

Sister Mary Joseph Nodule and peritoneal carcinomatosis from squamous cell cervical carcinoma

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Abstract

Sister Mary Joseph nodule is an eponym assigned to a nurse, first reported in 1949, denoting a rare form of cutaneous umbilical metastasis. The primary neoplasm is usually adenocarcinoma and the gastrointestinal tract is the most common primary site. The present case report describes a 79 year old woman with squamous cell cervical carcinoma. Her disease was managed with radiation and concurrent chemotherapy and no evidence of loco-regional recurrent disease was noted on follow-up examination. Eight months after treatment, gynecological examination suggested recurrent disease. Three weeks later, the patient presented with a painless umbilical nodule and the biopsy revealed a poorly differentiated carcinoma. Computed tomography showed ascites, diffuse peritoneal carcinomatosis including an umbilical nodule, multiple pulmonary nodules, bilateral pleural effusion, and retroperitoneal lymphadenopathy. She died 63 days after this manifestation. Review of the literature shows that a Sister Mary Joseph nodule is a sign of advanced neoplastic disease and is associated with poor prognosis. The treatment remains palliative either by radiation, chemotherapy or surgery.

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Introduction

The term “Sister Mary Joseph nodule” (SMJN) has been used since 1949 to denote umbilical metastases. Sister Mary Joseph was a nurse and the first observer to establish a correlation between umbilical nodules and carcinoma. SMJN is a rare but typical cutaneous periumbilical metastasis of internal malignancies, and its recognition is important because it may be the first presenting sign of unknown malignant disease in a patient.¹

The exact mechanism underlying metastases to the umbilicus remains unknown. Proposed routes for the spread of cancer to the umbilicus include direct extension from the peritoneum and spread via arteries, veins, or lymphatic channels.²

SMJN is most commonly found in association with adenocarcinomas, and the most common primary site is the gastrointestinal tract (35–65%). This metastasis impairs the quality of life and shortens survival.³

Although squamous cervical cancer

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is a common occurrence, the association of this condition with skin metastasis is rare.

A search of MEDLINE using search terms “umbilical metastasis* AND squamous cell cervix cancer” yielded 6 cases reported in the literature, none of them associated with peritoneal carcinomatosis.

Case Report

A 79-year-old Caucasian woman presented with postmenopausal intermittent vaginal bleeding and a vegetative mass external to the cervix. Gynecological examination revealed left parametrium infiltration.

Ultrasonography showed the presence of a 30-mm thickened endometrium, and cervical and

endometrial biopsy revealed a poorly differentiated tumor.

Immunohistochemistry showed that the cells were positive for p63 and Pan-Keratin and negative for cytokeratin 5/6 and S100. These features indicated the epithelial origin of the neoplastic cells and, as there were no criteria for squamous cell endometrial carcinoma, the patient was classified as having stage IIB cervical cancer, and the disease was managed with concurrent cisplatin-based chemoradiation. The pelvis was treated with 10 MV photons, 45 Gy in 25 daily fractions followed by brachytherapy. Follow-up examinations included Papanicolaou smear and pelvic examination for 6 months, which revealed no evidence of loco-regional residual disease.



Figure 1: Umbilical nodule

Eight months after chemoradiotherapy, gynecological examination revealed a pelvic mass and left parametrium infiltration, suggesting recurrent disease. Ultrasonography and magnetic

resonance imaging were requested.

Two weeks after this examination, the patient was evaluated for complaints of abdominal pain and a painless umbilical nodule (Figure 1).

On examination, a 3-cm exophytic necrotic lesion with mucoid and bloody discharge, ascites, and an abdominal mass growth were noted. A biopsy was performed and revealed a poorly differentiated carcinoma with extensive ulcerated necrotic areas infiltrating fibroadipose tissue.

As magnetic resonance was not available, computed tomography was performed and showed ascites, diffuse peritoneal carcinomatosis including an umbilical implant, multiple pulmonary nodules, bilateral pleural effusion, and retroperitoneal lymphadenopathy (Figures 2 and 3).

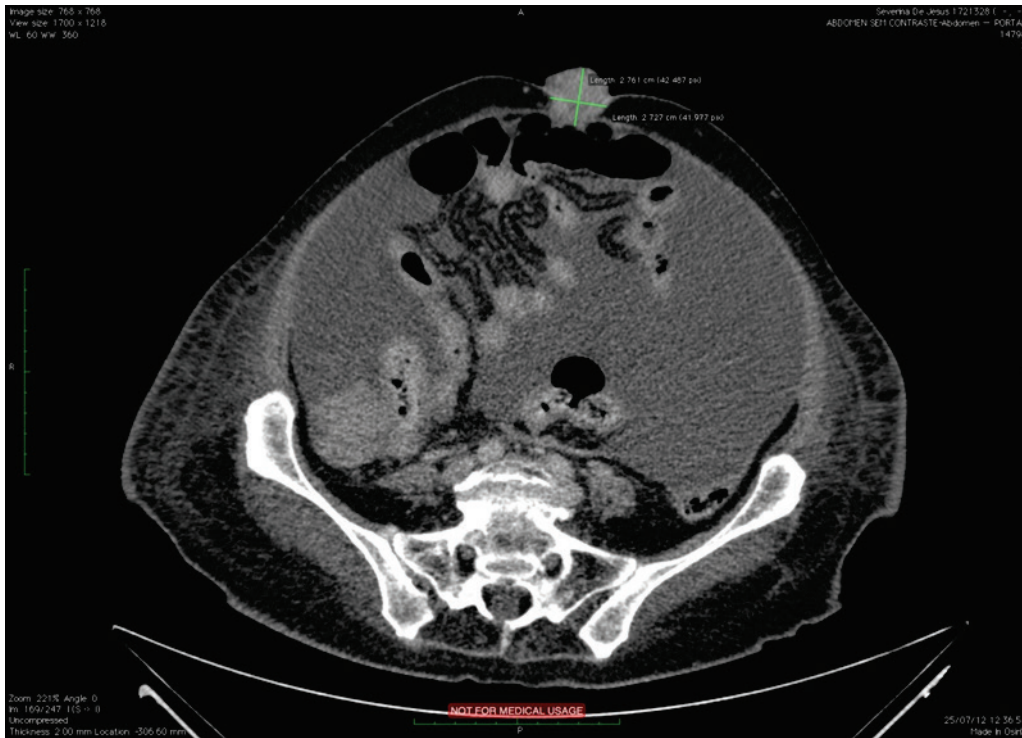


Figure 2: Computed tomography showing umbilical nodule

Paracentesis was performed, and hematic liquid had the following properties: mononuclear cells, 4.2 g/dL total protein, a serum ascites-albumin gradient of <1.1 g/dL, and negative culture.

The treatment was palliative with local excision.

The patient developed respiratory distress and intestinal sub occlusion. Death occurred 63 days after SMJN diagnosis.

Discussion

SMJN is an umbilical tumor usually associated with advanced cancer. An umbilical mass can present as a benign lesion, but also as a primary or metastatic malignant tumor.¹ Differential diagnoses include umbilical hernia, cutaneous endometriosis, pyogenic granuloma, melanocytic nevus, keloid, melanoma, squamous cell carcinoma, and basal cell carcinoma.³

Umbilical nodules are due to a

primary tumor and, despite being rare, they represent 38% of cases. Endometriosis is responsible for 32%, and metastases, 30%.⁴

SMJN is most commonly found in association with adenocarcinomas, and the most common primary site is the gastrointestinal tract (35–65%). The gynecological tract is responsible for 12–35% of cases, and in these cases, the ovary is the

most common primary site.³

SMJN may have different morphological characteristics, such as inflammatory erythematous thickening of the overlying skin, vascular appearance, fissures or ulceration, or features mimicking incarcerated umbilical hernia.³ In the present case, the nodule had a necrotic surface with bloody and mucoid discharge.

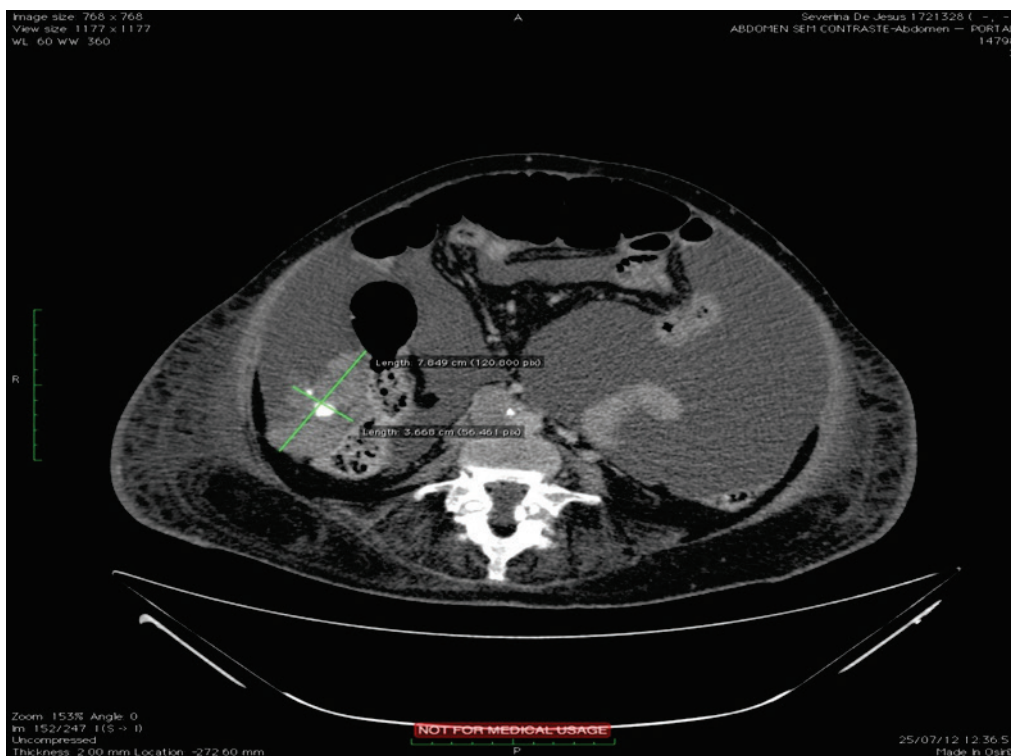


Figure 3: Computed tomography showing secondary peritoneal implantation in the right lower abdomen

Fagundes et al., in a retrospective analysis of 1211 patients with invasive cervical carcinoma, observed that clinical stage, endometrial extension prior to therapy, histology, and pelvic tumor control within each stage were indicators of distant dissemination. The frequency of metastases was greater when endometrial tumor extension was detected.⁵ Although

endometrial extension is not included in the International Federation of Gynecology and Obstetrics staging, this extension is associated with a high risk of lymph node metastases and decreased disease-free and overall survival.⁶

The superficial squamous tumor replaces the entire endometrium in the intrauterine spread of cervical carcinoma. The tumor must satisfy

three criteria (established by Fluhmann and modified by Kay) to be classified as primary endometrial carcinoma: 1) no coexistent endometrial adenocarcinoma, 2) no continuity between tumor and cervical squamous epithelium, and 3) no primary cervical carcinoma.⁷

Cervical carcinoma recurrence is most commonly loco-regional.⁸ Typical manifestations of recurrence involve the pelvis (preserved cervix, vagina or vaginal cuff, parametrium, bladder, rectum, ureters and ovaries) and lymph nodes.⁹ However, with the increasing use of pelvic irradiation, less typical patterns of recurrence such as peritoneal carcinomatosis can occur.¹⁰ These patterns are being recognized with greater frequency in patients with suspected recurrence because of the advancements in imaging studies.

Metastases from cervical carcinoma are usually predictable. Distant metastasis occurs commonly in the lung, liver, and bone.¹¹ Umbilical metastasis is extremely rare, and the histological type most frequently found is adenocarcinoma.¹²

Our patient recurred with ascites, diffuse peritoneal carcinomatosis, umbilical nodule, bilateral pleural effusion, multiple pulmonary nodules, and retroperitoneal lymphadenopathy.

Skin metastasis is a late manifestation in cervical cancer patients and is an indication of uncontrolled or widespread metastasis. Most skin metastases occur after an interval of less than 21 months. The mean survival of patients with umbilical metastasis is 3 months.⁸

We presume that the progressive involvement of the peritoneum due to squamous cell carcinoma might have led to spread to the umbilicus, and this was likely responsible for the unusual course of this case.

At the time of umbilical metastasis diagnosis, the cancer was deemed incurable and the umbilical metastasis was an indication of recurrence. SMJN treatment remains palliative either by radiation, chemotherapy, surgery alone or in combinations.⁸

Conclusion

Sister Mary Joseph nodule is a sign of uncontrolled disease. Although similar cases have been reported in the literature, this manifestation of the squamous cell cervical cancer is rare and this clinical course associated with endometrial extension and peritoneal carcinomatosis is unique.

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