Sister Mary Joseph Nodule as Presenting Complaint in First Diagnosis of Intra-Abdominal Malignancy

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INTRODUCTION

The Emergency Department is often where the diagnosis of suspected malignancy is first made. As clinicians, we must have a high index of suspicion when patients present with complaints or physical exam findings that may be associated with cancer. Patients with abnormal lab results or imaging that is concerning for malignancy should be admitted or referred for close follow-up for further workup and definitive diagnosis.

CASE REPORT

An 81-year-old woman with a history of COPD on three liters of home oxygen, hypertension, hyperlipidemia, pulmonary fibrosis and stroke presented to the emergency department with a chief complaint of bleeding from her umbilicus. Earlier in the evening, she noticed a small trickle of blood from her umbilical region. She had never had bleeding from this location. By the time she arrived, the bleeding had stopped. She denied any associated symptoms of nausea, vomiting, diarrhea, melena, fatigue, weight loss, abdominal distension or pain, headaches, chest pain or loss of appetite.

The patient had a 100-year pack smoking history; she quit in 2009. Her mother died of breast cancer at age 60. She lives with her two daughters and requires assistance with bathing secondary to debilitating arthritis.

When she presented to the emergency department, she was in no acute distress, and her vital signs were blood pressure of 124/80, pulse of 78 beats per minute, temperature of 98.4 degrees Fahrenheit, respiratory rate of 18, and pulse oximetry of 96%. She had a normal exam except for a 2x2 cm erythematous nodule protruding from the umbilicus, which was non-tender and not bleeding (**Image 1**). Her lab work did not show any significant abnormalities, except for slight elevation of her alkaline phosphatase at 120. The urine culture was positive for E. coli. Due to the concerns raised by the umbilical nodule, a CT scan of the abdomen and pelvis was ordered. The CT scan showed an umbilicus soft tissue nodule, extensive peritoneal carcinomatosis, extensive hepatic and intraperitoneal metastatic disease, lung nodules, and bilateral adnexal masses, most consistent with primary ovarian versus sigmoid malignancy (**Images 2 and 3**). Her umbilical nodule was diagnosed as a Sister Mary Joseph nodule in the context of extensive intra-abdominal carcinomatosis. She was admitted to the hospitalist service for oncology consult, biopsy of the nodule and initiation of cancer treatment.

Image 1. Umbilical nodule

A liver lesion biopsy revealed a metastatic high-grade serous carcinoma, consistent with spread from the female genital tract/Mullerian primary. A CT scan of the chest showed a right upper lobe mass and three left basilar pulmonary nodules, concerning for metastatic disease. A non-contrast CT scan of the brain did not show any lesions. The patient was started on antibiotics for her urinary tract infection.

She was started on carboplatin/paclitaxel for stage IV cancer and offered palliative chemotherapy and debulking pelvic surgery. The patient did not tolerate chemotherapy and consequently died from complications of her extensive malignancy.

Image 2. Axial CT Abdomen/Pelvis image demonstrating SMJ nodule and metastatic disease

Image 3. Sagittal CT Abdomen/Pelvis image of SMJ nodule and metastatic disease

DISCUSSION

An umbilical nodule may represent a benign umbilical hernia, or it may be the first visible manifestation of intra-abdominal malignancy. Presentation to the Emergency Department for a new umbilical mass is rare, but differentiating a benign from malignant nodule is critical. Umbilical metastases were reported as early as 1854 by Baluff and in 1860 by Nelaton; they were referred to as "trouser button navel".[1] However, the English surgeon, Hamilton Bailey, was the first to coin the popular eponym "Sister Mary Joseph's nodule" in 1949 in his textbook "Demonstration of Physical Signs in Clinical Surgery".[1,2] Sister Mary Joseph was a superintendent nurse at St. Mary's Hospital in Rochester, Minnesota (now Mayo Clinic) and reportedly pointed out to Dr. Bailey her finding that patients with intra-abdominal malignancy often had an umbilical nodule.[1,2]

The Sister Mary Joseph nodule is an umbilical mass that may be painful, firm, or ulcerated and may have serous, sanguinous, or purulent drainage.[2] These nodules tend to be small, 0.5 to 2 cm, but can get as large as 10 cm before presenting for evaluation. Patients who present with this nodule may have other symptoms of malignancy, including weight loss, abdominal pain, nausea, ascites, and bleeding per rectum.[3] CT scan is often the first step to characterize the extent of the umbilical mass and to look for other lesions and a primary malignancy. In 14%–33% of SMJ nodules, a new diagnosis of occult malignancy is made.[4] For patients with a known malignancy who present with this finding, 40% represent a recurrence of prior cancer.[4] In men, the source is often gastrointestinal (gastric, colonic, pancreatic), while in women, it is usually gynecologic (ovarian, uterine). The common primary malignancies are gastrointestinal (52%), gynecologic (28%), stomach (23%) and ovarian (16%). A high percentage of the cases, 15-29% are from an unknown primary.[3-5]

There are hypotheses about the spread of malignancy to the umbilicus from direct spread from peritoneal masses, hematogenous spread, and lymphatic spread via embryonic remnants (round ligament of the liver, urachus or obliterated vitelline artery).[6,7] Presence of an SMJ nodule is associated with advanced disease and a poor prognosis. Average survival is around 11 months with <15% survival reported at 2 years.[2] Therefore, it is critical to have a high index of suspicion when a patient presents with a new umbilical nodule, especially when there is a personal or strong family history of malignancy.

References

1. Abu-Hilal, Mohannad, James S. Newman. "Sister Mary Joseph and Her Nodule: Historical and Clinical Perspective." *The American Journal of Medical Sciences,* vol. 337, no. 4, 2009, pp.271-273.
2. Dar IH, Kamili MA, Dar SH, Kuchaai FA. Sister Mary Joseph nodule-A case report with review of literature. *Journal of Research in Medical Sciences : The Official Journal of Isfahan University of Medical Sciences*. 2009;14(6):385-387.
3. Urbano FL. Sister Joseph's nodule. (44). *Hospital Physician*. 2001;37(5):33–5.
4. Majumdar B, Wiskind AK, Croft BN, Dudley AG. The Sister (Mary) Joseph nodule: its significance in gynecology. *Gynecol Oncol*. 1991;40(2):152–9.
5. Larentzakis A, Theodorou D, Fili K, Manataki A, Bizimi V, Tibishrani M, et al. Sister Mary Joseph's nodule: three case reports. *Cases Journal*. 2008;1:182.
6. Srinivasan R, Ray R, Nijhawan R. Metastatic cutaneous and subcutaneous deposits from internal carcinoma. An analysis of cases diagnosed by fine needle aspiration. *Acta Cytol*. 1993;37(6):894–8.
7. Powell FC, Cooper AJ, Massa MC, Goellner JR, Su WP. Sister Mary Joseph's nodule: a clinical and histologic study. *J Am Acad Dermatol*. 1984;10(4):610–5.

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