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Umbilical metastasis mimicking symptomatic hernia: Report of a case of Sister Mary Joseph Syndrome



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ABSTRACT

INTRODUCTION: An umbilical mass can be the first symptom of an intra-abdominal neoplasia, and this condition is also defined as Sister Mary Joseph Syndrome.

CASE PRESENTATION: An eighty-year-old patient presented with a complicated umbilical hernia. CT scan abdomen revealed a pancreatic neoplasm and an umbilical biopsy confirmed diagnosis of metastasis. Patient started chemotherapy by Oncologists.

CONCLUSIONS: In this paper the Authors suggest a histopathological evaluation of umbilical fat in all cases of newly emerging umbilical hernia in the adult population.

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1. Introduction

An umbilical mass can represent an important clinical sign. Indeed, it can be an umbilical metastasis of intra-abdominal neoplasia, also defined as a Sister Mary Joseph Syndrome [1]. It is named from Mary Joseph (1856–1939), a superintendent of St. Mary's Hospital, in Rochester. She noticed a relationship between umbilical nodules, observed during skin preparation for surgical interventions, and intra-operative finding of gastric cancer [2]. We report an interesting case regarding a Sister Mary Joseph's nodule mimicking complicated umbilical hernia.

2. Case report

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

An 80 years old male patient, suffering with hypertension, psoriasis and diffuse arthrosis, was admitted in August 2016 to our Emergency Department because he complained of sudden mesogastric pain. On clinical examination, the patient presented a 2 × 2 cm umbilical non-reducible hernia. An abdominal wall CT SCAN was done: no bowel involvement was detected but, surprisingly, a 4.5 cm cystic-solid mass, localized in pancreatic tail, was revealed (Figs. 1 and 2). CA 19.9 and CEA values were 141.1 U/ml (normal range <35 U/ml) and 4.0 ng/ml (normal range <3 ng/ml) respectively. Further thoracic and abdominal CT SCAN with iodium



Fig. 1. Contrast imaging CT Scan: umbilical fat hernia.

contrast detected multiple pulmonary and liver metastatic lesions. Consequently, we performed an elective hernia repair according to the Mayo technique and biopsy of umbilical fat. Histological finding indicated infiltrating and moderately differentiated adenocarcinoma of umbilical adipose tissue. A diagnosis of umbilical metastasis deriving from intra-abdominal neoplasia, also defined as Sister Mary Joseph Syndrome, was made. Therefore, we planned a CT SCAN – guided pancreatic mass fine needle aspiration (FNA) obtaining a non – neoplastic cytological data (no atypical cells), probably secondary to aspiration in a cystic-necrotic area of neoplasia, with a satisfying mass reduction. Conclusive diagnosis

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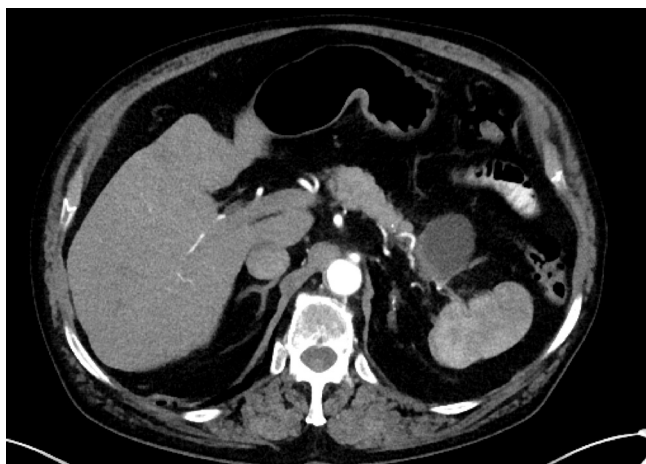


Fig. 2. Contrast imaging CT Scan: 4.5 cm diameter cystic-solid mass localized in pancreatic tail.

was G2 pancreatic cancer (adenocarcinoma) with multiple metastases in liver and umbilical adipose tissue. Both surgical hernia repair and pancreatic mass FNA resulted in no complications during post-operative course. The patient was finally transferred to our Oncology Department to receive adequate chemotherapeutic treatment. First line treatment consisted of gemcitabine from September 2016 to June 2017 plus umbilical radiotherapy according V-MAT technique (December 2016). Considering progression of metastases and increase of oncological markers, i.e. CA 19.9 276 U/ml and CEA 10 ng/ml, oncologists decided to modify chemotherapy by introducing a second line treatment defined FOLFIRI (fluorouracil and oxaliplatin). For this reason we performed a port-a-cath insertion and chemotherapy started on July 2017. At follow-up at 3 months no adverse effects were observed during oncological treatments.

3. Discussion

Sister Mary Joseph Syndrome is a clinical condition characterized by an umbilical nodule. It is rare but it is a really important pathological sign, because it can represent an umbilical metastasis of an intra-abdominal malignancy. These neoplastic nodules are 30% of all umbilical tumours and adenocarcinoma is the most common histological feature (90% of all specimens) [4,5]. Lesions usually arise from gastric (26%), ovarian (12%), colonic (10%) and pancreatic (7%) cancers [6]. A significant part of these syndromes remains of unknown origin (about 29%) [7]. Moreover, it is worthwhile considering that urological and gynaecological cancers can represent other rare causes of umbilical nodules, especially urachal duct carcinomas in male patients and ovarian tumours in females [8,9]. Pathophysiological mechanism can be explained by cancer cells migration through umbilical venous system, lymphatic drainage or round ligament [10]. We report an interesting example of a difficult diagnostic case because initially the nodule seemed to be only a complicated umbilical hernia. Other diseases can be considered among diagnostic hypothesis, such as sarcoma, endometriosis, lymphangioma, lipoma, granuloma [11]. In these challenging cases, the best diagnostic tools to characterize an umbilical mass are Ultrasound (US) SCAN and abdominal CT SCAN [12]. Surgical excision is the only option to obtain a correct histological evaluation.

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4. Conclusion

Patients with Sister Mary Joseph Syndrome are usually affected by an invasive and aggressive cancer and the majority of them have a poor prognosis. We recommend in our paper a histopathological evaluation of umbilical fat in all cases of newly emerging umbilical hernia in the adult population.

Conflict of interests

No conflict of interest.

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Ethical approval

None.

Consent

Written informed consent was obtained from the patient for 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.

Author contribution

Giovanni Cestaro Collection of case report data and design of publication.

Suleyman Alkayyali Grammar check.

Francesco Salmoiraghi References check.

Fabrizio Fasolini Supervisor.

Marco De Monti Supervisor and figures of case.

Guarantor

Giovanni Cestaro.

Marco De Monti.

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