gynecologic surgery (often cesarean section)[2]; however, an increased incidence of primary CE in patients without prior surgical history recently has been documented in the literature.[3] While secondary CE usually manifests at the site of a surgical scar, primary CE has a predilection for the umbilicus (Villar nodule). In both primary and secondary CE, patients present clinically with a solitary nodule and abdominal pain that may be exacerbated during menstruation. Bleeding without associated pain may be more common in primary CE, while bleeding with pain may be more common in secondary CE. Cutaneous endometriosis often is overlooked given its low incidence, leading to delayed diagnosis. Primary CE often is misdiagnosed clinically as a pyogenic granuloma, Sister Mary Joseph nodule, or keloid, while secondary CE may be mistaken for a fibroma, incisional hernia, or granuloma.[2]

Primary and secondary CE have identical histopathologic features. Glands of variable size consisting of a single epithelial layer of columnar cells are present in the reticular dermis or subcutis (quiz image).[4] The accompanying periglandular stroma often is uniform, consisting of spindle-shaped basophilic cells with abundant vascular structures. The stroma may contain moderate numbers of mitotic figures, a chronic inflammatory infiltrate, and extravasated red blood cells. The ectopic tissue may be inactive or display morphologic changes resembling those of the endometrium in the normal menstrual cycle.[4] As the ectopic tissue progresses through the stages of menstruation, the glandular morphology also transforms. The proliferative stage demonstrates increased epithelial mitotic figures, the secretory stage exhibits intraluminal secretion, and during menstruation there are degenerative epithelial cells and evidence of vascular congestion. A mixture of glandular stages may be seen in biopsy results. Robust immunohistochemical expression of CD10 in the endometrial stroma can aid in diagnosis (Figure 1). Estrogen and progesterone receptor immunostaining also shows strong nuclear positivity, except in decidualized tissue.[4] Unlike intestinal glands, endometrial glands do not express CDX2 or CK20.[5] Complete surgical excision of CE usually is curative; however, recurrence has been documented in 10% (3/30) of cases.[2]

Breast carcinoma is the most common internal malignancy associated with cutaneous metastasis and may develop prior to visceral diagnosis. It is possible that tumor cells travel through the communicating networks of the cutaneous lymphatic ducts and the mammary lymphatic plexus; however, cutaneous manifestation often is located on the ipsilateral breast, and therefore tumor expansion rather than true metastasis cannot always be ruled out. On histopathology, findings of breast adenocarcinoma include tumor cells that tend to show either interstitial, nodular, mixed, or intravascular growth patterns (Figure 2). Tumor cells may invade the stroma in clusters or as individual cells. Sites of distant metastasis may show an increased likelihood of vascular and lymphatic invasion.[6]

Nodular hidradenoma often manifests as a solitary nodule in the head or neck region, predominantly in women.[7] Pathology shows well-demarcated intradermal

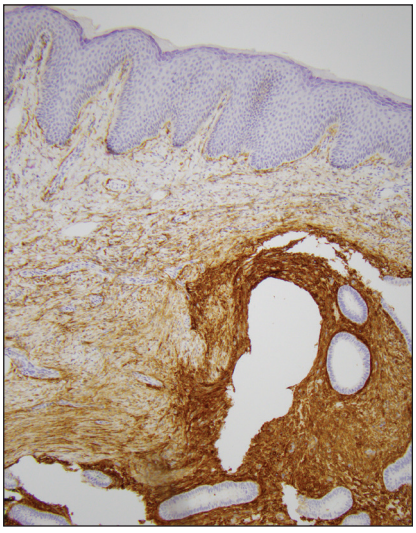

FIGURE 1. Cutaneous endometriosis shows CD10 expression in the stroma on immunohistochemical staining (original magnification ×100).

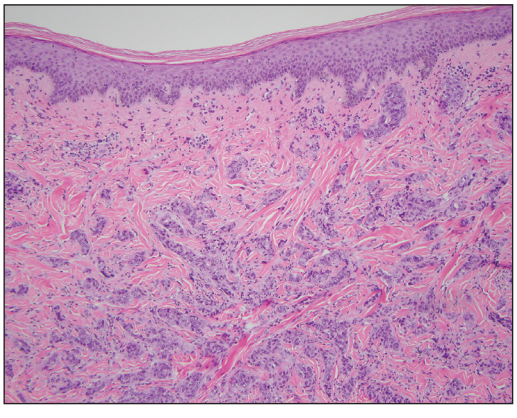

FIGURE 2. Metastatic breast carcinoma shows interstitial aggregates of tumor cells in the superficial and deep dermis (H&E, original magnification ×100).

aggregates of tumor cells within a hyalinized stroma; connection to the epidermis is not a feature of nodular hidradenoma. The epithelial component consists of polygonal cells with eosinophilic to amphophilic cytoplasm as well as large glycogenated cells with pale to clear cytoplasm (leading to the alternative term *clear cell hidradenoma*)(Figure 3). The cystic portion represents deterioration of tumor cells. Surgical excision usually is curative, although lesions may recur. Malignant transformation is rare.[7]

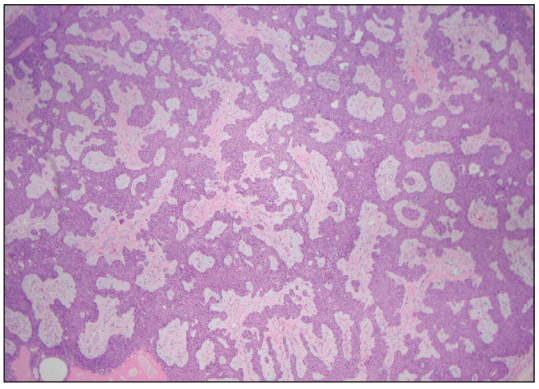

FIGURE 3. Nodular hidradenoma shows a large, well-demarcated, intradermal nodule composed of aggregates of monomorphic tumor cells (H&E, original magnification ×40).

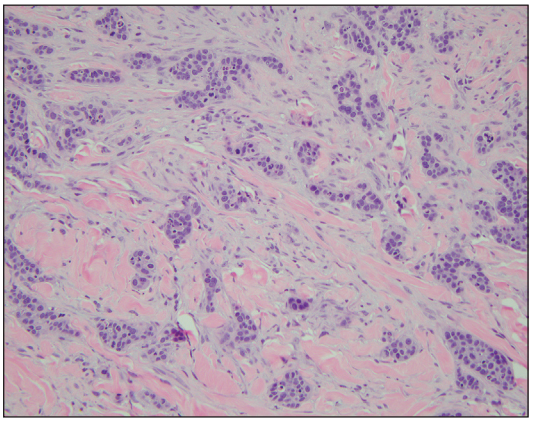

FIGURE 4. Sister Mary Joseph nodule shows dermal invasion of atypical squamous cells, replicating the primary tumor of cervical squamous cell carcinoma (H&E, original magnification ×100).

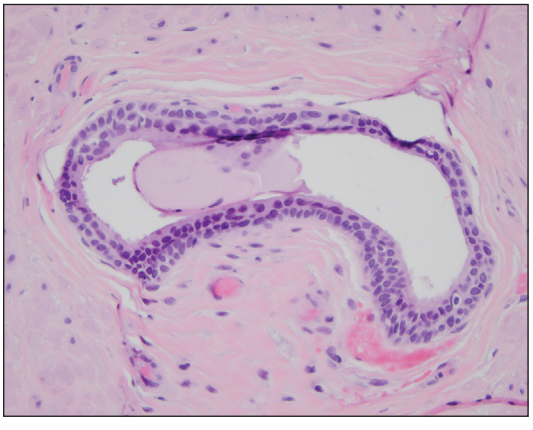

FIGURE 5. Urachal duct cyst shows cystically dilated structures lined with transitional epithelium that intraluminal secretions (H&E, original magnification ×400).

Sister Mary Joseph nodule is a cutaneous involvement of the umbilicus by a metastatic malignancy, often from an intra-abdominal primary malignancy (most commonly ovarian carcinoma in women and colonic carcinoma in men). Clinically, patients present with a solitary firm nodule or plaque within the umbilicus.[8,9] Histopathology recapitulates the primary tumor (Figure 4).[9] Sister Mary Joseph nodule portends a poor prognosis, with a survival rate of less than 8 months from the time of diagnosis.[10]

Urachal duct cyst develops from a remnant of the urachus that closed appropriately at the umbilicus and bladder but did not completely regress. It may manifest as an extraperitoneal mass at the umbilicus. Clinically, urachal duct cysts may be asymptomatic until an inciting event (eg, inflammation, deposition of calculus, or malignancy) occurs.[11] Histopathology shows cystically dilated structures lined with a transitional epithelium (Figure 5).[12] Urachal duct cysts usually are diagnosed in children or young adults and subsequently are excised.[11]

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