Case report

Sister Mary Joseph nodule as the only sign of silent caecal adenocarcinoma☆

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A R T I C L E   I N F O

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A B S T R A C T

Sister Mary Joseph nodule is a nodule localized in the umbilicus which represents a cutaneous metastasis from a primary cancer, mostly localized in the abdomen.

We report the case of an 82-year-old woman who presented with an ulcerated umbilical nodule. Imaging identified it as a Sister Mary Joseph nodule, representing the only distant metastasis from a silent caecal adenocarcinoma.

When an umbilical nodule is found at clinical examination imaging can play a crucial role in determining underlying pathology and in determining therapeutic strategy.

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Introduction

Sister Mary Joseph nodule is the name given to an umbilical nodule representing a cutaneous metastasis from a primary cancer. The name is a tribute to a nurse who first noticed that this finding was consistent with advanced stadia of malignancy [1]. The source of malignancy is often an adenocarcinoma from the gastrointestinal tract or from the genitourinary system, even though a hematological, pulmonary or breast source have been described in literature [2].

Case description

An 82-year-old caucasian woman accessed the ER for an umbilical nodule appeared 1 month before, which in the last 3 days showed ulceration of its surface with bleeding and serous-purulent discharge.

At examination the nodule was fibrous in consistency, and painful at palpation. The patient did not declare any changes in her bowel habit, which has always been constipated. She declared fatigue but not weight loss. Fever was not present at the time of examination and her blood tests were normal.

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A CT of the abdomen was prescribed in order to rule out a strangulated umbilical hernia or an umbilical abscess.

CT exam was performed with a 64-row CT Scanner (Optima CT 660, GE Healthcare, Milwaukee, WI, USA) before and after administration of 110 ml of iodinated contrast media (Iopamidol – Iopamiro, Bracco Imaging, Milano, Italy) at a rate of 3ml/sec.

CT demonstrated a soft-tissue density nodule arising from the umbilical scar measuring 18 × 16mm which showed intense enhancement after administration of contrast media (Fig. 1).

The nodule extended from the peritoneal reflection of linea alba to the skin surface; no signs of inflammation were noted in the nearby subcutaneous fat tissue.

The exam also showed enhanced thickening of caecal wall (maximum thickness 6 mm) extending for 25 mm along the length of the viscerum. Three enlarged nodes (maximum diameter 7mm) were also noted in the pericaecal fat (Fig. 2). A diagnosis of caecal adenocarcinoma with suspected nodal metastases was made.

No abnormalities were noted within the liver or in the peritoneum. The exam was then extended to study the chest and the brain; no other significant lesions were noted.

The umbilical nodule was then suspected to be a distant metastasis, a condition known as Sister Mary Joseph nodule.

An excisional biopsy of the umbilical nodule was then performed. At histology it showed clear signs of metastasis from adenocarcinoma.

A colonic endoscopy was also performed, which showed an ulcerated vegetating mass arising from the distal part of the caecum. At histology it was diagnosed as adenocarcinoma of the caecum.

The clinical finding was consistent as Sister Mary Joseph nodule as the only sign of caecal adenocarcinoma.

**Discussion**

Sister Mary Joseph nodules are rare cutaneous metastases from abdominal malignancies, encountered in 2% of all of them [3]. Extra-abdominal malignancies are even more rare.

Differential diagnosis of non-congenital umbilical nodules include non-neoplastic entities as abscess, umbilical hernia, umbilical endometriosis, umbilical scars with keloid formation, and neoplastic entities. In particular, primary neoplastic entities as melanoma, squamous cell carcinoma or basal cell carcinoma account for approximately 20% [1].

Gastrointestinal tumours are responsible for approximately 50% of malignant umbilical nodules and gynaecologic tumours (mainly ovarian, but also uterine) account for approximately 25% of them, for this reason they are more frequently encountered in women [1,4].

Although pathophysiology is unclear, it may involve venous or lymphatic drainage of neoplastic cells to the umbilicum, though direct extension through the peritoneum seems to be the preferred route of spreading for gastrointestinal tumours [2].

Generally, prognosis is poor because a Sister Mary Joseph nodule is a sign of advanced disease; often observed when distant metastases in other organs or extensive peritoneal involvement are present.
In rare cases as ours, when the umbilical nodule is the only distant metastasis, resection of the primary tumour is recommended [5].

In conclusion, when an umbilical nodule is found, imaging is crucial for determining underlying pathologies; in particular it helps in excluding potentially life-threatening pathologies as strangulated umbilical hernia and can diagnose malignancies with Sister Mary Joseph nodule being the only visible sign of it.

Moreover, imaging plays a pivotal role in determining therapeutic strategy, as surgical resection of primary tumour is recommended only when Sister Mary Joseph nodule is the only distant metastasis found.

**Patient consent statement**

Written Informed Consent for publication of this case report was obtained from the patient. The signed form is held by our institution.

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