

Gastric Carcinoma of the Umbilicus: Case Report of Sister Mary Joseph Nodule

Talley B. Whang, BS; Lina Wang, MD, MS; David H. Peng, MD, MPH

Sister Mary Joseph nodule is a metastatic lesion of the umbilicus, which is an uncommon phenomenon that carries an ominous prognosis. We describe a patient with gastric cancer who presented with asymptomatic papules on the umbilicus proven to be metastatic foci by biopsy. The lesion represented spread of the malignancy despite chemotherapy; thus, treatment was initiated with second-line agents. Sister Mary Joseph nodule can be the first presenting sign of an occult malignancy, or as demonstrated in this case, a sign of disease progression. Given the variable appearance of the lesions, a high index of suspicion is necessary to make an accurate diagnosis.

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Case Report

A 63-year-old man presented with asymptomatic bumps on the umbilicus of 2 weeks' duration, with no associated history of trauma, discharge, or ulceration. Topical treatments prescribed by a primary care physician initially were used for fungal disease, but no improvement was seen in the 2 weeks preceding presentation to the dermatology department. Five months prior to presentation, the patient was diagnosed with gastric cancer with peritoneal carcinomatosis. The patient was being treated with a chemotherapy regimen of oxaliplatin, 5-fluorouracil, and folinic acid (FOLFOX-6), and computed tomography obtained 6 weeks prior to this presentation demonstrated stable disease.

Physical examination revealed a pinkish tan papillomatous plaque around the umbilicus with

an indurated dermis (Figure 1). Foci of necrosis as well as a satellite lesion were present. There was no lymphadenopathy or organomegaly.

A 4-mm punch biopsy was performed. The biopsy specimen demonstrated superficial dermal infiltration of atypical cells with high nuclear-cytoplasmic ratios, irregular nuclear contours, and occasional prominent nucleoli. Signet ring cells with eccentrically located nuclei and abundant foamy cytoplasm also were noted (Figure 2). The results of immunohistochemical staining for pancytokeratin were positive, consistent with poorly differentiated primary gastric carcinoma (Figure 3).

Gastric carcinoma of the umbilicus, or Sister Mary Joseph nodule, was diagnosed. In light of the biopsy findings, the patient underwent repeated computed tomography, which showed overall stable disease. However, the development of the new metastatic lesion on the umbilicus represented progression of the disease despite chemotherapy. The patient was to begin treatment with second-line agents, irinotecan hydrochloride and docetaxel.

Comment

Approximately 5% to 9% of patients with internal carcinomas develop cutaneous metastasis,¹ and approximately 10% of these cutaneous metastases are localized to the umbilicus.² The eponym *Sister Mary Joseph nodule* was named after the surgical assistant who first noted the connection between umbilical nodules and abdominal malignancy.² The nodules usually are firm, irregular, and small, and may be painful and exudative. They also may present with diffuse infiltration and erythema.³

The differential diagnosis of an umbilical mass includes other benign and malignant tumors.³ The umbilicus is the most common site of endometriosis, with an estimated prevalence of 10% in females of childbearing age. Foreign body granuloma, papilloma, fibroma, epithelial inclusion cysts,

From University of Southern California, Keck School of Medicine, Los Angeles.

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Correspondence: David H. Peng, MD, MPH, University of Southern California, Keck School of Medicine, 1200 N State St GH8440, Los Angeles, CA 90033 (dpeng@usc.edu).



Figure 1. A pinkish tan papillomatous plaque around the umbilicus with an indurated dermis and foci of necrosis.

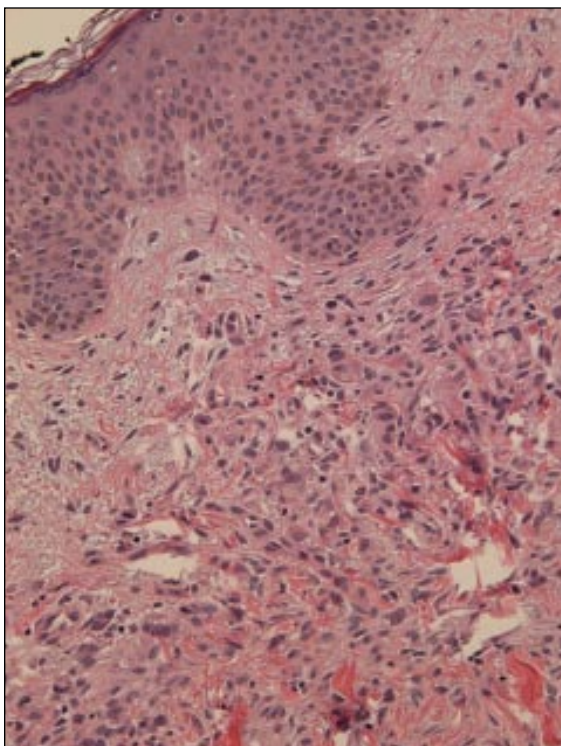


Figure 2. Atypical cells with large nuclei as well as signet ring cells with eccentrically located nuclei and abundant foamy cytoplasm were seen infiltrating the superficial dermis (H&E, original magnification $\times 20$).

epidermoid cysts, abscesses, seborrheic keratosis, myxoma, and hernia also often present as umbilical nodules. More rarely, primary umbilical carcinomas, such as melanoma; squamous and basal cell carcinoma; sarcoma; and adenocarcinoma also have been reported in this anatomic location.³ An alternative

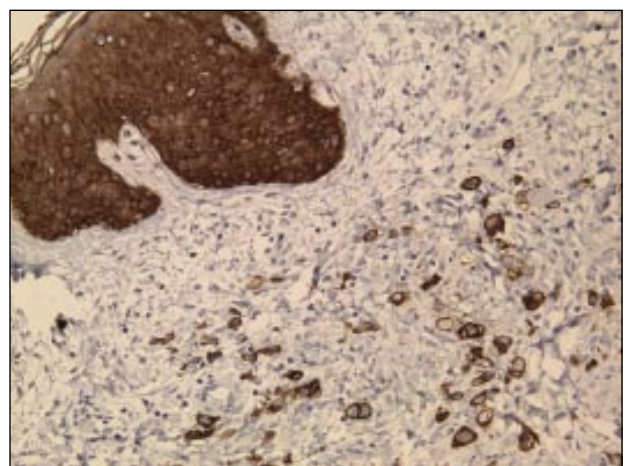


Figure 3. Immunohistochemical staining was positive for pancytokeratin (original magnification $\times 20$).

primary cancer should be excluded with a complete workup, as umbilical tumors are the first and only presenting sign of an occult malignancy in up to 30% of cases.² In other cases, their presence may indicate a recurrence in a patient with a medical history of an internal cancer. Once diagnosed, the presence of Sister Mary Joseph nodule represents advanced metastatic disease with an associated poor prognosis and an average survival time of 11 months.³

In adult males, the most frequent site of primary malignancy is the gastrointestinal tract (55%), most notably the stomach (30%).⁴ Gynecologic malignancies, especially ovarian and endometrial neoplasms, are the most common primary malignancies to metastasize to the umbilicus in women.

In 3% to 6% of cases, metastases originate from hematologic sources, the lungs, the cervix, and other sites. The primary neoplasm remains occult in 15% to 30% of cases.⁴ Metastatic processes to the umbilicus may follow several possible pathways, such as deep and superficial lymphatics, venous or arterial networks, embryologic remnants, contiguous extension, or iatrogenic presentation following laparoscopy.³

Gastric carcinomas, especially the signet ring cell subtype, occasionally are known to develop cutaneous lesions of varying clinical manifestations. Most present as firm nodules,⁵ but other cases resembling erysipelas,⁶ contact dermatitis,⁷ and epidermoid cyst⁸ have been reported.

Conclusion

Sister Mary Joseph nodule can represent an increased cancer burden in a patient with known internal malignancy. Although it is a rare clinical manifestation of metastatic cancer, it signifies a poor prognosis. Because the clinical presentation of these lesions is highly variable, histopathologic examination is highly encouraged to make an accurate diagnosis. Physicians must use a high degree of suspicion when evaluating an umbilical lesion in a patient with known internal malignancy.

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