CASE REPORT

SISTER MARY JOSEPH NODULE, A FORGOTTEN NODULE

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Metastatic cancer of the umbilicus, known as Sister Mary Joseph nodule, is typically associated with visceral malignancy. It is an uncommon and rare presentation. It indicates disseminated disease and poor prognosis. Physicians need to be aware of this rare clinical condition so that they can promptly diagnose the primary cancer.

Keywords: Sister Mary Joseph, umbilical nodule, gastric adenocarcinoma

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INTRODUCTION

Sister Mary Joseph nodule (SMJN) is an eponym to describe cutaneous metastasis of a visceral malignancy to the umbilicus.[1] Gastrointestinal malignancies account for about half of the underlying sources (gastric, colonic, pancreatic cancer), gynaecologic (ovarian, uterine cancer), unknown primary tumours and rarely bladder or respiratory malignancies cause umbilical metastasis.[2,3]

The proposed mechanisms for the umbilical metastasis include spread via the remnant structures like the falciform ligament, direct trans-peritoneal spread via the lymphatics which run alongside the obliterated umbilical vein or the haematogenous route.[2] It is a rare condition and an uncommon presentation of primary visceral cancer. The presence of umbilical metastasis indicates a grave prognosis and is a sign of advanced malignancy.

CASE REPORT

A 37 year old woman with epigastric pain and umbilical swelling was referred to gastroenterology clinic for further evaluation. She described epigastric pain after intake of solid foods and nonbilious vomitus. In the past several months, she noted a weight loss of 15 kg. On physical examination, she had palpable left supraclavicular lymph node (Virchow's node), umbilical swelling (Sister Mary Joseph nodule; Figure-1, Panel A) and 4cm hepatomegaly. Fine needle aspiration cytology (FNAC) of umbilical swelling indicated adenocarcinoma. CT scan abdomen revealed gastric malignancy with hepatic and vertebral metastasis and a mass through the umbilicus (Figure-1, Panel B). Endoscopy revealed diffuse, circumferential, ulcerated growth involving the body and antrum of stomach (Figure-1, Panel C). Histopathology showed signet ring cell adenocarcinoma of stomach (Figuer-1, Panel D). Patient was referred to oncologist for palliative care.

DISCUSSION

Umbilical tumours are relatively rare. The differential diagnosis includes malignant and benign lesions such as pyogenic granuloma, epidermal cyst, haemangioma, abscess, umbilical hernia, endometriosis, and primary umbilical carcinoma.[2] Malignant tumours can be primary or metastatic tumours.

Historically, Sister Mary Joseph (1856–1939) was a surgical assistant of Dr. William Mayo, a gastrointestinal surgeon at Saint Mary's Hospital in Rochester, Minnesota (at present, the Mayo Clinic), who noticed the presence of an umbilical nodule in intra-abdominal malignancies. In 1928 he reported her findings as pants button umbilicus during a lecture to the Cincinnati Academy of Medicine. Nevertheless, it took 21 years until 1949 when the English surgeon Sir Hamilton Bailey mentioned her observation in the 11th edition of his famous surgical textbook "Demonstrations of Physical Signs in Clinical Surgery" with the new name Sister Mary Joseph nodule.[4]

Sister Mary Joseph nodule can be a presenting symptom or sign of undiagnosed underlying malignancy, or an alarming symptom or sign of disease progression or recurrence in a known patient. Its incidence is 1–3% of all intra-abdominal or pelvic malignancies.[5] The lesion is seen more often in women.[6] Clinically, it usually presents as a firm, indurate often vascular swelling averaging in size from 1 cm to 5 cm, but occasionally enlarges enough to form a protruding tumor.[7]

A SMJN may be fissured or ulcerated and may have serous, mucinous, purulent or bloody discharge. The nodule has been described as white, bluish violet and brownish red and is occasionally pruritic. These metastatic lesions are predominantly from gastrointestinal (52%) and gynaecological neoplasm (28%), most commonly from the stomach (23%), colon (15%), pancreas (10%), and ovary (16%), and less frequently from the uterus, cervix, gallbladder, and small intestine.[8] The mechanism of umbilical seeding from primary tumours is not

clearly understood; however, authors worldwide have proposed several hypotheses. Metastatic lesions can reach the umbilicus via propagation through lymphatic ducts, the venous network, arterial spread, contiguous extension or extension along the ligaments of embryologic origin.[9]

Sister Mary Joseph nodule is an ominous sign of disseminated disease that is not amenable to cure. Mean life expectancy is 2–11 months without treatment.[10] Surgery is usually recommended only in patients with a solitary umbilical metastasis. Surgery should be avoided in cases with widespread disseminated disease; in such cases, effective palliation can be achieved with chemo radiotherapy.[5]

Sister Mary Joseph nodule is an uncommon manifestation of visceral and other malignancies. It is essential for all physicians to be aware that an umbilical nodule may be the first presenting sign of internal malignancy, its progression or recurrence and should prompt further clinical evaluation.

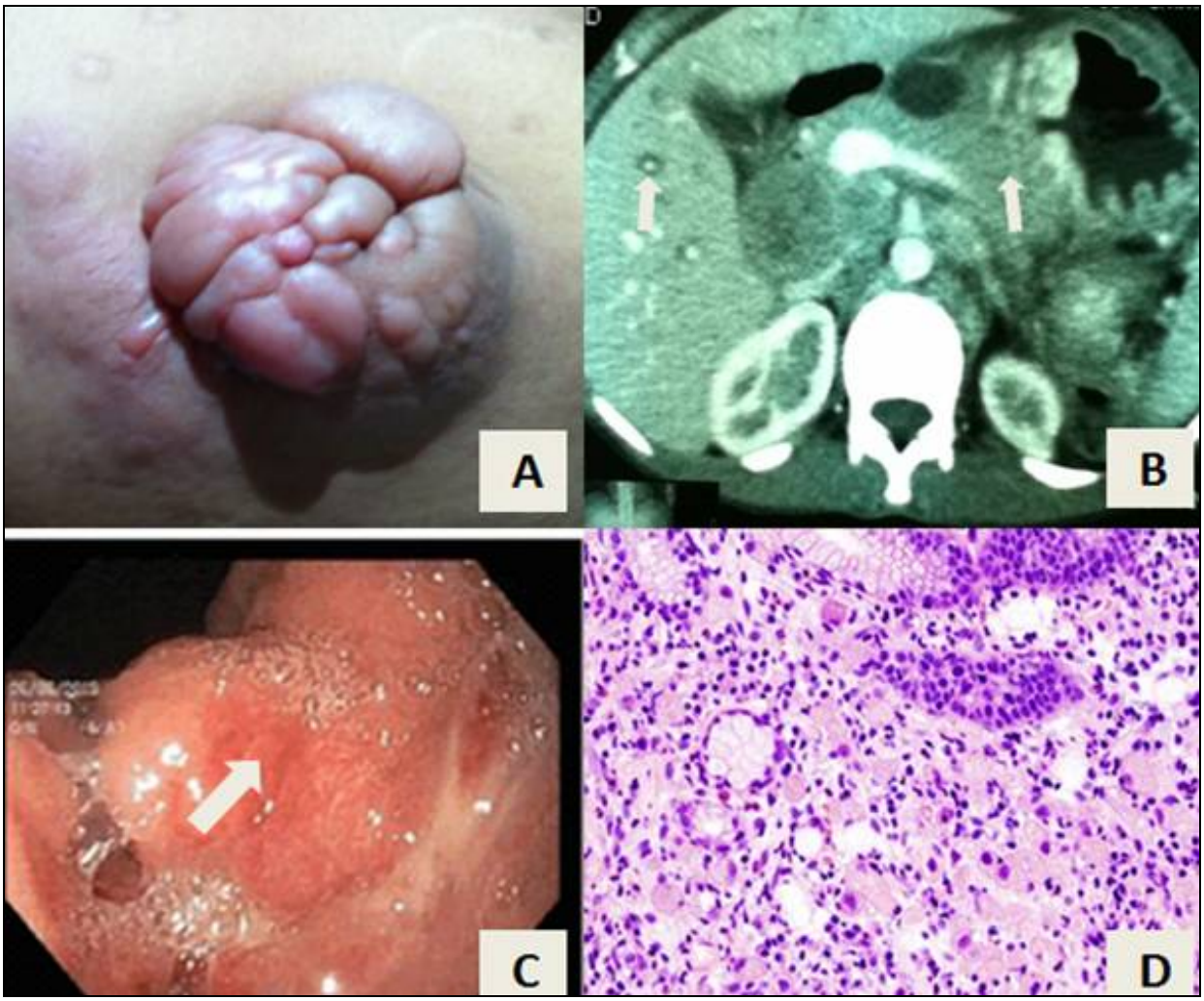

Figure-1: A: Sister Mary Joseph nodule, B: Gastric mass with liver metastasis on CT scan (indicated by white arrows), C: Gastric mass on endoscopy, D: Signet ring cell adenocarcinoma on histopathology

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