

## Case Report

# Pancreatic Cancer with Umbilical Metastasis (Sister Mary Joseph's Nodule)

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A 60-year-old woman was admitted to Nagasaki Prefectural Shimabara Hospital, with a painful tumor in her umbilical region. The tumor was about 1×1 cm in size. Histological examination of biopsied specimens revealed it to be a metastatic adenocarcinoma. Abdominal computed tomography and ultrasonography revealed a cystic tumor of about 23 mm in diameter in the pancreatic body. In addition, serum levels of cancer antigen 19-9 were elevated. With a tentative diagnosis of pancreatic tumor, she underwent surgery. When we opened her peritoneal cavity, there was no evidence of intra-abdominal disseminations, liver metastases or ascites. At that time, distal pancreatectomy accompanied by splenectomy was the procedure of choice. Histological examination revealed a moderately differentiated adenocarcinoma in the pancreatic body along with fatty replacement of the pancreatic tail. The umbilical tumor was a metastatic adenocarcinoma, which is referred to as a Sister Mary Joseph's nodule.

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## Introduction

A metastatic umbilical tumor is referred to as a Sister Mary Joseph's nodule. Sister Mary Joseph (1856-1939) worked as a surgical assistant to Dr. William Mayo at St. Mary's Hospital in Rochester, Minnesota. During that time, she recognized a metastatic umbilical nodule from an intra-abdominal malignancy. In her honor, Bailey<sup>1</sup> named metastatic umbilical tumors as Sister Mary Joseph's nodules. He stated that approximately 80% of malignant tumors in the umbilical region originated from carcinomas of stomach, colon and pancreas. We herein report a case of pancreatic cancer with Sister Mary Joseph's nodule, in which we diagnosed intraperitoneal malignancy from the presence of a metastatic umbilical tumor.

## Case report

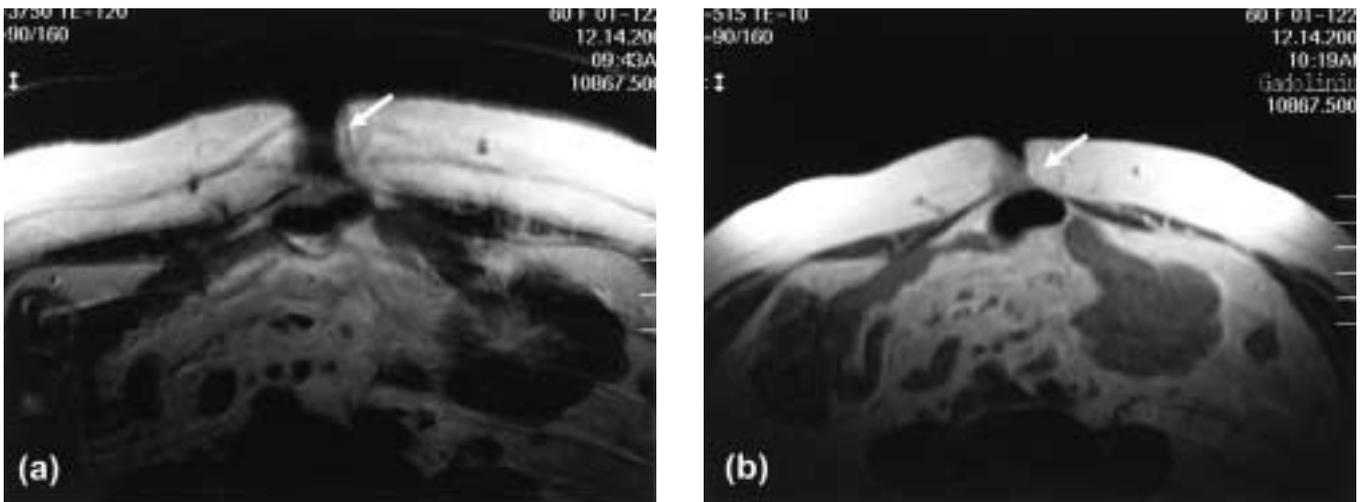
A 60-year-old woman was admitted to Nagasaki Prefectural Shimabara Hospital, with a painful tumor of about 1×1 cm in size in the umbilicus. The tumor had been gradually increasing in size over the previous 9 months. The tumor was found to be elastic and hard by pal-

pation, but its mobility was good. No infectious signs were detected. Complete blood counts and serum chemistries were within normal limits. In tumor markers, serum levels of cancer antigen 19-9 were 252 U/mL (normal level is 0 to 37 U/mL). Magnetic resonance imaging (MRI) revealed the umbilical tumor with a low intensity signal on T1- and T2-weighted images. In addition, it was evenly enhanced by gadolinium (Figure 1). Histological examination of biopsied specimens revealed a metastatic adenocarcinoma of unknown origin. Abdominal computed tomography (CT) revealed a cystic lesion of about 23 mm in diameter in the pancreatic body with fatty replacement of pancreatic tail and an elevated tumor at the fundus of gallbladder (Figure 2). Endoscopic retrograde cholangiopancreatography (ERCP) indicated disruption of the main duct in the pancreatic body, revealing a tumor of the gallbladder with smooth surface and gradual elevation (Figure 3). Abdominal ultrasonography detected a hypoechoic tumor in the pancreatic body that was contiguous to the cystic lesion. We considered her gallbladder tumor to be an adenomyomatosis. In addition, we judged that her umbilical tumor was a metastasis from pancreatic cancer. There was no evidence of metastasis in any other organ or regional lymph node, and no ascites were observed. The patient was considered to be a candidate for surgery, because imaging

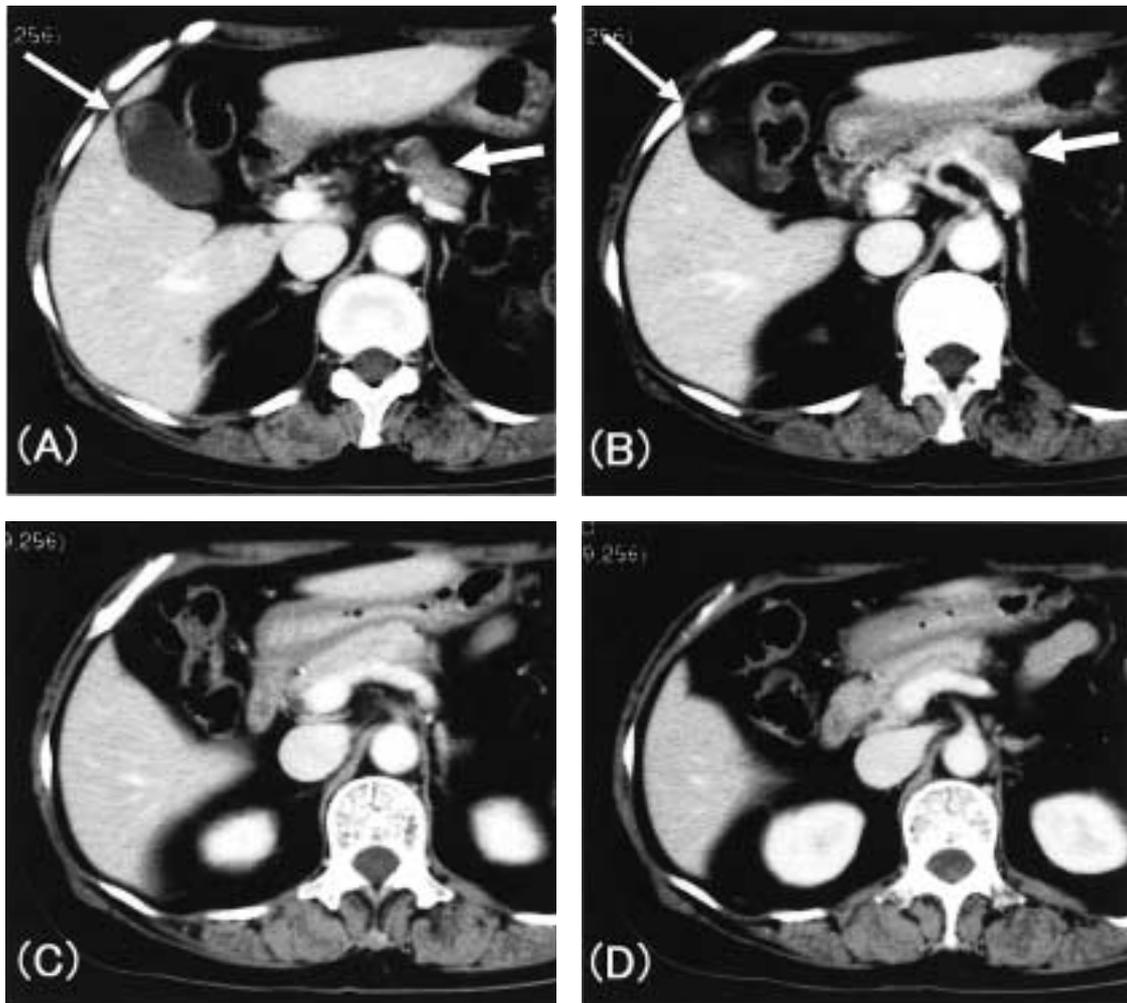
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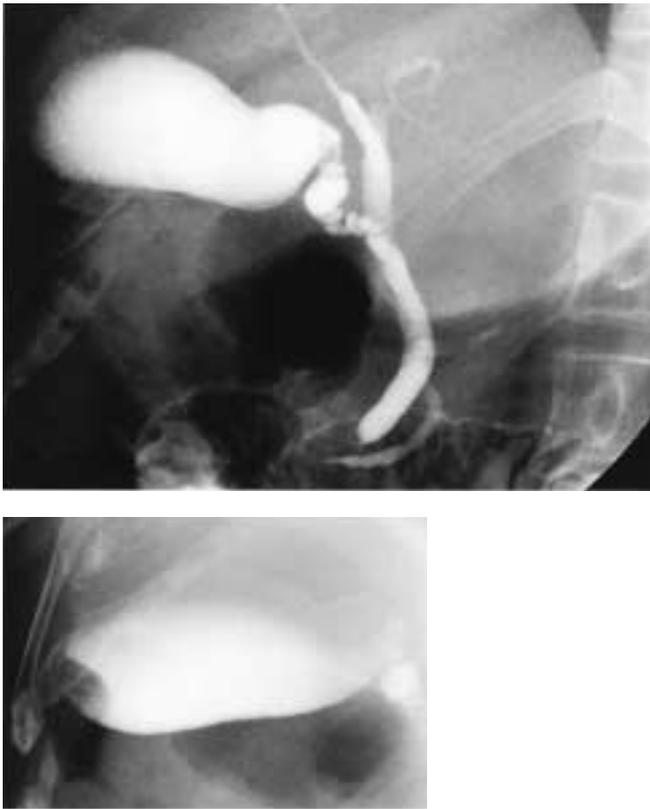
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**Figure 1.** Abdominal MRI revealed an umbilical tumor (arrows) of about 1×1 cm in size, with a low intensity signal on T2-weighted images (a) and with a gadolinium enhancement on T1-weighted images (b).



**Figure 2.** (A) and (B). Abdominal CT revealed a cystic low-density tumor of about 23 mm in diameter in the body of the pancreas (thick arrows). An adenomyomatosis was found at the fundus of the gallbladder (thin arrows). (C) and (D). The pancreatic tail was absent and was completely replaced by fatty tissue.



**Figure 3.** ERCP indicated disruption of the main duct in the pancreatic body, revealing a defect at the fundus of the gallbladder. The defect was gradually extended and its surface was smooth.

studies indicated that the main tumor of the pancreas and the metastatic umbilical tumor were localized. After the patient was informed of her condition, she chose surgery rather than chemotherapy or radiation therapy. On April 5, 2002, she underwent surgery. When we opened her peritoneal cavity, there was no evidence of any peritoneal disseminations, liver metastases or ascites. Cholecystectomy, distal pancreatectomy and splenectomy were performed on the patient. Histological examination of resected specimens revealed a moderately differentiated tubular adenocarcinoma of the pancreas along with a complete fatty replacement of the tail of the pancreas. Massive carcinomatous invasions were found in nerve fibers, in the main pancreatic duct and in fat tissues around the pancreas. Metastatic adenocarcinomas were also found in peripancreatic lymph nodes. In spite of postoperative adjuvant chemotherapy with Gemcitabine, both oophorectomies (a procedure used for removal of a Kruckenberg's tumor) were performed on August 23, 2002. About 6 months after the initial operation, she died as a result of a peritoneal recurrence of pancreatic cancer.

## Discussion

Umbilical metastatic tumors, which are referred to as Sister Mary Joseph's nodules, are relatively rare.<sup>1</sup> The primary site of this metastasis

found most frequently is gastric cancer, then followed by ovarian cancer, colorectal cancer and pancreatic cancer.<sup>2-10</sup> Of 80 collective cases in Japan, 15 cases (18.8%) originated from pancreas.<sup>10</sup> To our knowledge, only 20 cases (including our present case) have been reported in English as umbilical metastasis originating from pancreatic cancer.<sup>6-9</sup>

In pancreatic cancer, umbilical metastases are rarely found preceding diagnosis of the primary tumor.<sup>9,10</sup> Because pancreatic cancer is sometimes asymptomatic, an umbilical metastasis can be the first sign of the presence of pancreas cancer. With respect to the metastatic routes to the umbilicus, Steck and Helwig<sup>3</sup> have listed the following possibilities: (a) direct invasion from the peritoneum; (b) via lymphatic canals from the axillary, inguinal, or para-aortic nodes; (c) arterial or venous transportation; and (d) along embryonic ligaments. However, the etiology of umbilical metastases from the pancreas is still unclear. In pancreatic cancers that are accompanied by umbilical metastasis, primary tumors are commonly found in the body or tail of the pancreas.<sup>9</sup> This suggests that the metastatic route to the umbilicus can be explained by the anatomical formation of vessels or the proximity of the umbilicus (direct invasion). Because of the absence of any peritoneal dissemination during our patient's laparotomy, we have concluded that the metastatic route from pancreatic tumor to the umbilicus was likely due to via arterial, lymphatic vessels or embryonic ligament.

The average survival time after the diagnosis of primary tumor is 6 to 11 months,<sup>2,3,9,10</sup> so the prognosis of Sister Mary Joseph's nodule is considered to be poor. Clements<sup>11</sup> stated that the presence of metastatic umbilical tumor "almost certainly establishes the inoperability of the patient." However, some reports described that the outcome of these patients could be improved by surgical resection of both the primary organ and the skin tumor,<sup>3</sup> and that long survival time (18 years) was achieved by surgical intervention.<sup>4</sup> According to some reports, including research papers written in Japanese,<sup>12-16</sup> some patients with solitary metastasis of the umbilicus could survive for more than 1 year after the intervention of comprehensive therapy including surgery, chemotherapy and radiation. Obviously, if peritoneal disseminations or multiple distant metastases are present in addition to umbilical metastases, the prognosis will be very poor. However, in selected cases with umbilical metastasis only (as is the present case), surgical resection of primary tumor may be helpful for reducing the number of cancer cells to eradicate in subsequent chemotherapy. In our case, the patient survived for 15 months after umbilical tumor appeared.

Pancreatic cancer with Sister Mary Joseph's nodule is at advanced stage and is definitely a therapeutic challenge. However, extensive therapies, including surgery, might provide long-term survival in selected patients. So, when umbilical tumor is found, it is important to find the primary malignancy immediately; improvement in the patient's prognosis may depend on the site of this primary malignancy.

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