

# Umbilical metastasis: a case series of four Sister Joseph nodules from four different visceral malignancies

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# ABSTRACT

Sister Joseph nodule is a metastatic umbilical lesion secondary to a primary malignancy of any viscera. It can be a presenting symptom (a sign of undiagnosed malignancy) or a symptom or sign of progression or recurrence in a known case. Its incidence is 1%–3% of all intra-abdominal or pelvic malignancies. Here, we present 4 such cases, with Sister Joseph nodule as a finding of

- presentation in a case of gallbladder carcinoma,
- progression in a case of malignant gastrointestinal stromal tumour,
- recurrence in a case of ovarian carcinoma, and
- presentation in a case of rectal carcinoma.

The clinicopathologic features of all 4 patients are discussed, and the related literature is briefly reviewed.

## **KEY WORDS**

Sister Mary Joseph, umbilical nodule, gallbladder carcinoma, malignant GIST, ovarian carcinoma, rectal carcinoma

## 1. INTRODUCTION

The first and best-known description of umbilical metastasis was published by William James Mayo in 1928. In 1949, the condition was named Sister Mary Joseph nodule (SMJN) by the British surgeon Hamilton Bailey<sup>1</sup>. This metastatic lesion of the umbilicus spreads from a primary malignancy in any viscera—stomach and colon being most common in men, and ovary, in women. Cases of SMJN metastasized from organs such as the cecum, endometrium, pancreas, gallbladder, prostate, and lung, and from unknown primaries have also been reported <sup>2</sup>. The lesion is seen more often in women<sup>3</sup>.

Typically, SMJN will be a firm irregular nodule, averaging in size from 1 cm to 1.5 cm, and occasionally reaching a maximum of 10 cm in diameter<sup>4</sup>. Sister Mary Joseph nodule can be a presenting symptom or sign of undiagnosed underlying malignancy, or an alarming symptom or sign of disease progression or recurrence in a known patient <sup>5</sup>. Here, we report 4 cases of SMJN encountered at our institute.

## 2. CASE DESCRIPTIONS

## 2.1 Case 1

A 65-year-old man presented with a progressively enlarging nodular growth in the umbilicus and increasing abdominal distension for 2 months. Clinical examination revealed a firm-to-hard nodular growth in the umbilicus (Figure 1) and minimal ascites. Fine-needle aspiration cytology of the umbilical nodule was suggestive of metastatic adenocarcinoma (Sister Joseph nodule). Computed tomography (CT) imaging of the abdomen showed a thickened, distended gallbladder with luminal projections and adjacent liver parenchymal infiltrations. It also revealed minimal intrahepatic biliary radical dilatation, mild-to-moderate ascites, and a soft-tissue density infiltrating the muscle and fat planes of the anterior abdominal wall at the umbilicus. The patient was subsequently diagnosed with metastatic gallbladder carcinoma.



FIGURE 1 Sister Joseph nodule in gallbladder carcinoma.

## 2.2 Case 2

A 54-year-old man was evaluated for complaints of anorexia, nausea, and regurgitation for 2 months, and loose stools and weight loss for 1 month. On evaluation, CT imaging of the abdomen showed features suggestive of small-bowel gastrointestinal stromal tumour (GIST), with a solitary liver metastasis. Primary tumour resection with end-to-end anastomosis and metastasectomy of the solitary liver lesion was done. Histopathologic examination and immunohistochemistry of the surgical specimens were reported as malignant GIST (epithelioid variant) with focal neural differentiation.

The patient was started on imatinib mesylate. After 2 years and 3 months of regular follow-up, he developed disease metastatic to liver and peritoneum. He was then prescribed sunitinib, which had to be stopped after 4 months because of hand-foot syndrome. The patient then took *siddha* treatment for 3 months, after which he returned to our institute with a smooth umbilical swelling (Figure 2). Fineneedle aspiration cytology of that lesion was reported as metastatic malignancy (Sister Joseph nodule).

## 2.3 Case 3

A 47-year-old woman self-referred to our cancer institute for management of first recurrence of her ovarian carcinoma. Her earlier treatment (surgery and adjuvant chemotherapy) had been delivered elsewhere. We surgically removed her recurrent stage IIIC mass after 4 cycles of neoadjuvant liposomal doxorubicin. She then received 6 cycles of adjuvant gemcitabine and carboplatin. After about 5 months of follow-up, she presented with an ulcerated serosanguinous secretory umbilical nodule (Figure 3). Fine-needle aspiration cytology of the lesion showed metastatic carcinoma (Sister Joseph nodule).



FIGURE 2 Sister Joseph nodule in malignant gastrointestinal stromal tumour.

#### 2.4 Case 4

A 79-year-old man was brought to the institute with an umbilical mass and abdominal distension of 2 months' duration. The patient was sick, with a performance status of 4 at presentation. Clinically, he was pale and had marked ascites and a flat, dark, non-ulcerated serous-secreting umbilical growth (Figure 4). Imaging by CT showed a rectal wall thickening, with peritoneal and omental infiltration and infiltration of the anterior abdominal wall at the umbilicus. The rectal and umbilical lesions were biopsied, and the histopathology report indicated rectal adenocarcinoma with umbilical metastasis (Sister Joseph nodule).

#### 3. DISCUSSION AND CONCLUSIONS

Table I sets out the important clinical characteristics of our 4 patients. The incidence of SMJN is very low, being diagnosed in only 1%–3% of all intra-abdominal or pelvic malignancies <sup>6</sup>. Neoplastic processes such Paget disease and angioma, and non-neoplastic diseases such as umbilical hernia, endometriosis, hypertrophic scar, granuloma, pilonidal sinus, mycosis,



FIGURE 3 Sister Joseph nodule in ovarian carcinoma.



FIGURE 4 Sister Joseph nodule in rectal carcinoma.

Characteristic	Case			
	1	2	3	4
Age (years) at primary diagnosis	65	54	45	79
Sex	Male	Male	Female	Male
Time of nodule appearance	Presentation	Progression	Recurrence	Presentation
Nodule size (cm)	2×1.5	2.5×3	3×3	2.5×2.5
Nature of nodule	Nodular, non-ulcerated, non-secreting	Bulging, non-ulcerated, non-secreting	Nodular, ulcerated, serosanguinous secreting	Flat, non-ulcerated, serous secreting
Primary diagnosis	Gallbladder carcinoma	Malignant GIST	Ovarian carcinoma	Rectal carcinoma
Time to nodule appearance after primary diagnosis	NA	2 Years, 10 months	3 Years, 5 months	NA
Treatments	Gemcitabine, capecitabine	Imatinib, sunitinib, <i>siddha</i> medicines	Surgery, adjuvant chemotherapy, doxorubicin, surgery, gemcitabine+carboplatin, capecitabine	Advised supportive care only
Outcome (from date of nodule diagnosis)	Lost to follow-up at 4 months	Lost to follow-up at 1 week	On follow-up for 4 months	Died at 2 weeks

TABLE I Clinical characteristics of 4 patients with Sister Mary Joseph nodule.

GIST = gastrointestinal stromal tumour; NA = not applicable.

psoriasis, and eczema are other differential diagnoses of an umbilical nodule <sup>7,8</sup>. When a patient presents with an umbilical nodule, fine-needle aspiration biopsy is adequate to establish an easy and early diagnosis <sup>9</sup>. Studies have shown that most umbilical lesions are primaries <sup>4</sup>; when they are metastatic (as in SMJN), most are diagnosed as adenocarcinoma <sup>5</sup>.

Among all reported cases of SMJN, 35%–65% metastasize from gastrointestinal malignancies; 12%–35%, from the genitourinary tract; 15%–30%, from unknown sites; and 3%–6%, from other malignancies such as those of the lung and breast <sup>10</sup>. A SMJN can sometimes be fissured or ulcerated, and it can also secrete serous, mucinous, purulent, or bloody discharge, depending on the primary tumour inside. The lesion has also been reported to be variously coloured: white, bluish violet, brownish red, and so on, with or without pruritus <sup>11</sup>.

The mechanism of umbilical seeding from primary tumours is not clearly understood; however, authors worldwide have proposed several hypotheses. One proposal is that a seeding process can occur through one or more routes: contiguous spread of peritoneal infiltration (the most common route) or through arteries, veins, or lymphatic channels. Spread through embryonic structures (such as the urachus, round ligament of liver, vitello intestinal duct remnant, or obliterated vitelline artery) is also proposed as a possible mechanism in certain cases. Some routes are relatively specific to given cancers—for example, pancreatic tumours spread through lymphatic system, and urinary bladder malignancies, through the urachus. It has also been observed that most gastrointestinal malignancies with SMJN also metastasize to liver. In those cases, it is apparent that the venous and lymphatic channels between liver and umbilicus are the probable means of mutual seeding, but whether the liver spread occurs first and then affects the umbilicus, or vice versa, remains unexplained <sup>12,13</sup>.

However the spread occurs, SMJN is an ominous sign of disseminated disease that is not amenable to cure. Mean life expectancy is 2–11 months without treatment. Recent reports have suggested that prognosis is better with aggressive management, the mean survival being 17.6–21 months with surgery and adjuvant chemotherapy. But surgery is usually recommended only in patients with a solitary umbilical metastasis. Surgery should be avoided in cases with widespread disseminated disease; in such cases, effective palliation can be achieved with chemoradiotherapy. The aggressive strategies belie the traditional concept that patients, once diagnosed with SMJN, should receive supportive care only. However, prognosis also depends on the primary lesion type and the organ affected. It has been noted that patients with ovarian cancer have a better survival rate than do patients with other malignancies <sup>5,14,15</sup>.

Sister Mary Joseph nodule is an uncommon manifestation of visceral and other malignancies.

Physicians need to be aware of this rare clinical condition so that they can promptly diagnose the primary cancer or its progression or recurrence.

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