

Surprising Twist in the Plot – Sister Mary Joseph’s Nodule of Pancreatic Cancer Mimicking Wound Infection after Umbilical Hernia Repair

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Introduction

Umbilical hernia repair is the second most common type of hernia surgery after inguinal hernia repair. Depending on the surgical technique, complications occur in 5–25% and include wound infections and abscesses [1].

Pancreatic cancer is a fatal disease most commonly diagnosed at an advanced stage. It is typically asymptomatic or causes unspecific symptoms. Over 80% of the patients do not qualify for curative surgical resection due to locally advanced or metastasized disease. Common sites of metastasis include liver, lungs, peritoneum, and lymph nodes [2].

Umbilical metastasis originating from pancreatic cancer, also known as Sister Mary Joseph’s nodule (SMJN), is an extremely rare occurrence [3].

We report the cases of two patients who presented to our department with ulcerated umbilical lesions and had a history of umbilical hernia repair.

Case 1

A 62-year-old female patient was referred to our department complaining of an oozing and ulcerating umbilical lesion. She had a history of wound infection and open wound treatment

after external umbilical hernia surgery with mesh repair 3 months earlier. Additionally, she complained of an unintentional weight loss of 14 kg in the last few months, but denied other symptoms such as pain, night sweats, or autonomic changes.

Clinical examination revealed an ulcerating umbilical mass as the only remarkable finding (Fig. 1). The abdomen was soft, non-tender, and without distension. Ultrasound showed an inhomogeneous umbilical tumor with a diameter of 4 cm. Laboratory values were significant for white blood cell count ($12 \times 10^9/l$) and carbohydrate antigen (CA) 19–9 (730 U/ml). The tumor marker level was tested, because underlying malignancy was suspected due to her deteriorated general status. Suspicion of malignancy was then aggravated because of significantly elevated tumor marker levels. Therefore, a computed tomography was performed which depicted a tumor in the pancreatic tail measuring 4×6 cm infiltrating the celiac trunk and the splenic vessels (Fig. 2). Furthermore, hypodense masses were detected in liver segments 7/8 and 5. Suspecting an umbilical metastasis a biopsy of the umbilical mass was performed. Histological investigations confirmed a poorly differentiated adenocarcinoma with immunohistological signs of pancreatic cancer (Fig. 3). Palliative chemotherapy was suggested by the interdisciplinary tumor board. The patient received one cycle of chemotherapy with the FOLFIRINOX regimen. Unfortunately, her general condition worsened rapidly. She developed a small bowel obstruction due to peritoneal metastases and was transferred to a hospice where she died three months after the primary diagnosis of pancreatic cancer.

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Case 2

An 87-year-old female patient presented to our department with an ulcerating and oozing umbilical lesion. She had a history of umbilical hernia repair with mesh implantation

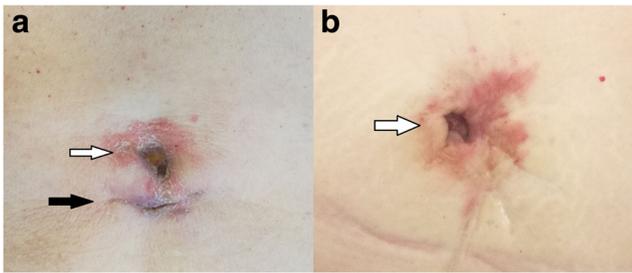


Fig. 1 Images of metastatic umbilical lesions in patients 1 and 2. White arrow: umbilical metastasis; black arrow (photo a): scar after umbilical hernia repair, which is not clearly seen in patient 2 (photo b)

12 years ago. She further complained of a weight loss of 10 kg within the last 10 weeks, loss of appetite, abdominal fullness and intermittent nausea.

Clinical examinations revealed a soft abdomen with a palpable ulcerating umbilical mass and no significant tenderness (Fig. 1). Ultrasound showed an inhomogeneous mass of 3 cm at the umbilicus surrounded by liquid strands above the mesh and ascites. Laboratory values were significant for gamma-glutamyl-transferase (274 U/l), and C-reactive protein (27 mg/l). CA 19–9 and carcinoembryonic antigen levels were extremely elevated measuring 19,195 U/ml and 261 ng/ml, respectively. Tumor marker levels were primarily tested, because a malignancy was suspected due to a worsened general condition of the patient. Further investigations were initiated to identify the origin of the suspected malignancy. A CT-scan of the abdomen revealed a huge mass in the pancreatic tail measuring 4.5×7.7 cm with infiltration and encasement of the celiac trunk, the splenic vessels and the superior mesenteric artery (Fig. 2). Furthermore, diffuse ascites and a hypodense liver mass in liver segment 4a were detected. Colonoscopy, gastroscopy, and the gynecologic examination were unremarkable.

During the hospital stay, the patient suffered a stroke with resulting motoric aphasia. She was transferred to the neurologic ward for 2 days and received conservative treatment. Back on the surgical ward, the patient refused further diagnostic workup and was referred to the palliative ward. Although not histologically proven, the patient suffered from hepatic and peritoneal metastases of pancreatic origin, based on the results of the CT-scan and the extremely elevated CA 19–9

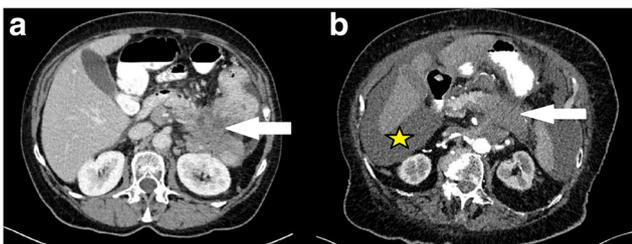


Fig. 2 Contrast-enhanced CT-scans of patients 1 and 2. White arrow: pancreatic ductal adenocarcinoma of the pancreatic tail; yellow star: perihepatic ascites

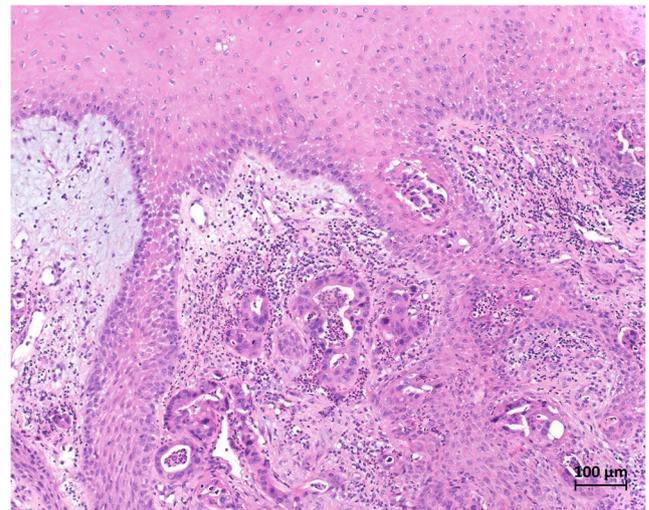


Fig. 3 Pathohistological findings of a poorly differentiated pancreatic ductal adenocarcinoma infiltrating the subcutaneous tissue at the umbilicus

level. She died two months after the primary diagnosis of pancreatic cancer.

Discussion

SMJN refers to an umbilical cutaneous metastasis typically originating from an abdominal or pelvic malignancy. It is named after Sister Mary Joseph, born Julia Dempsey in 1856 and first surgical assistant to William James Mayo. She observed the association between firm umbilical nodules and metastasized abdominopelvic malignancies which was published by Hamilton Bailey in 1949 [4].

Abdominopelvic malignancies rarely metastasize to the umbilicus; however the presence of a metastatic umbilical lesion leads to the diagnosis of the primary malignancy in 64%. Typical sites of origin include the stomach in men (30%), the ovaries in women (34%), and the small and large bowel (20%). The pancreas is involved in 7–10% of all cases with SMJN [3, 5].

Several theories regarding the pathophysiological background of SMJN have been proposed. According to Sugarbaker and Powell, umbilical metastasis develops via contiguous extension of abdominopelvic tumor cells associated with peritoneal metastases. Tumor spread through lymphatic channels is considered highly unlikely since there are no lymph nodes in and around the umbilicus. Hematogenous metastasis is considered improbable in the absence of other blood borne metastatic sites [6, 7].

Pancreatic cancer is a fatal disease most commonly diagnosed at an advanced stage. More than 80% of all patients suffer from locally advanced or metastasized tumors at the time of diagnosis [2]. The average survival for patients with SMJN is less than 4 months with an overall 2-year survival

rate of 3.5%. Typical regions of metastasis include regional lymph nodes, liver, lung, and peritoneum [5].

The two patients in our report presented with ulcerated, oozing, firm umbilical nodules. Both had been referred to our department due to a history of umbilical hernia repair and suspected complication following this repair. Up to date, no similar reports have been reported in literature.

According to the local findings (Fig. 1), postoperative complications were a possibility, especially in patient 1 who had just recently undergone hernia repair. However, the history of deteriorated general status with significant weight loss led to further diagnostic studies. Extremely elevated CA 19–9 tumor marker levels raised the suspicion of an underlying malignancy. Tumor marker levels should always be tested if abdominopelvic cancer is suspected.

In our cases, complete staging including a contrast-enhanced multidetector CT-scan of the abdomen, gastroscopy, colonoscopy and gynecological examinations were performed.

The CT-scan revealed malignancy of the pancreatic tail in both cases, which is in accordance with the literature where 91–92.3% of pancreatic tumors in patients with cutaneous metastases are found in the pancreatic body or tail [5, 8].

A biopsy of the umbilical mass to confirm the underlying malignancy is easily performed and should be considered in all patients with a suspected advanced tumor stage. Immunohistological staining for CK 7 and 19 allows the diagnosis of pancreatic cancer [9]. In those cases, obtaining tumor specimens via abdominal surgery to confirm malignancy becomes dispensable.

Although surgical techniques and palliative therapy including chemo- and chemoradiation therapy have improved, survival rates are still low in pancreatic cancer. Palliative therapy is often the only option for these patients [2]. SMJN can be highly suspected based on the visual diagnosis and the case history. It is associated with an advanced tumor stage and a poor prognosis. The mean survival rate in our patients was 2.5 months. Survival of up to 19 months after the primary diagnosis of pancreatic cancer with umbilical metastasis was reported in a single case report [10].

Conclusion

A history of umbilical hernia repair should not exclude umbilical metastasis originating from an abdominopelvic

malignancy as the underlying cause for an ulcerating umbilical lesion.

SMJN can present as an initial sign of advanced malignancy with poor prognosis and facilitate the diagnosis of the primary disease.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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Human and Animal Rights and Informed Consent This article does not contain any studies with human participants or animals performed by any of the authors.

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