A 64-year-old man presented with a 1-month history of painless nodule on the umbilicus (Fig. 1A). He had no history of abdominal pain, poor appetite, bowel habit change, or body weight loss. The skin biopsy of the umbilical nodule revealed dermal infiltration of tubuloglandular neoplastic cells, which were stained strongly positive for cytokeratin 7 and weakly positive for cytokeratin 20. All laboratory test results were within normal ranges, except a remarkable elevation in the tumor marker CA19-9 level of 12000 U/mL (normal: 37 U/mL). The abdominal computed tomography confirmed a 3.8 cm mass at the pancreatic tail (Fig. 1B, arrow), with pancreatic duct dilatation, multiple foci of liver metastasis, and peritoneal carcinomatosis. Gastrointestinal endoscopy, chest X-ray, and whole-body bone scan showed negative results for tumor surveillance. Under the diagnosis of pancreatic adenocarcinoma with multiple metastases, the patient was transferred to the oncology department for chemotherapy.

Cutaneous metastases from an internal malignancy have been reported in 0.7–10.4% of patients. Sister Mary Joseph nodule (SMJN) is a cutaneous nodule resulting from metastasis of malignant tumors affecting the umbilicus. SMJN is commonly associated with a primary adenocarcinoma of the gastrointestinal or genitourinary tract. Umbilical involvement in an internal malignancy may result from lymphatic or hematogenous spread, direct invasion into the
skin, or iatrogenic implantation by surgical procedures. We reviewed 465 SMJN cases, which were reported by 2012 (table not shown), and found that the most common primary site of malignancy was the stomach (20.65%), followed by the ovary (17.85%), large intestine (12.04%), and pancreas (9.25%). It was worth noting that the primary tumor of unknown origin accounted for 15% of the cases.

Searching for the sites of primary malignant tumors is essential when the diagnosis of SMJN is made. Immunohistochemical markers are useful to define tumor lineage from tissue specimens for unknown primary cancer. Monoclonal antibodies to cytokeratin 7 and cytokeratin 20 have been used to classify tumors because of their distinct expression by different organs. The cytokeratin 7+/20+ phenotype narrowed the differential diagnoses to urothelial tumors, ovarian mucinous adenocarcinoma, cholangiocarcinoma, and pancreatic adenocarcinoma. Based on the results of imaging studies and cytokeratin staining, it is clinically rational to conclude that the primary tumor site of this patient was the pancreas, without performing further biopsy of pancreatic tail or hepatic nodules.

The presence of SMJN often means a poor prognosis. The survival time without treatment ranged from 2 months to 11 months (average 10 months). Only 13.5% of patients with SMJN stayed alive after 2 years of umbilical metastasis, and the longest survival time reported was 18 years.

In summary, we delineated a case with an end-stage pancreatic cancer with SMJN as the first presenting sign. SMJN is a characteristic but easily overlooked dermatological sign of metastatic intra-abdominal tumors. Awareness of such a specific manifestation enables the physicians to investigate the underlying malignancy promptly.

References