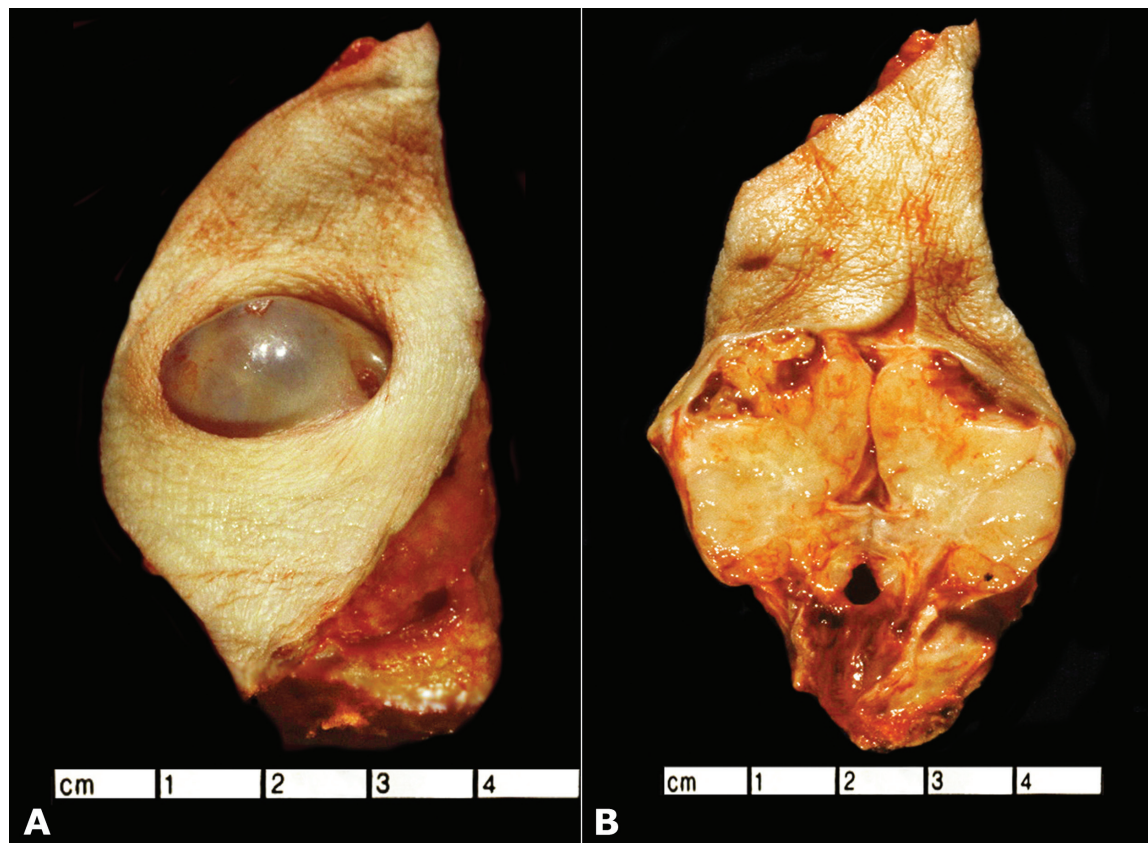


Sister Mary Joseph nodule: it does not bode well

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Sister Mary Joseph, the superintendent nurse and surgical assistant of Dr. William Mayo at St Mary's Hospital (presently Mayo Clinic), was the first to note the association of umbilical nodules with intra-abdominal malignancy.^{1,2} Dr. Hamilton Bailey, a British surgeon, in his classic book *Physical Signs in Clinical Surgery*³ named the lesion for Sister Mary Joseph;

the palpable bulging usually firm nodule distorting and invading into the umbilicus as a manifestation of metastatic malignancy from the abdomen or the pelvis.

Sister Mary Joseph's nodule (SMJN), was at the time of its description, recognized as metastatic disease.³

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Histologically, SMJNs are predominantly adenocarcinoma (75% of the cases), mostly metastatic from primary gastrointestinal (GI) tract or gynecological sites (12-15%) and, as in the case shown here, mucus-secreting.³

In the GI tract the stomach is the leading malignant primary site, followed by colorectal and pancreas (more often the body and tail, rather than the head), whereas ovarian serous papillary cyst adenocarcinoma is the most common site in females.⁴⁻⁶ Occurring rarely, SMJNs may be metastatic from a wide variety of other primary sites: gallbladder, liver, breast, lung, prostate, penis, peritoneum, lymphoma, bladder, kidney, endometrium, cervix, vagina, vulva, and fallopian tube.^{6,7}

SMJNs may be diagnosed on routine physical examination, during surgical workup or may represent the patient's presenting complaint. In some series⁸ this umbilical lesion was the initial manifestation of the internal malignancy. The umbilical metastasis may be direct extension from a contiguous tumor, by hematogenous or lymphatic spread, and uncommonly, as extension along the remnants of embryonic ligaments. Direct implantation after laparoscopy may also spread tumors to the umbilicus.⁹⁻¹¹ Clinically; the lesion is often a painful, hard lump with irregular margins. It is sometimes purplish, eventually ulcerates and varies in size from 0.5 to 2 cm. Tumors as large as 10 cm have been reported.^{4,11,12}

Differential diagnosis should include benign causes such as endometriosis, melanocytic nevi, dermatofibroma, fibroma, urachal duct cyst, pilonidal sinus, keloid, foreign body, granuloma, myxoma, omphalitis and umbilical hernia,^{5,13,14} as well as primary malignant umbilical skin tumors.^{5,13} Benign umbilical nodules (endometriosis, fibroma, foreign body granuloma, keloid, myxoma) are sometimes called "pseudo SMJN"¹⁵ although using the term in this manner may be unnecessarily confusing and should be avoided. Each of the benign lesions has distinctive gross and microscopic features.

SMJN usually represents widespread advanced cancer and therefore is associated with poor prognosis, almost always indicating inoperability although, particularly with direct extension from a primary site, this may not always obtain. Depending on the patient's general conditions as well as the characteristics of

the primary neoplasm, the mean survival time is approximately 11 months. Fewer than 15% of patients survive more than 2 years after diagnosis.⁴

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