

Sister Mary Joseph Nodule: A Case Report

Andrea Baratta, DO; Risa J. Gorin, DO; Richard Costa, DO

GOAL

To understand Sister Mary Joseph nodules (SMJNs) to better manage patients with these lesions

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Explain the mechanisms of umbilical metastasis.
2. Describe the clinical presentation of SMJN.
3. Discuss the histopathology of lesions in the umbilical region.

CME Test on page 479.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: November 2007.

This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education through the joint sponsorship of Albert Einstein College of Medicine and Quadrant HealthCom, Inc. Albert

Einstein College of Medicine is accredited by the ACCME to provide continuing medical education for physicians.

Albert Einstein College of Medicine designates this educational activity for a maximum of 1 AMA PRA Category 1 Credit™. Physicians should only claim credit commensurate with the extent of their participation in the activity.

This activity has been planned and produced in accordance with ACCME Essentials.

Drs. Baratta, Gorin, and Costa report no conflict of interest. The authors report no discussion of off-label use. Dr. Fisher reports no conflict of interest.

A metastatic malignancy of the umbilicus is commonly termed Sister Mary Joseph nodule (SMJN). It is a rare occurrence but may represent the first sign of a visceral malignancy and therefore should prompt a thorough search for the primary tumor. Typically, the most common origin of an umbilical metastasis is an adenocarcinoma from a gastrointestinal or gynecologic primary malignancy. The presence of SMJN carries a poor prognosis with

the average survival time at the appearance of an umbilical metastasis being 10 months. We report a case of a 66-year-old man who was referred for evaluation of an enlarging umbilical lesion. Histopathology revealed adenocarcinoma. After a full metastatic workup, the tumor of origin was identified as adenocarcinoma of the sigmoid colon. Benign tumors of the umbilicus are uncommon. This case report serves to emphasize the importance of obtaining a histologic diagnosis when any new lesion presents in the umbilical region.

Cutis. 2007;80:469-472.

Accepted for publication November 16, 2006.

Dr. Baratta was and Drs. Gorin and Costa are Clinical Assistant Professors, Family Practice Residency, University of Medicine & Dentistry of New Jersey, School of Osteopathic Medicine, Stratford. Reprints: Andrea Baratta, DO, St. John's Episcopal Hospital—South Shore, 327 Beach 19th St, 6th Fl, Far Rockaway, NY 11691-4423 (e-mail: abcalleo@hotmail.com).

Case Report

A 66-year-old white man presented for evaluation of an umbilical lesion of 3 weeks' duration. The lesion

**Figure not
available online**

Figure 1. Umbilical mass on patient's abdomen. Reprinted with permission from <http://www.surgical-tutor.org.uk>.¹ Photograph courtesy of Sampurna Tuladhar, B&B Hospital, Katmandu, Nepal.

was asymptomatic and nontender but was bleeding and oozing. The lesion, diagnosed by the patient's primary care physician as an infected pyogenic granuloma, was treated with oral antibiotics, without relief. His past medical history was unremarkable. He took no medications at that time and had an allergy to quinolones. His dermatologic history was positive for numerous actinic keratoses and a basal cell carcinoma on the forehead.

Physical examination upon presentation revealed a 3-cm fungating polypoid umbilical mass with copious purulent exudates similar to Figure 1.¹ The clinical differential diagnosis at that time included a pyogenic granuloma versus a Sister Mary Joseph nodule (SMJN). A shave biopsy was performed. Histopathology revealed multiple cystic and ductal spaces lined by atypical epithelial cells with numerous mitotic figures in the dermis (Figures 2 and 3). These changes favored a gastrointestinal primary lesion, supporting the clinical diagnosis of SMJN. The patient subsequently underwent a full metastatic workup. A computed tomographic scan of the abdomen and pelvis demonstrated no definite metastases to the lungs or liver but suggested a near-obstructing distal bowel primary tumor. A colonoscopy demonstrated a near-obstructing sigmoid colon carcinoma. The patient subsequently underwent an exploratory laparotomy, sigmoid resection, and primary colonic reanastomosis. Liver biopsies and

resection of the omentum and the periumbilical abdominal wall including the eroding metastasis were performed as well as multiple peritoneal biopsies. Pathologic evaluation confirmed the presence of extensive carcinomatosis. The primary tumor was a 5.0×3.0-cm moderately differentiated adenocarcinoma of the sigmoid colon with extension through the bowel wall. Biopsies of the liver, lymph nodes, periumbilical soft tissue, omentum, and peritoneum were all positive for metastatic carcinoma. Postoperatively, the patient was started on the FOLFOX regimen incorporating folinic acid, 5-fluorouracil, and oxaliplatin. Currently, the patient remains clinically stable and without evidence of progressive sigmoid colon carcinoma.

Comment

SMJN is the term applied to an uncommon metastatic malignancy of the umbilicus. This eponym, coined by Sir Hamilton Bailey in his book *Physical Signs in Clinical Surgery*,² honors Sister Mary Joseph Dempsey, a nun in the Franciscan order who was the first surgical assistant to Dr. William J. Mayo at St. Mary's Hospital in Rochester, Minnesota.^{3,4} Sister Mary Joseph made the astute observation that patients with gastrointestinal and gynecologic tumors may have involvement of the umbilicus; if there is involvement of the umbilicus, the prognosis is poor.⁵ Although Dr. Mayo gives credit to Sister Mary Joseph, contrary to what some believe, he is not responsible for the eponym. Dr. Mayo used the term *pants button umbilicus* in his article; Sir Bailey definitively coined the term SMJN. Some authors argue that the correct name is *Sister Joseph's nodule*; however, SMJN remains the more commonly used term.⁶

Frequency—Cutaneous metastasis, in and of itself, is a rare feature of visceral malignancy, occurring in about 1% to 2% of cases, with umbilical metastasis occurring even less frequently. There are approximately 265 cases of SMJN reported in the literature up to 1990, emphasizing its infrequency. The most common origin of an umbilical metastasis is an adenocarcinoma from a gastrointestinal or gynecologic primary malignancy. In a review of the world literature on metastatic umbilical tumors, Barrow⁷ found that the most common primary tumor identified was carcinoma of the stomach. The second most common was ovarian tumor,⁷ with colon, rectal, and pancreatic cancers following in descending order of frequency.⁸ In very rare cases, the primary tumor involved the breast, cervix, endometrium, small bowel, liver, gallbladder, lung, prostate, kidney, fallopian tube, appendix, and penis.⁹ In approximately 20% to 30% of cases, the primary tumor could not be identified.⁹

Umbilical Anatomy—A neoplasm may metastasize to the umbilicus by one of many mechanisms, most commonly direct extension from the anterior peritoneal surface from adjacent organs. Other mechanisms of metastasis include vascular spread via arterial and venous channels, lymphatic spread, and umbilical ligamentous spread.

The rich vascular supply to the anterior abdominal wall facilitates metastasis of a neoplasm. The main arteries supplying the umbilical region are the inferior epigastric, deep circumflex iliac, and superior epigastric arteries. Venous drainage of the umbilical region stems from a network of veins radiating from the umbilicus, including the axillary vein above and the femoral vein below the umbilicus. There also are small paraumbilical veins that connect with the portal system along the ligamentum teres hepatis, the superior portion of the falciform ligament.¹⁰

The superficial lymphatic drainage of the peri-umbilical region leads to axillary lymph nodes above and inguinal lymph nodes below the peri-umbilical region. The deep lymphatic drainage runs along the falciform ligament of the liver and then enters the anterior mediastinum by traversing the diaphragm. Inferiorly, the deep lymphatics communicate by coursing to the lymph nodes of the iliac arteries. Because the lymphatic channels of the periumbilical region communicate with the deep inguinal, axillary, deep femoral, and periaortic lymph nodes, once a neoplasm is established at the umbilicus, it can metastasize to any part of the body,⁸ supporting the notion that the presence of SMJN portends a poor prognosis. In the fetus, the umbilicus serves as the opening for the umbilical vessels. In the adult, there are several ligaments that contain remnants of obliterated fetal structures and connect to the umbilicus. For example, the median umbilical ligament, also known as the urachus, represents the obliterated umbilical arteries that formerly connected the umbilicus to the bladder. The falciform ligament in the adult contains the obliterated umbilical vein and runs along the hepatic surface. Therefore, a firm command of the anatomy and embryology of the periumbilical region is essential for an accurate clinical workup once the diagnosis of SMJN has been established.

Physical Examination—Clinically, SMJN usually presents as a firm indurated plaque or nodule that is 0.5 to 2 cm in size, though lesions up to 10 cm have been reported. The lesion is occasionally painful and may have a fibrous consistency with irregular edges that are attached to the anterior abdominal wall. It tends to have a vascular

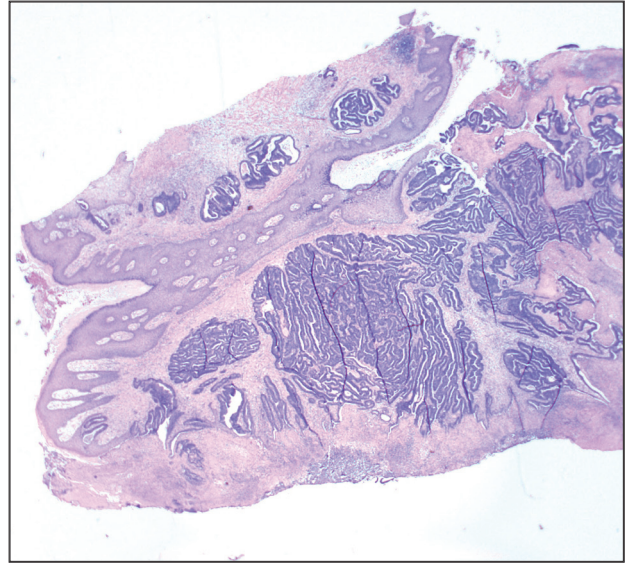


Figure 2. Polypoid pedunculated mass composed of multiple cystic and ductal spaces (H&E, original magnification $\times 10$).

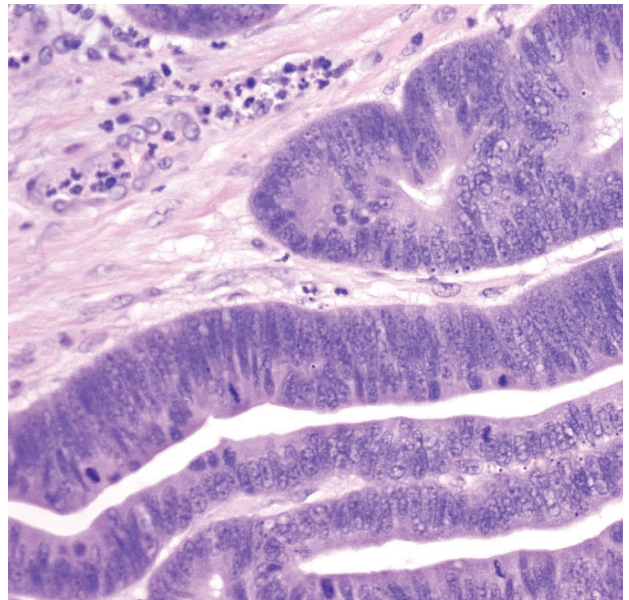


Figure 3. High-power image demonstrating multiple cystic and ductal spaces lined by atypical epithelial cells with numerous mitotic figures in the dermis (H&E, original magnification $\times 40$).

appearance and can be fissured, ulcerated, or necrotic. Purulent discharge may be present if it is secondarily infected. Typically, there are no gross features to distinguish between a primary and secondary umbilical tumor.

Differential Diagnosis—The differential diagnosis of SMJN includes a primary carcinoma of the umbilicus, a benign umbilical lesion, and a metastatic

lesion of the umbilicus. Primary umbilical carcinomas are rare and include malignant melanoma, basal cell carcinoma, and omphalomesenteric duct carcinoma. Endometriosis is the most common benign lesion of the umbilicus. Other benign umbilical lesions include papillomas, epidermal/inclusion cysts, seborrheic keratoses, dermal nevi, polyps, congenital malformations, foreign bodies, talc granulomas, angiomas, pyogenic and pilonidal granulomas, keloids, incarcerated hernias, angiokeratomas, and lymphangiomas.¹¹

Histopathology—In general, the histology of a cutaneous metastatic lesion is of finite value in determining the site of the primary malignancy. In contrast, when speaking of an umbilical metastasis, histologic sampling more often aids in determining the derivation.¹¹ The histopathology is dependent on the primary tumor and is most often adenocarcinoma, which was the case with our patient.

Prognosis—Although the presence of umbilical disease indicates metastasis, it does not suggest unresectability and, in fact, may be the first sign of a recurrence.¹¹ However, in general, SMJN portends a poor prognosis and is considered an ominous sign. The presence of SMJN should prompt a thorough metastatic workup and appropriate clinical staging. Survival time in untreated patients ranges from 2 to 11 months, with 10 months as the average survival time from the appearance of an umbilical metastasis.^{6,9} This rapid deterioration may be, in part, caused by the rich lymphatic network in the abdominal wall providing communication with the deep inguinal, axillary, deep femoral, and periaortic lymph nodes. Therefore, from the umbilicus, a tumor may spread to many parts of the body.

Conclusion

Cutaneous metastasis is a rare occurrence, making SMJN exceptionally infrequent. The finding of SMJN should prompt a thorough search for the primary malignancy. As demonstrated by our patient, SMJN may be the first sign of a visceral malignancy, making proper and timely diagnosis lifesaving.

REFERENCES

1. Parker S. Gastric carcinoma, Sister Mary Joseph nodule. Available at: <http://www.surgical-tutor.org.uk>. Accessed September 14, 2006.
2. Bailey H. *Demonstrations of Physical Signs in Clinical Surgery*. 11th ed. Baltimore, Md: Williams and Wilkins; 1949.
3. Gabriele R, Borghese M, Conte M, et al. Sister Mary Joseph's nodule as a first sign of cancer of the cecum: report of a case. *Dis Colon Rectum*. 2004;47:115-117.
4. Beynet D, Oro AE. Leukemia cutis presenting as a Sister Mary Joseph nodule. *Arch Dermatol*. 2004;140:1170-1171.
5. Schiffer JT, Park C, Jefferson BK. Cases from the Osler Medical Service at Johns Hopkins University. Sister Mary Joseph nodule. *Am J Med*. 2003;114:68-70.
6. Skellchock LE, Goltz RW. Umbilical nodule. metastatic adenocarcinoma (Sister Mary Joseph nodule). *Arch Dermatol*. 1992;128:548-549.
7. Barrow MV. Metastatic tumors of the umbilicus. *J Chron Dis*. 1966;19:1113-1117.
8. Pieslor PC, Hefter LG. Umbilical metastasis from prostatic carcinoma—Sister Joseph nodule. *Urology*. 1986;27:558-559.
9. Khan AJ, Cook B. Metastatic carcinoma of umbilicus: "Sister Mary Joseph's nodule." *Cutis*. 1997;60:297-298.
10. Coll DM, Meyer JM, Mader M, et al. Imaging appearances of Sister Mary Joseph nodule. *Br J Radiol*. 1999;72:1230-1233.
11. Chen P, Middlebrook MR, Golman SM, et al. Sister Mary Joseph nodule from metastatic renal cell carcinoma. *J Comput Assist Tomogr*. 1998;22:756-757.

DISCLAIMER

The opinions expressed herein are those of the authors and do not necessarily represent the views of the sponsor or its publisher. Please review complete prescribing information of specific drugs or combination of drugs, including indications, contraindications, warnings, and adverse effects before administering pharmacologic therapy to patients.

CONFLICT OF INTEREST STATEMENT

The Conflict of Interest Disclosure Policy of Albert Einstein College of Medicine requires that authors participating in any CME activity disclose to the audience any relationship(s) with a pharmaceutical or equipment company. Any author whose disclosed relationships prove to create a conflict of interest, with regard to their contribution to the activity, will not be permitted to present.

The Albert Einstein College of Medicine also requires that faculty participating in any CME activity disclose to the audience when discussing any unlabeled or investigational use of any commercial product, or device, not yet approved for use in the United States.