

Sister Mary Joseph Nodule and Ovarian Adenocarcinoma - A Case Report and Literature Review

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Abstract

SMJN is a rare superficial umbilical tumor metastasis that is most often associated with gynecological or gastrointestinal origin and usually reflects a poor prognosis. In this study, we report a case of Sister Mary Joseph Nodule in a patient with recurrent ovary adenocarcinoma. The patient, 56-year-old woman underwent ovarian cancer surgery 7 months ago. She received paclitaxel and carboplatin chemotherapy for a total of 5 courses after operation. As the tumor index continued to rise, considering the primary drug resistance, the patient was treated with oral etoposide. During the follow-up visit, the patient presented with an umbilical protuberance, measuring about 2 cm in diameter with a yellowish liquid spill which was malodorous. No contact bleeding was noted. Based on laboratory monitoring and imaging studies, ovarian cancer with a Sister Mary Joseph nodule (SMJN) was considered. The umbilical mass was resected and histo-pathological report showed a metastatic ovarian serous adenocarcinoma. The current incision healed well. In order not to miss a diagnosis of SMJN, it is very important to adequately examine patients.

Keywords: Sister Mary Joseph Nodule; Ovary Adenocarcinoma; Cutaneous metastasis; Case Report

Introduction

The metastasis or infiltration of a malignant tumor into the umbilicus is called a Sister Mary Joseph's Nodule (SMJN), a relatively rare lesion named after Dr. Mary Joseph's surgical assistant, Sister Mary Joseph [1]. She discovered the relationship between umbilical tumors and intra-abdominal malignancies, and subsequent studies have shown that SMJN is caused by intra-abdominal and or pelvic malignancies [2]. The incidence of SMJN is relatively low, accounting for 1%-3% of all intra-abdominal or pelvic malignancies. Most commonly it arises from a gastrointestinal tumor, in which case it accounts for 52%. Gynecological cancer as a primary source stands second, after gastrointestinal tumor. From the gynecological primary lesion, most commonly a SMJN comes from an ovarian or uterine cancer, which account for 28% of cases [3]. In ovarian cancer, approximately 0.91%-0.37% of patients have skin metastasis [4,5], however, in this case report, we bring forward a SMJN as a metastasis of an ovarian cancer. Furthermore, we also discuss the diagnosis, treatment and follow-up [6].

Case Presentation

Patient, 56 years old lady, self-noticed a mass of about 2 cm in diameter protruding from her umbilicus 9 months back (January 2019). It was accompanied by a bearable umbilical pain with a yellowish, foul smelling discharge. The patients do not complain about any contact bleeding. The patient underwent surgery 7 months prior to this, for excision of a pelvic mass suspected to be a high-grade serous adenocarcinoma of the ovary. During surgery, tumor infiltration was observed on the surface of the greater omentum and on the rectum.

Collected specimen were sent to the laboratory and the histopathological report was consistent with an ovarian serous adenocarcinoma. As a result, the final diagnosis was set as an ovarian cancer stage III C.

Physical examination: The general condition of the woman was satisfactory; no palpable superficial lymph nodes were noted.

Gynecological examination: the vulva and the vagina were normal, the cervix was absent and no definite connection between the umbilical mass and the pelvis was noted.

The blood analysis showed the following findings: Cancer

Antigen (CA) 12-5 level was 1520 U/mL, CA 19-9 level was 9.50 U/mL, CA 15-3 level was 114.2 U/mL, White Blood Cells Count: $3.09 \times 10^9 / L$.

Imaging study: Computer Tomography (CT) showed low-density nodules under the liver and soft tissue nodules under the skin of the umbilicus leading to the possibility of metastasis. Multiple metastatic lesions were seen in the abdominal cavity. Abdominal aorta showed enlarged lymph nodes, about 0.82×1.30 cm, and enlarged lymph nodes were also seen on the pelvic wall on both sides. The larger one was about 0.91×1.14 cm. Considering the patient's medical history and imaging examination findings, the diagnosis of a SMJN was established. The woman underwent an exploratory laparotomy.

During the operation: the umbilicus tumor could be seen reaching a size of 3.0×3.0 cm (Figure 1), located outside the peritoneum. The peritoneal surface and the surface of the intestine both contained miliary multiple tumor implantation. The umbilical tumor was completely resected and the surgery was uneventful.



Figure 1: Complete excision of the 2-3cm periumbilical mass.

Histopathological report showed a high-grade adenocarcinoma nodule under the skin, which was consistent with the implantation or metastasis of high-grade serous adenocarcinoma of the ovary (Figure 2). The incision healed well and the patient was discharged 7 days after surgery.

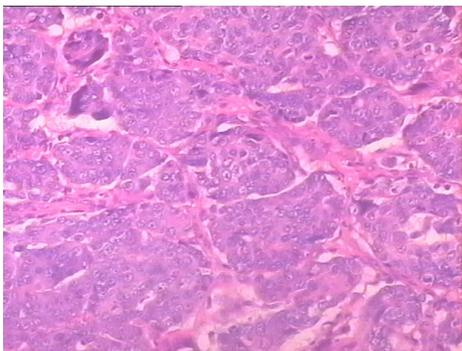


Figure 2: Histopathological slide.

Discussion

SMJN is named after Sister Mary Joseph (1856-1939). She was a nurse at the St. Mary's Hospital in Rochester, Minnesota (now the Mayo Clinic). She first observed a tumor or pelvic malignancy in the abdomen that was accompanied by an umbilical tumor and assessed the risk of umbilical metastasis [2]. In many reports, the most common primary cancer of SMJN is gastric cancer, followed by gastrointestinal cancer, and genital cancer. Among gynecological cancers, ovarian cancer is the most common [7,8]. Skin metastasis of ovarian cancer in the form of SMJN usually occurs in advanced ovarian cancer with extensive peritoneal involvement and has a poor prognosis [6]. It usually manifests itself as umbilical pain and ulcer formation, sometimes with hard nodules of pus, blood or serum. The average diameter is about 0.5 cm to 2 cm, however some nodules can go up to 10 cm in size [9]. The pathogenesis of SMJN in ovarian cancer may be blood-borne metastasis of arteries or veins, direct infiltration of peritoneal dissemination [10], accidental implantation after surgery, and diffusion of adjacent tumor cells through lymphatic vessels. The most common form of ovarian cancers that spread to the skin are those retrograde and come from lymphatic metastasis of the inguinal or para-aortic lymph nodes [10-12]. Studies have also reported that the rate of skin metastases in patients with ovarian lesions was as high as 17% [13], which was significantly higher than the previous study 0.91% - 3.7% [4,5]. However, the reason for this difference remains unclear.

SMJN is a late manifestation of malignancy [14]. In the absence of treatment, the average survival time of such women is 2 - 11 months [15]. It has been reported that the most appropriate treatment regimen is a combination of surgical resection, radiotherapy and chemotherapy. The average survival time is 17.6 - 21 months. However, because due to the progress of the disease and the fact that it is a metastatic tumor, sometimes it might require only palliative care [15-17]. In our case, this patient had a stage III C ovarian cancer and had undergone ovarian tumor cytoreductive surgery and chemotherapy. This time, the periumbilical metastasis has been removed and no further chemotherapy sessions were prescribed. The woman is now under follow up in the outpatient clinic and attends the hospital on a regular basis for further examinations and investigations, as and when required.

Conclusion

In routine clinical practice, patients with advanced ovarian tumors accompanied by an umbilical mass, the possibility of SMJN should not be excluded and should be adequately investigated for timely and proper treatment. In addition, timely detection of SMJN can yield a better outcome and prognosis from treatment offered.

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