

Crusted Plaque in the Umbilicus

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A 74-year-old man presented to our outpatient dermatology clinic with an asymptomatic umbilical lesion of unknown duration. The patient believed the lesion was a scar resulting from a prior laparoscopic repair of an umbilical hernia. However, the patient reported epigastric abdominal pain and diarrhea of 1 month's duration that he believed was due to the stomach flu. The patient denied fever, chills, loss of appetite, or weight loss. History was remarkable for hypertension, hyperlipidemia, coronary artery disease, chronic kidney disease, and emphysema. The patient had a surgical history of percutaneous transluminal coronary angioplasty in addition to the laparoscopic umbilical hernia repair. The patient's medications included pantoprazole, ondansetron, diphenoxylate-atropine as needed, amlodipine, lisinopril-hydrochlorothiazide, simvastatin, and aspirin. Physical examination revealed a 1x2-cm pink, nodular, firm plaque with crust at the umbilicus that was tender on palpation. A shave biopsy of the umbilicus was performed and sent for both pathological and immunohistochemical analysis.

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What's the diagnosis?

- nonmelanoma skin cancer
- psoriasis
- pyogenic granuloma
- scar
- Sister Mary Joseph nodule

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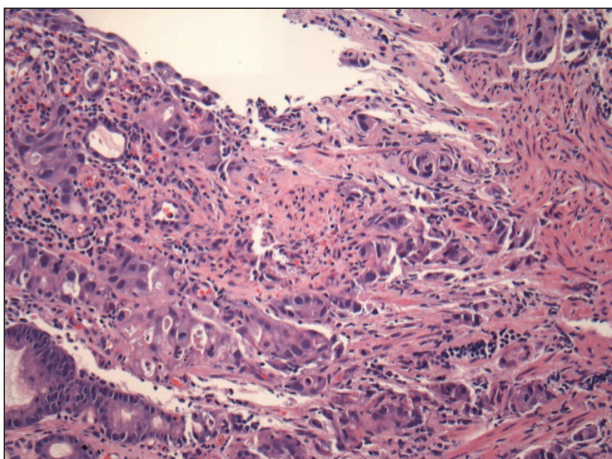
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The Diagnosis: Sister Mary Joseph Nodule

The umbilical skin biopsy revealed a moderately differentiated adenocarcinoma (Figure) that was positive for cytokeratin 20 and CDX2 and negative for cytokeratin 7 and transcription termination factor 1. The patient subsequently underwent computed tomography of the abdomen and pelvis, which showed multiple soft-tissue nodules on the greater omentum, a soft-tissue density at the umbilicus, and thickening of the gastric mucosa. An upper endoscopy was then performed, which revealed a large fungating ulcerated mass in the stomach. Biopsy of this mass showed an invasive moderately differentiated adenocarcinoma, which was *ERBB2* (formerly *HER2*) negative. Histopathologically, these pleomorphic glands looked similar to the glands seen in the original skin biopsy. With this diagnosis of metastatic gastric adenocarcinoma, our patient chose palliative chemotherapy but declined precipitously and died 2 months after the initial skin biopsy of the umbilical lesion.

When encountering a patient with an umbilical lesion, it is important to consider benign and malignant lesions in the differential diagnosis. A benign lesion may include scar, cyst, pyogenic granuloma, hemangioma, umbilical hernia, endometriosis, polyp, abscess, or the presence of an omphalith.¹ Inflammatory dermatoses such as psoriasis or eczema also should be considered. Malignant lesions could be either primary or secondary, with metastatic disease being the most common.²



Skin biopsy from the umbilical lesion (H&E, original magnification $\times 40$).

Sister Mary Joseph nodule (SMJN) is the eponym given to an umbilical lesion representing metastatic disease. Sister Mary Joseph was a nurse and surgical assistant to Dr. William Mayo in Rochester, Minnesota, in what is now known as the Mayo Clinic. She is credited to be the first to observe and note the association between an umbilical nodule and intra-abdominal malignancy. Metastasis to the umbilicus is thought to occur by way of contiguous, hematogenous, lymphatic, or direct spread through embryologic remnants from primary cancers of nearby gastrointestinal or pelvic viscera. It is a rare cutaneous sign of internal malignancy, with an estimated prevalence of 1% to 3%.³ The most common primary cancer is gastric adenocarcinoma, though cases of metastasis from pancreatic, endometrial, and less commonly hematopoietic or supradiaphragmatic cancers have been reported.⁴ It is more common in women, likely due to the addition of gynecologic malignancies.¹

The use of dermoscopy has been advocated as an adjuvant tool in delineating benign and malignant umbilical lesions when an atypical polymorphous vascular pattern indicating neovascularization has been observed with neoplastic growth.⁵ Once a suspicious umbilical lesion is identified, the first step should be to obtain a skin biopsy or to use fine needle aspiration for cytology.⁶ Biopsy is especially relevant in the background of cancer history because SMJN may present with cancer recurrence.³ Once one of these is obtained, histological and immunohistochemical analysis will guide further workup and diagnosis of the umbilical lesion.

The importance of reviewing such cases lies in the variable presentation of cutaneous metastases such as SMJN and the grim prognosis that accompanies this finding. It presents as a firm indurated plaque or nodule that may present with systemic symptoms suggestive of malignancy, though in 30% of cases it is the sole initial sign.⁷ The nodule may be painful if ulcerated or fissured. Bloody, serous, or purulent discharge may be present. After diagnosis of an SMJN, most patients succumb to the disease within 12 months. Thus, it is vital for dermatologists to investigate umbilical lesions with great caution and a high index of suspicion.

REFERENCES

1. Chalya PL, Mabula JB, Rambau PF, et al. Sister Mary Joseph's nodule at a University teaching

- hospital in northwestern Tanzania: a retrospective review of 34 cases. *World J Surg Oncol*. 2013;11:151.
2. Papalas JA, Selim MA. Metastatic vs primary malignant neoplasms affecting the umbilicus: clinico-pathologic features of 77 tumors. *Ann Diagn Pathol*. 2011;15:237-242.
 3. Palaniappan M, Jose WM, Mehta A, et al. Umbilical metastasis: a case series of four Sister Joseph nodules from four different visceral malignancies. *Curr Oncol*. 2010;17:78-81.
 4. Zhang YL, Selvaggi SM. Metastatic islet cell carcinoma to the umbilicus: diagnosis by fine-needle aspiration. *Diagn Cytopathol*. 2003;29:91-94.
 5. Mun JH, Kim JM, Ko HC, et al. Dermoscopy of a Sister Mary Joseph nodule. *J Am Acad Dermatol*. 2013;68:e190-e192.
 6. Handa U, Garg S, Mohan H. Fine-needle aspiration of Sister Mary Joseph's (paraumbilical) nodules. *Diagn Cytopathol*. 2008;36:348-350.
 7. Abu-Hilal M, Newman JS. Sister Mary Joseph and her nodule: historical and clinical perspective. *Am J Med Sci*. 2009;337:271-273.