



Case report

Sister Mary Joseph's nodule revealing a jejunal adenocarcinoma: A case report

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ABSTRACT

Introduction and importance: Adenocarcinoma of the jejunum is a diagnostic challenge for the physician because of its extreme rareness and the fact that it classically presents with vague clinical symptoms. On the other hand, the Sister Mary Joseph's nodule is a rare clinical sign that refers to umbilical metastasis of an internal malignancy. We here report a rare case of jejunal adenocarcinoma revealed by a Sister Mary Joseph's nodule.

Case presentation: A 55-year-old man presented with an ulcerated umbilical tumor, which was found to be a secondary lesion of an advanced jejunal adenocarcinoma invading the transverse colon. He underwent surgical resection of the umbilical tumor and the intestinal primitive.

Clinical discussion: The presence of umbilical metastasis usually represents advanced disease but can be its first manifestation. Gastro-intestinal tract tumors such as jejunal adenocarcinomas and gynecologic malignancies are the most common primary sites.

Conclusion: Practitioners must be aware of clinical implication of Sister Mary Joseph nodule. Aggressive surgery when feasible can be beneficial for survival.

1. Introduction

Small bowel cancers are rare tumors that represent less than 5% of all gastrointestinal malignancies [1]. Moreover, only 29% of cases are located in the jejunum [1]. Their clinical presentations are nonspecific, which usually leads to delay in diagnosis.

Although rare, the umbilicus can be the site of malignancy. The majority (83%) of malignant umbilical neoplasms are secondary tumors which are much more common than primary malignancies [2]. Secondary umbilical malignancies are known as Sister Mary Joseph's nodule or sign. We here report a case of a man who presented at the National Teaching Hospital of Cotonou (CNHU-HKM) with an umbilical tumor that revealed a jejunal adenocarcinoma.

This work has been reported in line with the SCARE 2020 criteria [3].

2. Case presentation

A 55-year-old black man with no particular medical history

presented at the visceral surgery department of the National Teaching Hospital of Cotonou (CNHU-HKM) in December 2020 for an umbilical mass. He experienced spontaneous umbilical pain ten months earlier, which worsened with contact. A few days later, the patient noticed a nodule on the umbilicus that progressively increased in size until it became ulcerated.

The patient had a good general condition. An umbilical tumor, ulcerated over its entire surface, measuring 6 cm * 6 cm, painful and bleeding on contact, was observed (Fig. 1). On deep palpation of the abdomen, a firm, poorly limited mass (estimated 8 cm) was found in the right hypochondrium.

A biopsy of the umbilical tumor was performed, and the pathology report was consistent with a well-differentiated, infiltrating adenocarcinoma. Abdominal CT scan suspected an irregular, circumferential thickening in the right third of the transverse colon measuring approximately 8.31 cm and confirmed the homogeneously enhanced umbilical tissue growth (Fig. 2). Total colonoscopy did not reveal any endoluminal tumor.

A laparotomy was performed in February 2021, by two senior

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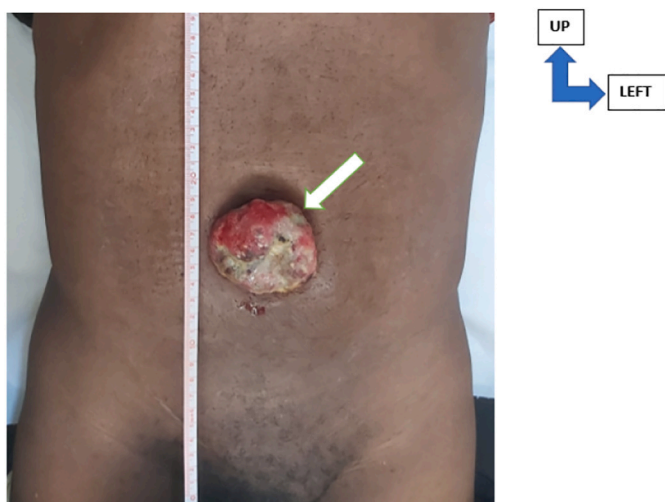


Fig. 1. Sister Mary Joseph's nodule, ulcerated umbilical mass (white arrow).

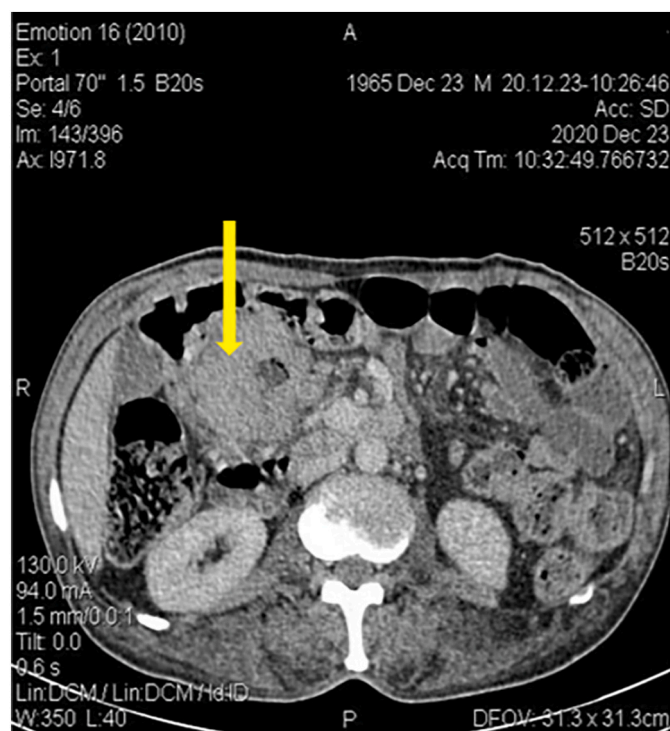


Fig. 2. Computed tomography showing the intra-abdominal tumor (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

digestive surgeons. An omphalectomy was carried out at first instance (Fig. 3A). Exploration revealed a tumor of the second jejunal loop invading the right transverse colon and, omental nodules. There was no peritoneal carcinosis. The surgery, therefore, consisted of a monobloc jejunal resection with a right hemicolectomy and a radical omentectomy. The post-operative course was uneventful.

Pathology report concluded a jejunal lieberkhanian adenocarcinoma (Fig. 4) measuring 6.5 cm, invading the peritoneum and the mesocolon by contiguity. Five nodes were metastatic with capsular invasion out of 48 examined.

Adjuvant FOLFOX (5-fluorouracil, leucovorin, and oxaliplatin) regimen chemotherapy was indicated, but unfortunately could not be performed due to a lack of financial resources. Eight months after

surgery there is no evidence of recurrence. The patient confirmed that he was feeling well.

3. Discussion

Sister Mary Joseph, assistant to Dr. William James Mayo (from 1890 to 1915), noted a relationship in patients with umbilical nodules and the presence of intra-abdominal malignancy. Dr. Mayo published this observation in 1928. The term 'Sister Mary Joseph's nodule' was used for the first time in 1949 by the English surgeon Hamilton Bailey, in acknowledgement of her work [4,5].

Metastatic umbilical tumors present in 20% of cases as the first manifestation of internal undiagnosed malignancy. They may also occur during known malignancy indicating disease recurrence or progression [6,7]. They classically present as a growing nodule that can be associated with erythema, induration, ulceration, or tenderness. Typically, its size is below 5 cm in diameter. Some spectacular cases up to 10 cm have been reported [5,7].

The most common histological variant of secondary umbilical tumors is adenocarcinoma although there are reports of umbilical metastases originating from squamous cell carcinoma, sarcoma, mesothelioma, and melanoma [2]. Differential diagnosis includes primary umbilical malignancy, benign lesions such as keloid scars, endometriosis, granuloma, abscesses, and umbilical hernia [5].

The Sister Mary Joseph nodule is more often associated with gastrointestinal malignancies (35–65%) mainly stomach, colon, and pancreas. Small bowel represents only 2% of primary sites. Since small bowel, especially jejunal, malignancies, are rare in comparison to more common gastrointestinal malignancies such as colon or gastric cancers, umbilical metastases of jejunal tumors are much more rare. Our patient's case is therefore exceptional.

Approximately 12 to 35% of cases are related to gynecological malignancies, primarily ovarian. It has also been reported that nodules may originate, rarely, from hematological malignancies, lung, and breast cancers. Identifying a primary site may be challenging and, in some cases, (15–30%) it remains unknown [5,6,8].

The underlying mechanisms of metastatic spread to the umbilicus are varied. They include direct transperitoneal spread, lymphatics and hematogenous spread, or spread via remnants of embryological structures such as falciform ligament, median umbilical ligament, and remnants of the vitelline duct [9]. The presence of peritoneum invasion of the primary tumor in this case, suggest a direct transperitoneal spread to the umbilicus. The absence of hepatic lesions at diagnostic and at follow up, makes a spread through the portal system less evident.

The presence of umbilical metastasis usually represents advanced disease with an associated poor prognosis. The average survival from diagnosis is 11 months. A combination of aggressive surgery and chemotherapy results in better survival, particularly when the nodule of the umbilicus is a solitary metastasis [2,6].

Diagnosis at an advanced stage and the inaccessibility of systemic therapies such as chemotherapy are obstacles to optimal management of these patients in developing countries [10]. In our case the patient couldn't afford chemotherapy.

4. Conclusion

Sister Mary Joseph's nodule may be the first presenting sign in a patient with an unknown malignant disease. The primary tumor, although very exceptionally, can be a cancer of the small intestine. Although rare, it is important to be aware of the presentation and significance of this key clinical sign. Aggressive surgery can improve the outcome even in advance disease.

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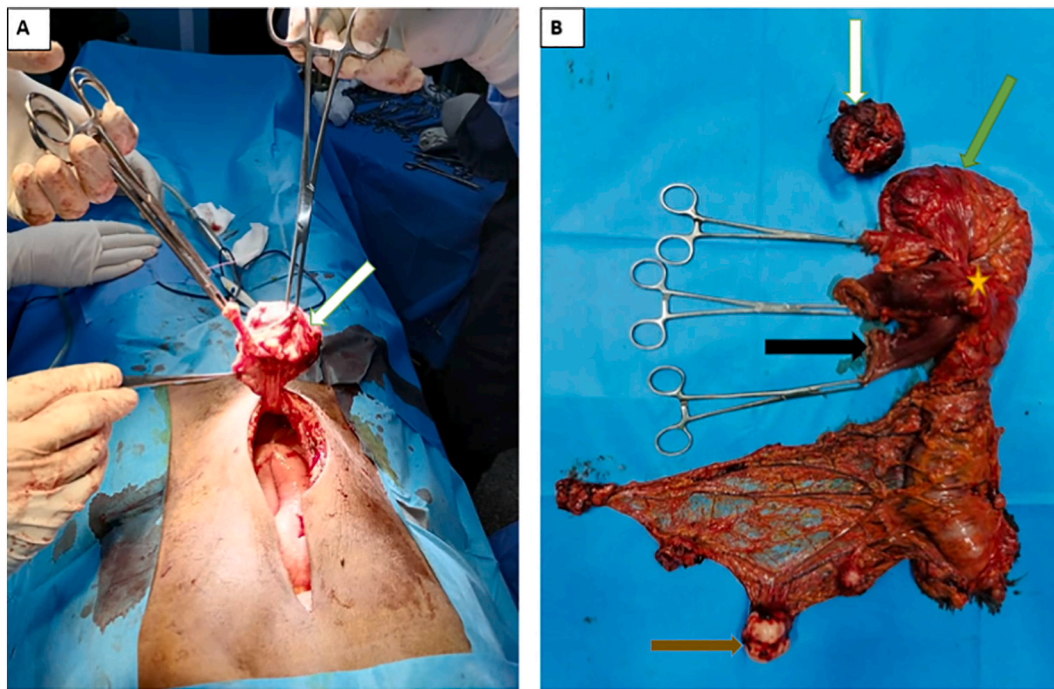


Fig. 3. A. Resection of umbilical tumor (white arrow). B. the resected specimen showing the tumor (yellow star) deriving from the jejunum (black arrow) and invading the transvers colon (Green arrow). Epiploic nodules (brown arrow) were seen. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

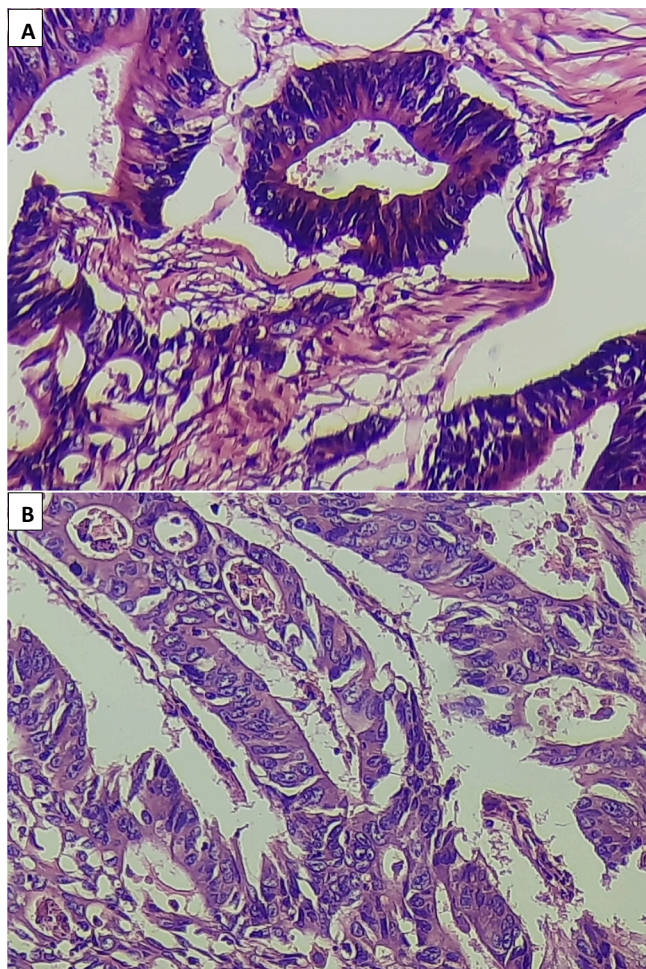


Fig. 4. Microscopic images of the tumor from the pathologic specimen; haematoxylin and eosin staining. A well differentiated jejunal adenocarcinoma ($\times 40$). B. umbilical metastasis ($\times 20$).

agencies in the public, commercial, or not-for-profit sectors.

Consent for publication

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-chief of this journal on request.

Ethical approval

Not applicable.

Guarantor

The Guarantor who is responsible for the present case report is GBESSI DG.

Registration of research studies

Not applicable.
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Analysis and interpretation of data: DG Gbessi and I Lawani.
Drafting of manuscript: FHR Gngangnon and DG Gbessi.
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Declaration of competing interest

The authors declare that they have no known competing financial

interests or personal relationships that could have appeared to influence the work reported in this paper.

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