

Sudden Onset of a Sister Mary Joseph's Nodule: A Case Report

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ABSTRACT

Sister Mary Joseph's nodule is an umbilical metastasis rarely seen in cancer patients. The primary focus usually is on the abdomen or pelvis. We aimed to present an umbilical metastasis detected in a 34-year-old female patient who was referred to our clinic for an umbilical mass without complaint.

ÖZET

Sister Mary Joseph nodülü kanser hastalarında nadiren görülen bir umbilikal metastazdır. Birincil odak genellikle karın veya pelvis üzerindedir. Bu olguda kliniğimize umbilikal kitle şikayeti ile başvuran 34 yaşındaki kadın hastada saptanan umbilikal metastazı sunmayı amaçladık.

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Introduction

Skin metastasis seen on umbilicus is defined as the Sister Mary Joseph's nodule. Sister Mary Joseph's nodule is a poor prognostic indicator in cancer patients and survival is reported to be 2-11 months in the patients who diagnosed of any cancer [2]. Primary cancer is frequently originated from gastric, colon and pancreas. The most commonly seen gynecologic malignancies with umbilical mass are ovarian and uterine cancers [3].

Case Presentation

A 34-year-old female patient admitted to clinic because of an umbilical mass. She had no systemic disease and had cesarean one year ago and there was no pathological finding then. Patient has complaining of umbilical mass that developed within 2 weeks and she has no abdominal distension or unexplained weight loss. Her umbilical nodule was about 2 centimeters diameter an irregular, ulcerated, painless nodule with blood (**Figure 1**). The lower abdomen CT revealed diffuse lesions in the right ovary and periovarian region, implants in the peritoneum and a mass of 8,5x5,5 cm in size, with cystic areas, heterogeneous contrast enhancement, and a clearly indistinguishable between uterine and bowel. Upper and lower gastrointestinal endoscopy was normal. Ca125 level was 691 U/mL. During laparotomy, primary tumour was on the right adnex. There was infiltration on the bladder, Douglas peritoneum and intestinal surfaces. Intraoperative frozen section was



Figure 1: Sister's Mary Joseph Nodule

resulted as high-grade serous carcinoma. Total abdominal hysterectomy, bilateral salpingo-oophorectomy, bilateral pelvic and para-aortic lymphadenectomy, infracolic omentectomy, peritonectomy, appendectomy and debulking surgery were performed.

Histopathologic examination revealed bilateral tubo-ovarian high grade serous carcinoma with invasion of the uterine serosa, myometrium, omentum, peritoneum (**Figure 2**). The subcutaneous tissue metastasis around umbilicus and one lymph node metastasis in the para-aortic region were found (**Figure 3**). The tumoral areas demonstrated solid, labyrinthine and cribriform architecture containing markedly atypical cells with large nuclei and high mitotic activity (**Figure 4**). Immunohistochemically, p53 diffuse strong nuclear positivity was detected in the tumor cells (**Figure 5**). Estrogen receptor (ER) and progesterone receptor (PR) were negative.

The patient received adjuvant chemotherapy containing carboplatin and paclitaxel after surgery but since the patient's treatment was performed in another center, other data could not be accessed. There is no recurrence in the patient's last PET-CT. The patient is living in good health in the 6th year after the surgery.

Discussion

Sister Mary Joseph (1856-1939) was born in New York in 1856 and was born in St. She started working as a nurse at Mary's Hospital.

She made a connection between intra-abdominal cancers and umbilical nodules. Spread to the umbilicus in intra-abdominal cancers occurs in 4 ways: 1) through the superficial or deep lymphatic system, 2)

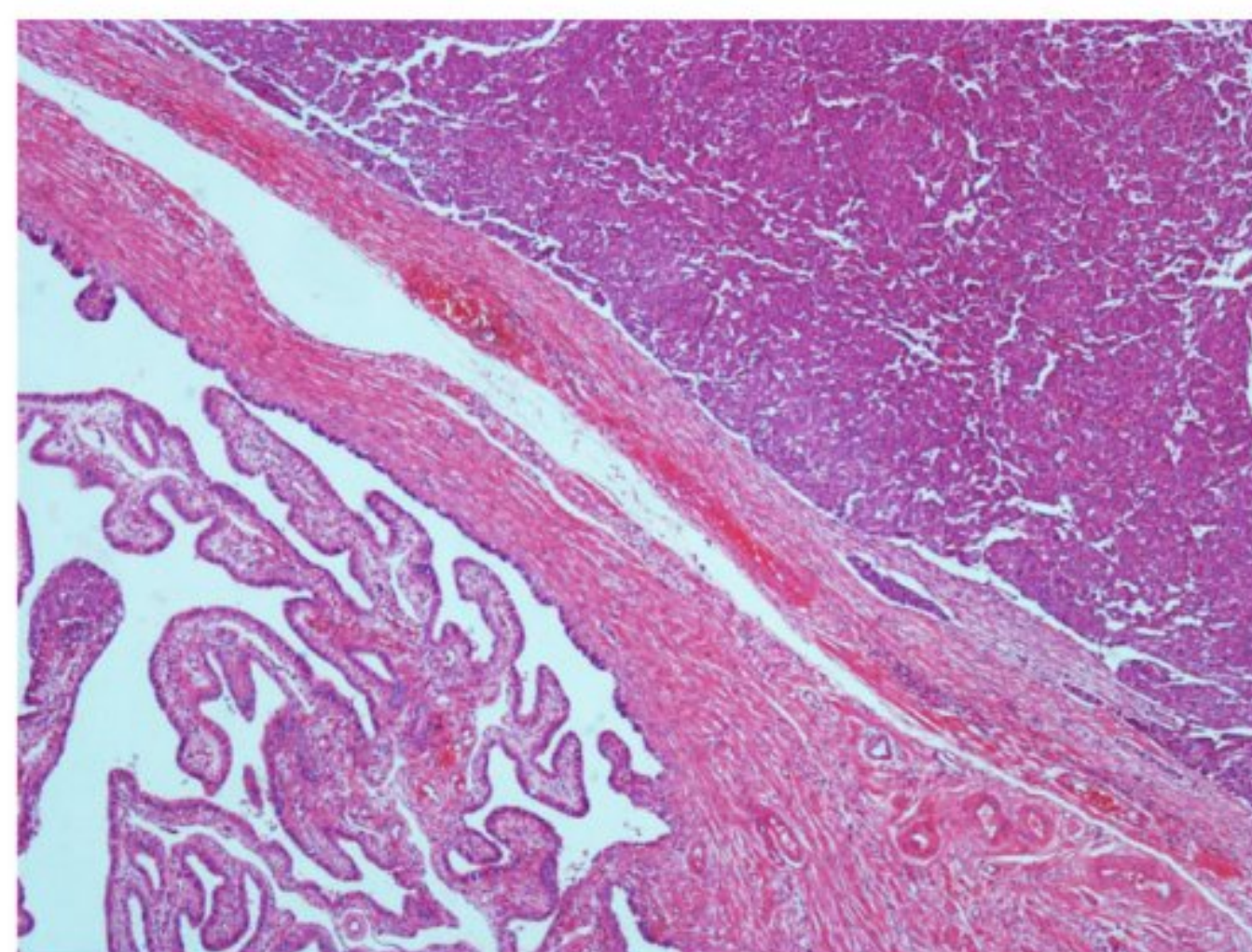


Figure 2: High grade serous carcinoma on the right and tubal epithelium on the left (H&E).

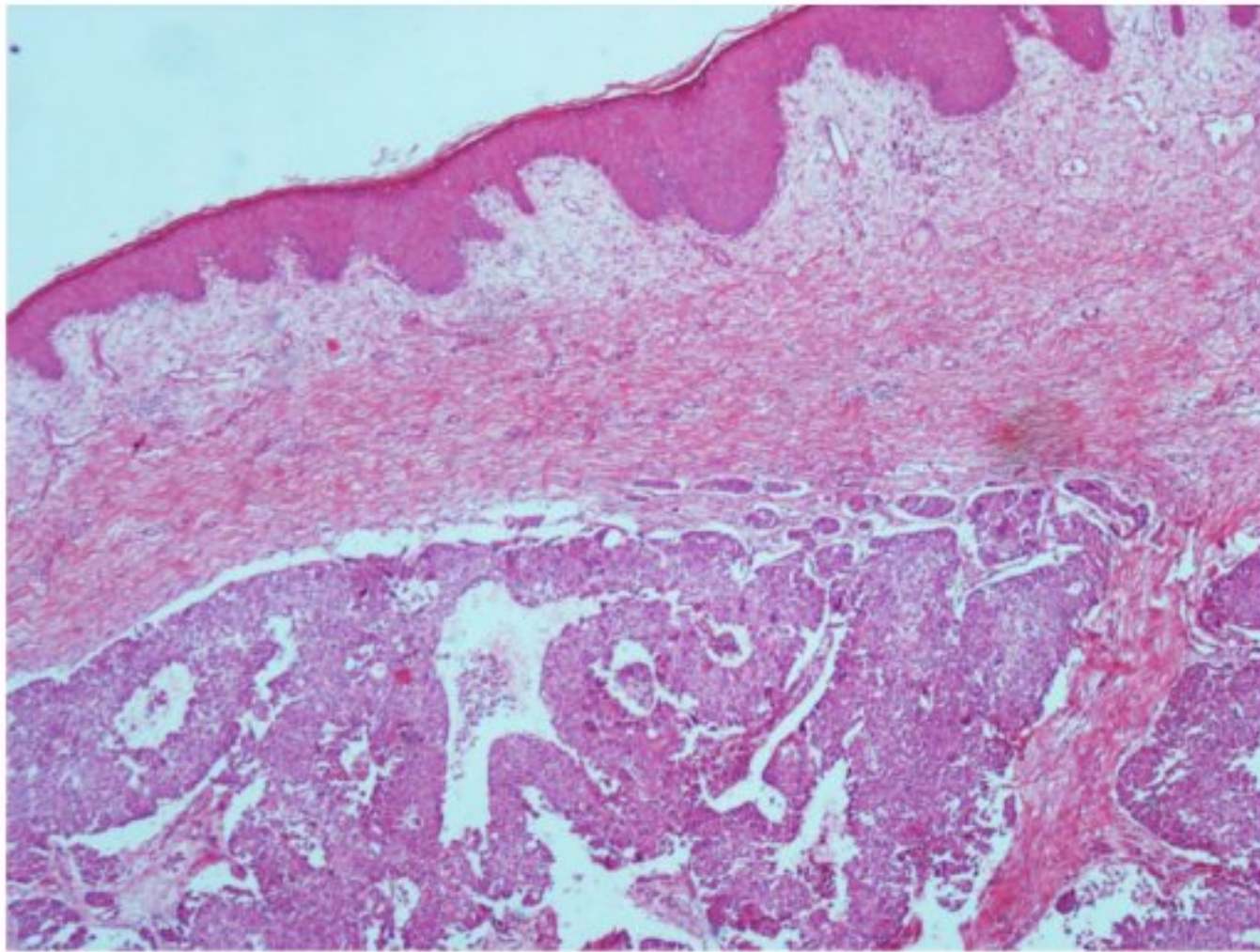


Figure 3: High grade serous carcinoma metastasis in the subcutaneous tissue around umbilicus (H&E).

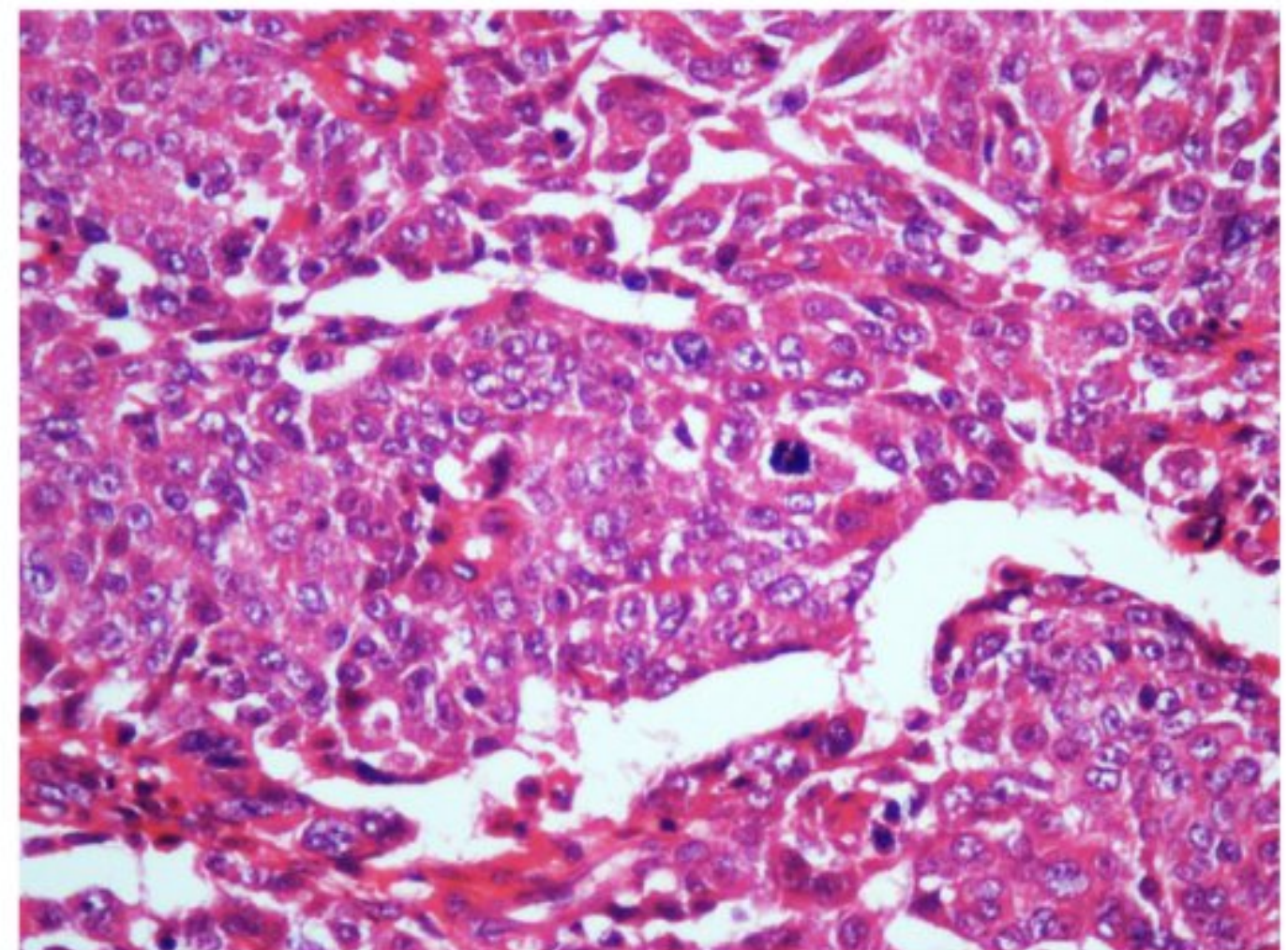


Figure 4: High grade serous carcinoma. Pleomorphic cells with large atypical nuclei and high mitotic activity (H&E).

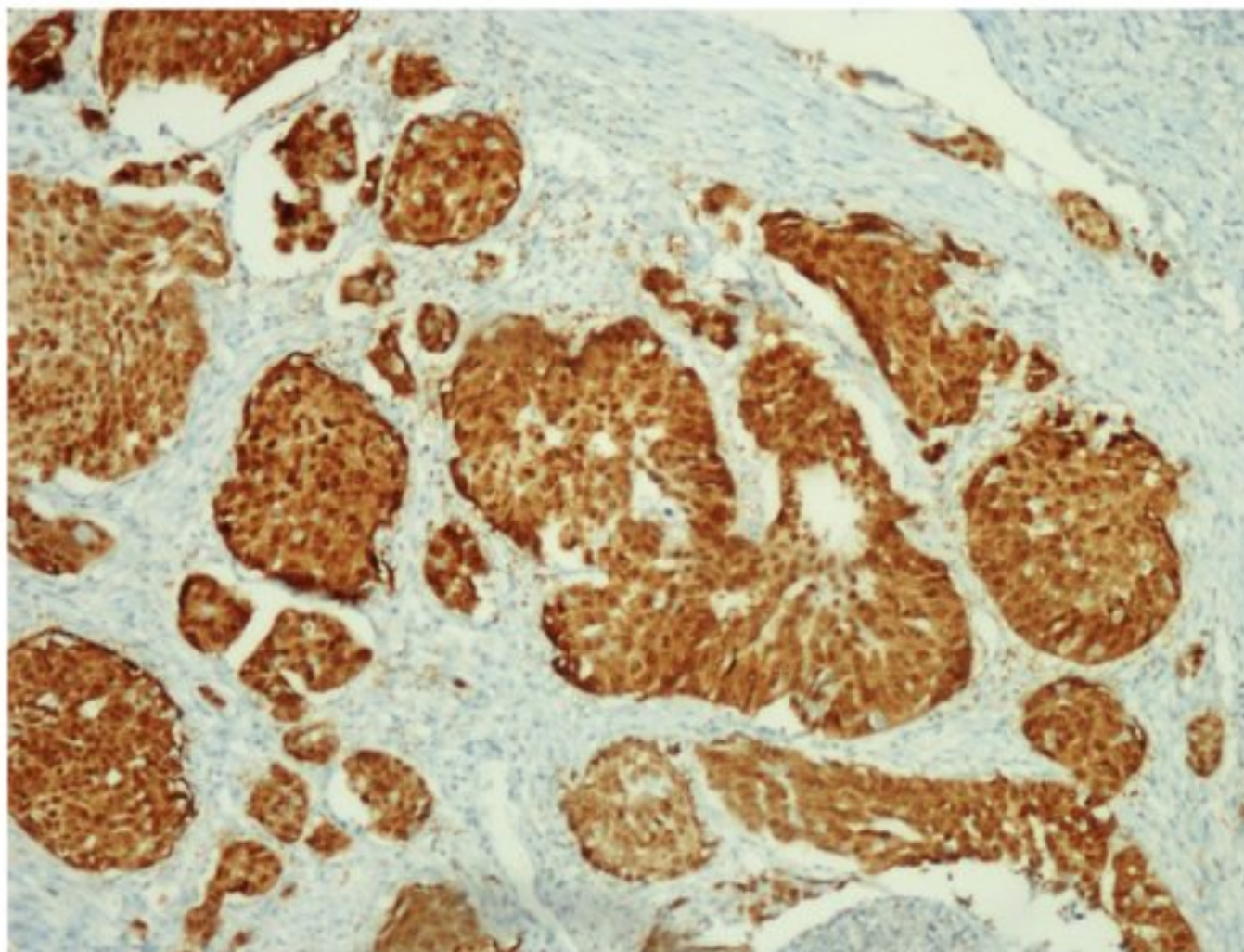


Figure 5: Mutant-pattern p53 immunostaining with diffuse strong nuclear expression.

by direct intraperitoneal spread, 3) by hematogenous spread, and 4) through the embryonic ligaments (4).

Galvan detected 24 uterine cancers in 407 umbilical tumor cases and reported that these were recurrent cases(5).

Primary tumors of the umbilicus are rare. It mostly occurs with metastasis of gastrointestinal system tumors. Distant metastases are thought to be caused by the hematogenous pathway. Umbilical hernia, endometriosis, granuloma and pilonidal sinus should be considered in the differential diagnosis. The tumoral mass may also be seen after the diagnosis of the primary tumor. The most common histological type is adenocarcinoma. The prognosis is bad. The average survival time has been reported to be 11 months (6). There is no consensus on treatment.

Malignancies originating from the pelvic organs should be kept in mind in people presenting with an umbilical nodule in the reproductive age. Differential diagnoses should be considered. Tissue sampling and imaging methods should be used for diagnosis. Systemic pelvic and paraaortic lymphadenectomy has been widely used in the surgical treatment of patients with advanced ovarian cancer, although supporting evidence from randomized clinical trials has been limited(7). During surgery was not associated with longer overall or progression-free survival than no lymphadenectomy and was associated with a higher incidence of postoperative complications(7).

Conclusion

Umbilical masses may occur with different clinical presentations like pyogenic abscess, epidermal cyst, hemangioma, abscess, umbilical hernia, endometriosis and primitive umbilical carcinoma. The physician should be careful about the umbilical mass that may be misdiagnosed as an umbilical hernia. Umbilical metastasis of primary malignancies is called as Sister Mary Joseph's nodule and it is rarely seen. The underlying causes of umbilical nodules should be well researched. As a result, although there are no specific clinical features of underlying condition, every umbilical mass should be examined carefully due to primary malignancies.

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