

## Sister Joseph's Nodule

Frank L. Urbano, MD

**U**mbilical tumors are relatively rare and can be benign or malignant.<sup>1</sup> Malignant tumors can be classified as primary tumors or metastatic tumors. Metastatic tumors can arise from a variety of primary malignant tumors. The term *Sister Joseph's nodule* is used to describe a malignant umbilical tumor. The tumor is usually associated with advanced, metastasizing intra-abdominal cancer. It generally indicates a poor prognosis.<sup>2</sup> This article describes Sister Joseph's nodule, gives a brief overview of umbilical tumors, and discusses the significance of Sister Joseph's nodule as a clinical sign.

### HISTORICAL PERSPECTIVE

Sister Mary Joseph, born Julia Dempsey in 1856, entered the Third Order Regular of Saint Francis of the Congregation of Lady of Lourdes in Rochester, MN, in 1878.<sup>3</sup> This order was significantly involved in the operation of St. Mary's Hospital, which was built in 1889 and which was the earliest incarnation of the now famous Mayo Clinic. Sister Mary Joseph was trained in nursing at St. Mary's Hospital and soon after became head nurse. In 1892, she became superintendent and served in that capacity until her death in 1939. However, prior to becoming superintendent, Sister Mary Joseph became a surgical assistant under the guidance of Dr. William Mayo, serving in this capacity from 1890 to 1915. Dr. Mayo relied a great deal on her assistance and wisdom. Once, when asked a technical question regarding what he would do in a certain clinical circumstance, he replied, "I would consult Sister Joseph and follow her advice."<sup>4</sup>

As a surgical assistant, Sister Mary Joseph's responsibilities included preparing the abdomens of patients scheduled to undergo surgery. It was while performing this act that she first identified the clinical sign that was to bear her name: she noted that the presence of an umbilical nodule usually indicated advanced intra-abdominal cancer. The term *Sister Mary Joseph's nodule* did not exist, however, until 1949 (10 years after her death), when Hamilton Bailey coined the term in the 11th edition of his textbook, *Physical Signs in Clinical*

### SISTER JOSEPH'S NODULE

A firm, indurated umbilical nodule that is usually associated with advanced, metastasizing intra-abdominal cancer. It generally indicates a poor prognosis.

*Surgery.* Because Sister Mary Joseph had removed *Mary* from her name, the sign has also been referred to as *Sister Joseph's nodule*. The correct name of the sign has been debated for years, with many authors believing that the more common designation, *Sister Mary Joseph's nodule*, is incorrect.<sup>5</sup>

### DESCRIPTION OF SISTER JOSEPH'S NODULE

Sister Joseph's nodule (**Figures 1 and 2**) has been described extensively in the literature. The nodule usually presents in a firm and indurated state, often with a vascular appearance. It may be fissured and ulcerated and may have a bloody, mucinous, serous, or purulent discharge.<sup>6,7</sup> The nodule has been described as white, bluish-violet, or brownish-red, and it is occasionally pruritic.<sup>8</sup> It is usually irregular in shape and generally painless when palpated, except when the overlying skin has ulcerated. It is typically less than 5 cm in diameter but occasionally enlarges enough to form a protruding tumor.<sup>9</sup> In general, however, the gross appearance of an umbilical nodule is largely unreliable for diagnostic purposes, because umbilical nodules with different causes can have a similar appearance.

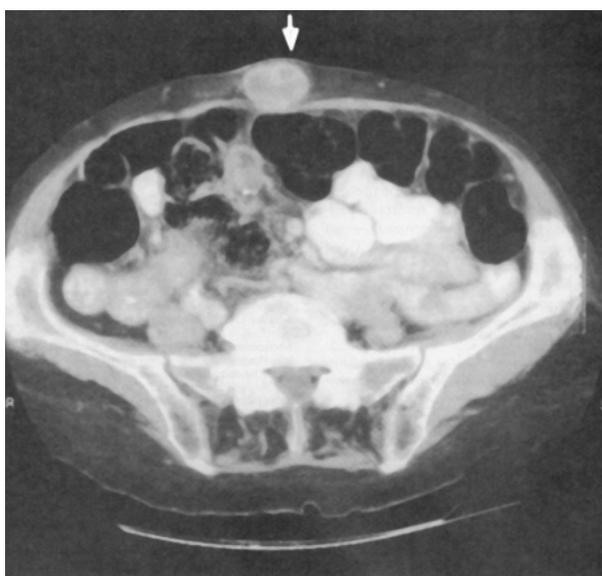
Patients with Sister Joseph's nodule may present with a number of other clinical symptoms consistent with intra-abdominal cancer, including epigastric pain, abdominal distension, weight loss, nausea, and ascites.<sup>10</sup> Such symptoms can assist the examiner in determining the clinical significance of a nodule that upon gross

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*Dr. Urbano is in general internal medicine, Mount Laurel Primary Care Physicians, Mount Laurel, NJ.*



**Figure 1.** Sister Joseph's nodule. Reprinted with permission from CJS 1997;40:167. Copyright 1997 Canadian Medical Association, www.cma.ca.



**Figure 2.** Computed tomography scan showing an umbilical subcutaneous nodule (arrow), Sister Joseph's nodule. Reprinted with permission from Moll S. Sister Joseph's node in carcinoma of the cecum. N Engl J Med 1996;335:1568. Copyright 1996 Massachusetts Medical Society. All rights reserved.

examination seems to be Sister Joseph's nodule. That is, when a patient has an umbilical nodule and these symptoms, there is a high probability that the nodule is caused by a metastasizing cancer of the intra-abdominal area. Some patients, however, may present with an umbilical nodule, which could be Sister Joseph's nodule, but they may have no other signs or symptoms of an internal malignancy. Thus, a histologic examination of a

biopsy specimen from the nodule becomes important in determining a diagnosis.

## OVERVIEW OF UMBILICAL TUMORS

### Noncancerous Tumors

Although an umbilical nodule is often associated with a malignant process, it may also be associated with nonmalignant conditions that produce benign tumors. Benign tumors of the umbilical region, which make up about 57% of all umbilical tumors, include nonmalignant melanocytic tumors (dermal nevi), the most common type, and many other less prevalent nonmalignant lesions, including fibroepithelial papillomas, epithelial inclusion cysts, seborrheic keratoses, dermatofibromas, and polyps.<sup>1,6,9</sup>

Congenital malformation of the omphalomesenteric duct or urachus can also cause an umbilical nodule, as can foreign bodies and talc granulomas—all of which may be seen in newborns or young children. Hypertrophic umbilical scars with keloid formation can likewise cause an umbilical nodule as can umbilical hernias.

Another nonmalignant lesion that may cause an umbilical nodule is umbilical endometriosis.<sup>1,9</sup> This is quite rare and produces a solitary nodule that can be dusky red, blue, purplish, or brown. It may change in size during the menstrual period and may also exhibit tenderness and bleeding, a fact that can help in its identification.

### Cancerous Tumors

Malignant tumors of the umbilical region make up about 43% of all umbilical tumors. Primary malignant umbilical tumors can be melanomas, basal cell carcinomas, squamous cell carcinomas, myosarcomas, and adenocarcinomas; primary tumors of the umbilical region are exceedingly rare, constituting only 17% of malignant umbilical tumors.<sup>1</sup> Adenocarcinomas and myosarcomas are assumed to arise in ectopic tissue, including remnants of the omphalomesenteric duct or urachus.

Metastatic tumors of the umbilical region, constituting 83% of all malignant umbilical tumors, are much more common than primary malignant tumors of this area.<sup>1,6,11</sup> The histologic nature of metastatic umbilical tumors usually suggests adenocarcinoma; however, there have also been reports of umbilical metastases from sarcoma, mesothelioma,<sup>12</sup> and melanoma.<sup>13</sup> In approximately 29% of the cases of metastatic umbilical tumors, the specific site of the primary tumor is unknown.<sup>14</sup> Known common sites of origin for Sister Joseph's nodule include the stomach (25%), ovary (12%), colorectal

region (10%), and pancreas (7%). Primary tumors in many other sites have also been reported to lead to Sister Joseph's nodule, including those in the gallbladder, uterus, liver, endometrium, small intestine, fallopian tube, appendix, cervix, penis, prostate, breast, kidney, and lung.

The spread of metastatic cancer to the umbilical region has been hypothesized to occur in several ways. Contiguous spread of peritoneal cancer is thought to be the most simplistic and important method.<sup>9,10</sup> One report described an ovarian carcinoma that presented with a Sister Joseph's nodule; by histologic sectioning, the authors were able to prove that the spread of carcinoma occurred by direct extension,<sup>15</sup> a finding that had not been shown before in the literature. Hematogenous spread through the arterial and venous systems is another postulated method by which metastasis occurs, with the persistence of the paraumbilical veins being a possible conduit for metastatic spread. Lymphatic spread to the umbilical region is also a probable method of metastasis, considering that 4 sets of lymphatics pass from the umbilical region. In particular, metastases from pancreatic cancer are believed to cause Sister Joseph's nodule in this manner, because pancreatic cancer rapidly invades the lymphatic system.<sup>16</sup> Lastly, direct extension along the ligaments of embryonic origin is a presumed mode of metastatic spread, including the round ligament of the liver, the urachus, the vitellointestinal duct remnant, and the obliterated vitelline artery.<sup>15</sup> This method may be important in metastatic tumors of the small intestine.<sup>9</sup>

#### CLINICAL SIGNIFICANCE OF SISTER JOSEPH'S NODULE

The presence of Sister Joseph's nodule (verified histologically) usually signifies advanced, metastasizing cancer and therefore a poor prognosis. In his classic article on metastatic carcinoma of the umbilical region, Clements wrote "the finding of a metastatic nodule at the umbilical site almost certainly establishes the inoperability of the patient."<sup>17</sup> However, other authors have disputed this conclusion, noting that the presence of a malignant umbilical tumor is insufficient proof of widespread metastatic disease, which would prevent an operation. Specifically, some contend that a Sister Joseph's nodule may sometimes represent a solitary metastasis or possibly a primary tumor that has not yet metastasized.<sup>8,18</sup> Both of these instances are generally believed to be uncommon.

Sister Joseph's nodule usually represents widespread metastasis. Consequently, the treatment of patients with the nodule commonly involves palliation. Several authors have advocated other possible treatments,

including wide excision with extensive search for the primary lesion,<sup>18,16</sup> radiotherapy,<sup>14</sup> and surgery combined with adjunctive therapy.<sup>10</sup> In the latter case, the authors showed that patients with Sister Joseph's nodule who were treated aggressively with both surgery and adjunctive therapy lived an average of 17.6 months, which was more than with surgery alone (7.4 months), adjunctive therapy alone (10.3 months), or no treatment (2.3 months). In any event, the overall prognosis for patients presenting with Sister Joseph's nodule is generally poor, and the treatment plan for each patient must be individualized.

#### SUMMARY

Sister Joseph's nodule is a time-honored clinical sign, with a rich history, that often indicates advanced, metastatic intra-abdominal cancer. When found on physical examination, this nodule may provide physicians with both a site on which a biopsy can be easily performed and a crude indicator of the patient's prognosis. The ease of identifying the nodule speaks to the importance of a careful physical examination of the abdomen and a knowledge of the normal appearance of the umbilical region. Although advanced technology now exists to more easily diagnose malignancies, Sister Joseph's nodule nevertheless remains an interesting and useful diagnostic tool in modern medicine. **HP**

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