Sister Mary Joseph’s nodule as a first sign of pancreatic cancer

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Abstract

Sister Mary Joseph’s nodule (SMJN) refers to a metastatic tumor of the umbilicus. It is a rare entity which arises from a malignancy in the intra-abdominal cavity. We herein describe a patient who presented with SMJN as his first sign of pancreatic cancer. It is an even more unusual case of SMJN. We therefore, suggest that pancreatic cancer should be included in the differential diagnosis when an umbilical mass is found. With the progress made in surgical procedures and other modalities, an early diagnosis will dramatically improve the prognosis of the patients.

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Key words: Sister Mary Joseph’s nodule; Pancreatic cancer; Umbilical metastasis; Diagnosis; Management

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INTRODUCTION

Umbilical tumors are rare and 30% of them are metastatic[3]. Sister Mary Joseph’s nodule (SMJN) refers to a metastatic lesion of the umbilicus originating from intra-abdominal or pelvic malignant diseases. The most common primary sites of the metastasis are stomach, ovaries, colon, rectum and pancreas. Pancreas accounts for 7%-9% of the SMJN cases[2]. Almost 90% of the cases arise from the body and tail of the pancreas, but not the head[3].

We herein describe a male patient with pancreatic cancer who presented with SMJN as his first clinical sign. We also reviewed the published literatures on this disease from PubMed. Only 20 cases (including our case) of SMJN with pancreatic cancer as the first symptom and sign have been reported in English. The aim of this report is to provide new insight into the identification of pancreatic cancer presenting as SMJN.

CASE REPORT

The patient is a 40-year-old man who presented with...
swelling, redness and induration at umbilicus. The patient was first diagnosed with eczema at a local hospital and was treated without improvement. After six months, the patient consulted a dermatologist at our hospital. Physical examination showed an inflamed umbilicus with a nodule of 2 cm × 1.5 cm without ulcer (Figure 1A). An umbilical skin biopsy was taken and sent for pathologic examination. The pathology was suggestive of adenocarcinoma infiltrating dermis and epidermis (Figure 1B). Immunohistochemistry (IHC) showed cytokeratin (CK) (AE1/AE3) +++, carcinoembryonic antigen ++++, and CK7 +. A metastatic adenocarcinoma was suspected based on all these findings.

Serum marker of carbohydrate antigen 19-9 (CA19-9) was elevated (202.1 U/mL). Other tumor markers were all in the normal range. An abdominal multi-detector computed tomography (MDCT) revealed a tumor at the neck of the pancreas with retroperitoneal lymph nodes enlargement as well as an umbilical soft tissue mass (Figure 2). An abdominal computed tomography (CT) angiography also showed involvement of retroperitoneal lymph nodes, superior mesenteric artery and vein, celiac trunk, proximal splenic artery, splenic vein and part of portal vein. A fluorodeoxyglucose (FDG) positron emission tomography-CT (PET-CT) scan demonstrated a mass located at the neck and body of pancreas with increased FDG uptake, with a maximal standardized uptake value (SUV) of 3.69, and thus malignancy was suspected. The umbilical mass with a maximal SUV of 2.85 was also detected and a metastatic malignancy was diagnosed by PET-CT (Figure 3).

Preoperative abdominal CT angiography showed the
tumor encasing celiac trunk and superior mesenteric artery, which, however, did not cause the occlusion or the stricture of theses arteries. According to our previous experience, the patient may still have a chance of resection. An exploratory laparotomy was then performed. Mild abdominal ascites was present. A hard mass of 8 cm at the neck and body of pancreas was discovered, with invasion of celiac trunk, superior mesenteric artery, superior mesenteric vein and portal vein. Several enlarged mesenteric lymph nodes and an umbilical mass of 2 cm × 1.5 cm as well as tiny diffused seeding nodules were observed. Diaphagmatic tumor invasion was visible. A biopsy was taken and a frozen section was sent for pathology. Adenocarcinoma infiltrating the striated diaphagmatic muscle tissues was confirmed. Surgical resection was abandoned because of distant metastasis of the cancer. The patient refused chemotherapy, and died four months later.

**DISCUSSION**

When a clinician observes an umbilical mass, his/her differential diagnosis should include both benign and malignant tumors. Benign tumors can be caused by a number of factors, including polyps, papilloma, myoma, fibroma, hemangioma, dermoid cyst, teratoma, pyogenic granuloma, omphalith, or endometriosis. Malignant lesions account for 38% of all umbilical tumors, including both primary and secondary cancers. These can be differentiated by means of biopsy collection and histopathological examination. Cytology can not distinguish between primary and secondary malignancies.

The incidence of metastatic tumor originating from intra-abdominal malignancy to the umbilicus (SMJN) is very low. The most common primary sites of SMJN in men are stomach, followed by colon, rectum, pancreas and others. In women, the most common origins are the ovaries, followed by stomach, colon, rectum, pancreas and others. Pancreas is the fourth or fifth most common primary site of SMJN. The percentage of SMJN arising from pancreas ranges from 7% to 9%, among which SMJN presenting as the first sign of pancreatic cancer is even unusual. To our knowledge after literature review, only 20 such cases, including our case, have been reported in English.

The metastatic routes of the pancreatic cancer to the umbilicus are thought to be by direct invasion from the peritoneum, through lymphatic or blood vessels and along embryonic remnants. Generally, the prognosis of pancreatic cancer with SMJN is poor. The mean survival is 6-11 mo. However, with early diagnosis and surgical intervention, a prolonged survival of 18 years was reported. Currently, with the improvement of surgery, the resection rate of advanced pancreatic cancer has been increased. Moreover, with advances made in multidiscipline therapeutic modalities including chemotherapy, radiotherapy, targeted molecular therapy, immunotherapy, and others, the survival of the patients has also been improved.

It has been reported that resection of SMJN and the primary cancer of pancreas combined with other therapeutic modalities has improved the prognosis of the patients. Therefore, an appropriate diagnostic algorithm is very important for early detection and rational treatment.

As mentioned above, for patients with umbilical nodules as the first clinical manifestation, biopsy should be performed first if the diagnosis is uncertain. Almost all the cases of SMJN from pancreatic cancer reported in the literature are adenocarcinomas. Serum tumor markers such as CA19-9 should be examined and elevation of CA19-9 is considered as a strong evidence of the disease. Once SMJN deriving from pancreatic cancer is suspected, contrast-enhanced MDCT or magnetic resonance imaging (MRI) on pancreas should be done. Contrast-enhanced MDCT or MRI is sensitive enough to display the pancreas mass and its involvement in the adjacent organs and vessels. PET-CT is also recommended to detect other distant metastasis. However, PET-CT is not sensitive to the identification of tiny seeding nodules in the intra-abdominal cavity. As in this case, PET-CT failed to display intra-abdominal diffusion.

An earlier diagnosis could have caught the tumor in its early phase, hence improving the prognosis of the patient. This paper is to remind physicians and surgeons of the importance of keeping in mind pancreatic cancer as one of their initial differential diagnosis when an umbilical nodule is presented. The recommended diagnostic algorithm is hopefully useful for the early detection of pancreatic cancer which appears as SMJN as its first sign.

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