A long-term survival case of Sister Mary Joseph’s nodule caused by colon cancer and treated with a multidisciplinary approach

Yoshinori Iwata1,4, Takashi Kinoshita1, Kenya Kimura1, Koji Komori1, Daisuke Hayashi1, Tomoyuki Akazawa1, Itaru Shigeyoshi1, Masayuki Tsutsuyama1, Jiro Kawakami1, Akira Ouchi1, Seiji Natsume1, Norihisa Uemura1, Yuichi Ito1, Kazunari Misawa1, Yoshiki Senda1, Tetsuya Abe1, Seiji Ito1, Masahiro Tajika2, Yasushi Yatabe3, Kazuhiro Yoshida2, and Yasuhiro Shimizu1

1Department of Gastroenterological Surgery, Aichi Cancer Center Hospital, Nagoya, Japan
2Department of Endoscopy, Aichi Cancer Center Hospital, Nagoya, Japan
3Department of Pathology and Molecular Diagnostics, Aichi Cancer Center Hospital, Nagoya, Japan
4Department of Surgical Oncology, Gifu University School of Medicine, Gifu, Japan

ABSTRACT

Umbilical metastasis from intra-abdominal or pelvic malignancy, which is called Sister Mary Joseph’s nodule (SMJN), is rare, and it has a poor prognosis. Its most common primary sites are the stomach and ovaries. SMJN caused by colon cancer is uncommon. A 42-year-old woman visited local clinics with complaints of an umbilical mass. After a detailed examination, she was diagnosed with peritoneal and umbilical metastasis caused by colon cancer. A radical surgery was performed after 12 months of chemotherapy. 6 months later, local recurrence and ovarian metastasis were suspected. Further radical surgery was performed, and 14 months after that (50 months after starting treatment), no recurrences have been observed. We experienced a long-term survival case of SMJN caused by colon cancer and treated with a multidisciplinary approach.

Keywords: umbilical metastasis, Sister Mary Joseph’s nodule, colon cancer, multidisciplinary treatment

BACKGROUND

Umbilical metastasis from intra-abdominal or pelvic malignancy is rare and is called Sister Mary Joseph’s nodule (SMJN) after Sister Mary Joseph who reported the association between umbilical nodules and advanced intra-abdominal malignancy.1,2 Its most common primary sites are the stomach and ovaries.3,4 SMJN caused by colon cancer is less common5,6 SMJN has been known to have a poor prognosis because most patients with SMJN have been diagnosed in their terminal stage.7,8 These cases can receive only limited treatment, including palliative surgery, mild
chemotherapy, or best supportive care. Herein, we report a long-term survival case of SMJN treated with a multidisciplinary approach.

CASE PRESENTATION

A 42-year-old woman visited local clinics with complaints of an umbilical mass. After a detailed examination, she was diagnosed with colon cancer, and the skin biopsy for umbilical tumor revealed adenocarcinoma. She was referred to our hospital for treatment. A contrast-enhanced CT showed ascending colon wall thickening, mesenteric lymph nodes swelling, an umbilical tumor lesion and an intraperitoneal nodule (Fig. 1). The tumor markers were found to be elevated (CEA, 272.7 ng/ml; CA19-9, 133.6 U/ml). We administered a combined chemotherapy regimen with fluorouracil, leucovorin, oxaliplatin, and bevacizumab. After 12 months of chemotherapy, both tumor markers were found to be in the normal range, and all lesions had shrunk or showed no change on CT (Fig. 2). In the next month, the tumor markers were found to be elevated again. Right hemicolectomy, omentectomy, and resection of umbilical tumor were then performed. Although two peritoneal nodules were found on the greater omentum, R0 resection was performed. Histopathological examination showed moderately differentiated adenocarcinoma in the ascending colon, peritoneal nodules, and umbilical tumor (Fig. 3). The grade of pathologic response was grade1b. Her postoperative course was uneventful, and we continued chemotherapy with the same regimen. After 4 months of postoperative chemotherapy and two months of follow-up, the tumor markers had elevated again. Fluorodeoxyglucose-positron emission tomography (FDG-PET) showed high accumulation on the anastomosis and right ovary (Fig. 4). Partial colectomy including anastomosis, bilateral oophorectomy, and the resection of the peritoneal nodule, which was found during surgery, were performed. Histopathological examination showed moderate adenocarcinoma in the anastomosis and peritoneal nodule, but not in the ovaries. After that, we administered only 3 courses of chemotherapy with fluorouracil, leucovorin, and oxaliplatin. No recurrences have been observed for 14 months after second surgery (50 months after starting treatment).

Fig. 1  A contrast-enhanced CT findings before treatment. CT showed ascending colon wall thickening (a), mesenteric lymph nodes swelling (a), an umbilical tumor lesion (b) and an intraperitoneal nodule (c).
Fig. 2  A contrast-enhanced CT findings after chemotherapy. All lesions had shrunk or showed no change (a, b, c).

Fig. 3  Histopathological examination findings. Histopathological examination showed moderate adenocarcinoma in the ascending colon (a) and umbilical tumor (b).

Fig. 4  FDG-PET findings. FDG-PET showed high accumulation on the anastomosis (a) and right ovary (b).
DISCUSSION

SMJN is a rare tumor metastasized to the umbilicus from intra-abdominal or pelvic malignancy. Umbilical metastasis was first reported by Walsh et al in 1846. In the 1920s these metastases were described as “trouser button navel” by William James Mayo. The association between umbilical nodules and advanced intra-abdominal malignancy was identified by Sister Mary Joseph; therefore, these nodules have been named SMJN. The most common primary site of SMJN in men is the stomach, followed by the colon, pancreas, and others. In women, the most common origin is the ovaries, followed by the stomach, colon, pancreas, and others. Although the mechanism of umbilical metastasis is not clearly understood, it is said that direct invasion including peritoneal dissemination is the most common pathway in gastrointestinal tumors.

The reported median survival after systemic 5-fluorouracil-based chemotherapy for colorectal cancer with peritoneal dissemination varies from 5.2 to 12.6 months. SMJN has a poor prognosis, and the overall survival is expected to be 2–11 months without treatment. Because most cases of SMJN have multiple metastases, they can receive only palliative surgery, mild chemotherapy, or best supportive care. A few cases of SMJN can receive aggressive chemotherapy or radical surgery as in the present case. In the present case, we had to select treatment considering the poor prognosis of SMJN and the resectable Stage IV colon cancer. In the Japanese Society for Cancer of the Colon and Rectum Guidelines 2014 for treatment of colorectal cancer, the following are described for distant metastasis associated with peritoneal dissemination:

- If both the distant metastases and the primary tumor are resectable, curative resection of the primary tumor is performed, and resection of the distant metastases is considered for Stage IV colorectal cancer.
- Complete resection is desirable for P1.
- Complete resection is considered for P2 when easily resectable.

In the present case, an umbilical tumor and an intraperitoneal nodule were the only distant metastases identified on CT. Both of the lesions were considered easily resectable. We considered performing radical surgery first, but considering the poor prognosis of SMJN, we feared that we would find multiple peritoneal dissemination during surgery or early recurrence after surgery. We decided that chemotherapy rather than surgery is preferable in this case. After 12 months of chemotherapy, we performed radical surgery because disease progression had been well controlled and no new lesions had appeared. It might be preferable that we should perform surgery before tumor markers were elevated. As a result, we could perform R0 resection although another peritoneal nodule was found on the greater omentum during surgery. The efficacy and safety of adjuvant chemotherapy after resection of distant metastases in colorectal cancer have not been established. We decided to use oxaliplatin again which was stopped before initial surgery. While we continued chemotherapy for four months, recurrences were suspected at 18 months after surgery. Although consensus on the treatment for local recurrence hadn’t established yet, previous systematic review suggested that long-term survival would be expected in about one half of patients who undergo R0 resection. We selected radical surgery for the recurrences because of the sufficient relapse-free survival time from the previous surgery and the high possibility of radical resection. In the present case, it was essential to perform chemotherapy for as long as possible and to perform R0 resection at the appropriate time.

CONCLUSION

Although SMJN has a high risk of recurrence and a poor prognosis, we could perform R0
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resection while continuing chemotherapy. We experienced a long-term survival (50 months) case of SMJN caused by colon cancer and treated with a multidisciplinary approach.

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