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CASE REPORT | PANCREAS

Sister Mary Joseph Nodule as an Initial Presentation of Pancreatic Adenocarcinoma

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ABSTRACT

Sister Mary Joseph (SMJ) nodule is a metastatic umbilical nodule seen in primary tumors of the gastrointestinal or genitourinary tract. The stomach and colon are the common gastrointestinal cancers associated with SMJ nodule. The pancreas is a rare primary site for umbilical metastasis. Moreover, malignant umbilical nodules as the first presentation in pancreatic cancer is rare. Pancreatic adenocarcinoma that metastasizes to umbilicus usually arise from the body or tail of the pancreas. The presence of SMJ nodule usually indicates poor prognosis. Umbilical nodule is a simple physical finding to the presence of an advanced intra-abdominal malignancy. Although rare, pancreatic cancer should be considered as one of the primary sites in such a situation.

INTRODUCTION

Sister Mary Joseph (SMJ) nodule is a metastatic umbilical nodule seen in primary tumors of the gastrointestinal or genitourinary tract.[1,2] Pancreas is a rare primary site for umbilical metastasis. We report SMJ nodule as the initial clinical presentation in an elderly man with pancreatic cancer and review the relevant literature on this topic.

CASE REPORT

A 79-year-old man presented with painful nodule in the umbilicus for 4 weeks. He gave a history of back pain and weight loss during this period. There was no epigastric discomfort, vomiting, gastrointestinal bleed, jaundice, or palpable mass in the abdomen. On examination, a hard pigmented nodule measuring 2 × 2 cm in size was noted in the umbilicus. The skin over it was fixed, and there was no discharge (Figure 1). Complete hemogram showed a hemoglobin of 10 g/dL. His blood sugar, renal, and liver function tests were normal. Abdominal computed tomography (CT) showed a nodule at the umbilicus and a heterogeneously enhancing mass measuring 7 × 5 cm, occupying the body and tail of the pancreas (Figure 2). His CA19-9 was 1,937 U/mL. Fine needle aspiration cytology of the nodule showed cohesive clusters of tumor cells, which were large with moderate to abundant pale blue cytoplasm, irregular nuclear membrane, large vesicular nucleus with opened up chromatin, and few conspicuous nucleoli, and the cells were positive for cytokeratin (CK)-19 in immunocytochemistry (Figure 3). Endoscopic ultrasound showed a hypoechoic mass in the body and tail of the pancreas encasing the splenic artery, compressing the portal confluence and adjacent segments of the superior mesenteric and splenic vein. Few celiacs and peripancreatic group of nodes were enlarged, the largest measuring 1.5 × 1.1 cm. Minimal ascites was present. Pancreatic adenocarcinoma was confirmed by endoscopic ultrasound-guided biopsy. The diagnosis of an advanced primary pancreatic malignancy with metastasis to the umbilicus was considered. He was referred to oncology services for palliative treatment.

DISCUSSION

Umbilical metastasis is rare in clinical practice and represents only 10% of all cutaneous metastasis in cancer.[2] Two-third of primary lesions arise from the gastrointestinal tract, stomach, and colon being the more common sites. The origin of the primary

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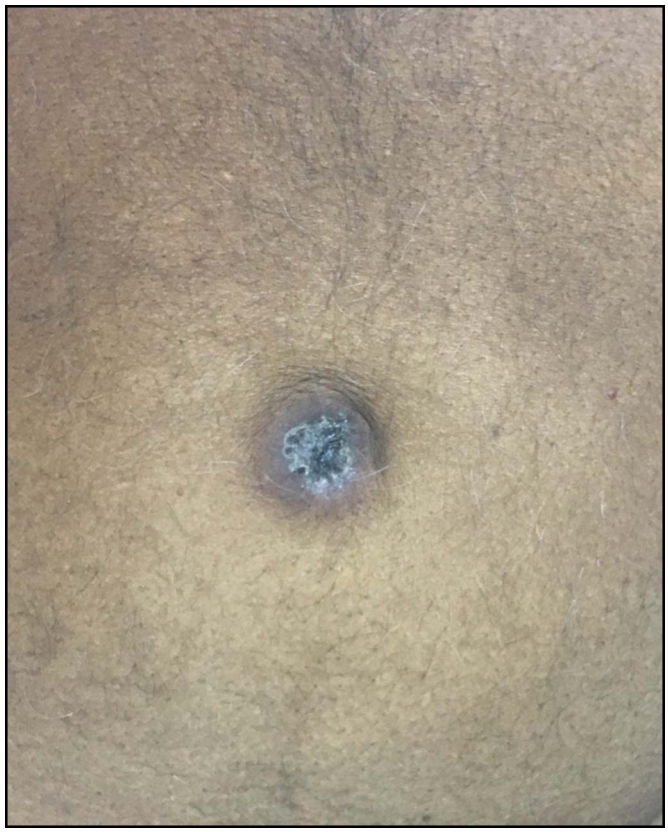

Figure 1. Abdominal examination showing a pigmented umbilical nodule.

tumor is unknown in 15%–30% patients.[1–3] The pancreas is a rare primary site for SMJ nodule. Approximately 6% of umbilical metastases are pancreatic in origin.[4]

The common sites of metastasis in pancreatic cancer are lymph nodes, peritoneum, liver, lung, kidney, bone, and brain.[5] Unlike the usual site of distribution of pancreatic tumor, pancreatic adenocarcinoma that metastasize to umbilicus arise from the body or tail of the pancreas.[3,6] Patients with pancreatic malignancy often have a dismal prognosis owing to presentation in the later stages of the disease.[1,3]

SMJ nodules are usually detected during or years after the diagnosis of the primary neoplasm. However, they may be the initial or only presenting sign of an advanced malignancy. Malignant umbilical nodules as the first presentation in pancreatic cancer is rare.[1,6–9] In a large published review of umbilical tumors, 9% of cases were from the pancreas.[10] The exact mechanism of its spread to the umbilicus is not known. Intraperitoneal dissemination and implantation of exfoliated pancreatic tumor cells on to the umbilicus or direct invasion from peritoneal metastasis are the most common mechanisms responsible for a SMJ nodule. Other postulated modes of spread to the umbilicus include invasion through arteries, veins, lymphatics, or via the umbilical ligament.[3,9]

SMJ nodules are painful, indurated, irregular, and often hard in consistency. The surface may be ulcerated or necrotic with serous, serosanguinous, or pus discharge. They are usually 0.5–2 cm in size but can progressively enlarge up to 10 cm.[1] Umbilical nodules can also occur in other conditions such as mycosis, angioma, endometriosis, pyogenic granuloma, eczema, Paget disease, teratoma, dermoid cyst, or hypertrophic scar.[2,3] The diagnosis is usually made by fine needle aspiration cytology, which has excellent sensitivity and positive predictive value.

Adenocarcinoma constitutes 75% of SMJ nodules. Some of the other histological findings reported include squamous cell carcinoma, anaplastic carcinoma, non-Hodgkin lymphoma, and cholangiocarcinoma.[1] Immunohistochemistry studies of SMJ nodule for CK will help in the evaluation and classification of unknown primary tumors. Although CK 8 and CK 18 expression is from exocrine, endocrine, and ductal epithelial cells of the pancreas, CK 7 and CK 19 are usually from the ductal cells. In pancreatic ductal adenocarcinoma, more than 90% of cases show expression of CK7 and 50% show positivity for CK 19.[11,12] The expression of CK 20 was variable, which had an impact on the clinical outcome. In addition, the elevation of CA19-9 is a strong evidence of pancreatic cancer. Our patient had a pancreatic body and tail mass, significantly elevated CA 19-9, tumor cells in umbilical nodule positive for CK 19, and histopathology of the pancreatic mass lesion

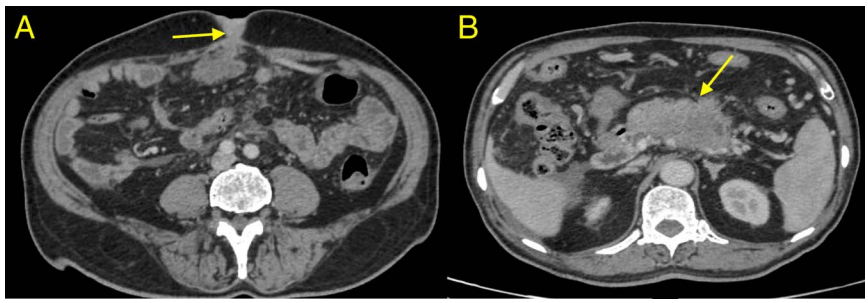

Figure 2. Computed tomography of the abdomen showing (A) Sister Mary Joseph nodule (arrows) and (B) a mass involving the body and tail of the pancreas (arrows).

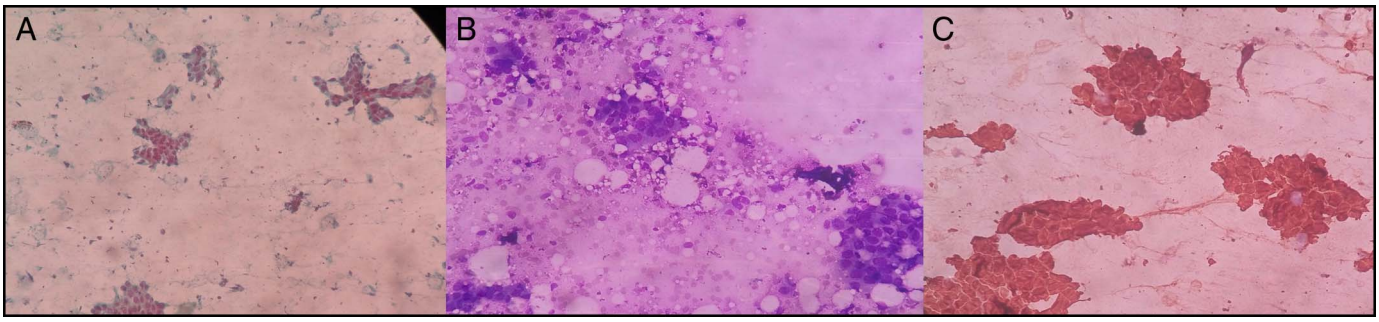

Figure 3. Fine needle aspiration cytology–(A) pap stain, (B) Giemsa stain showing cohesive clusters of large tumor cells with moderate to abundant pale blue cytoplasm, irregular nuclear membrane, large vesicular nucleus with opened up chromatin and few conspicuous nucleoli, and (C) immunocytochemistry of umbilical nodule positive for cytokeratin 19.

suggestive of adenocarcinoma. Imaging modalities such as ultrasound, CT scan, magnetic resonance imaging, or positron emission tomography CT and tumor markers are useful in the detection of the primary lesion.

The presence of a SMJ nodule usually indicates a poor prognosis, with mean survival often less than a year. The outcome is even worse in pancreatic cancer because the average survival is less than 3 months. The recommended treatment is often palliative, requiring chemotherapy, radiotherapy, or both. However, aggressive surgery and adjuvant therapy have been reported to improve survival, especially in patients presenting with SMJ as a solitary metastasis.[1,3]

In conclusion, umbilical metastatic nodule is a useful physical finding and a clue to an underlying advanced gastrointestinal malignancy. Although rare, pancreatic cancer should be considered as one of the primary sites to look out for in patients presenting initially with SMJ nodule.

DISCLOSURES

Author contributions: All authors contributed equally to this manuscript. P. Mohan is the article guarantor.

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