

Case Report

Sister Mary Joseph nodule due to carcinosarcoma

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ABSTRACT

The Sister Mary Joseph Nodule is an eponymous term which describes a palpable umbilical nodule occurring as a result of metastasis of an intra-abdominal or pelvic malignancy. In approximately 50% of cases, this sign is associated with gastrointestinal malignancies. These include gastric, colonic and pancreatic (mainly body and tail) carcinoma. Gastrointestinal malignancy is found in 52% of the metastatic cases and gynecologic and genitourinary origin in 28%, with gastric and ovarian cancers being the most common. The next report is about a 62 year-old woman whose was referred to the surgery department due a painful mass on the abdominal wall that has appeared 3 months before as a fast-growing, with suspicious diagnosis of strangulated umbilical hernia.

Keywords: Sister Mary Joseph, Nodule, Carcinosarcoma, Malignancy, Metastasis

INTRODUCTION

The Sister Mary Joseph nodule or Sister Mary Joseph sign is an eponymous term which describes a palpable umbilical nodule occurring as a result of metastasis of an intra-abdominal or pelvic malignancy.¹ In approximately 50% of cases, the Sister Mary Joseph nodule is associated with gastrointestinal malignancies. These include gastric, colonic and pancreatic (mainly body and tail) carcinoma.² Gastrointestinal malignancy is found in 52% of the metastatic cases and gynecologic and genitourinary origin in 28%, with gastric and ovarian cancers being the most common. Uterine carcinosarcomas are dedifferentiated carcinomas that comprise carcinomatous and sarcomatous elements and arise from a single malignant clone.⁵ Uterine carcinosarcomas are rare tumors that account for less than 5% of all uterine malignancies. Carcinosarcomas occur in older women (age at diagnosis ranging from 62 to 67 years).⁶ Due to the aggressive nature of carcinosarcomas, over 10% of patients will present with metastatic disease at the initial diagnosis, and up to 60% will have extrauterine disease according to imaging.⁷

CASE REPORT

A 62 years old woman was referred to the surgery department due to painful mass on the abdominal wall that has appeared 3 months before as a fast-growing, with suspicious diagnosis of strangulated umbilical hernia. The patient was otherwise asymptomatic. On physical examination, she had a hard erythematous nodule on the umbilical region, measuring 30×25 mm, with central crusting (Figure 1). CT reported: Peritoneal implants with nodular aspect and diffuse distribution mass that commitments the major and minor epiplon, mesentery and the peripancreatic tissues with "omental cake" sign (Figure 2). A 33 mm wall defect is observed in the umbilical ring, which protrudes material with soft tissue density due to secondary peritoneal implants (Figure 3). Exploratory laparotomy revealed frozen abdomen with large and diffuse peritoneal implants. Peritoneal biopsy reported a Metastasis due to carcinosarcoma (Malignant Mullerian Tumor) (Figure 4). Due to distant metastases and extensive local invasion, the tumor was unresectable. She decided to pursue hospice care without any further aggressive measures and she died 3 months after diagnosis due to the tumor progression.



Figure 1: Umbilical nodule. Well circumscribed nodule, hard, rounded umbilical mass measuring 30 × 25 mm diameter lifting the skin with inflammatory signs characterized by central crusting and violaceous patches.

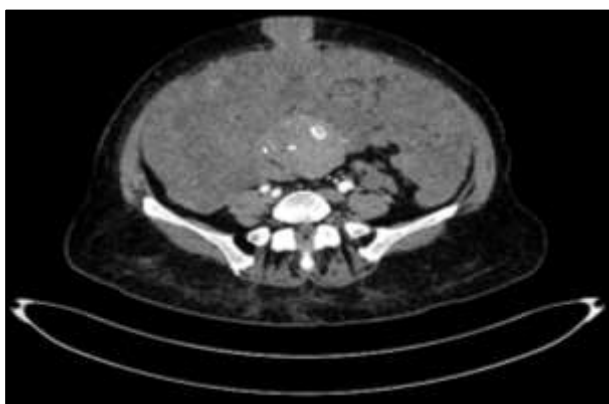


Figure 2: Axial view. Peritoneal implants with nodular aspect and diffuse distribution mass that commitments the major and minor epiploon, mesentery and the peripancreatic tissues with "Omental Cake" sign.

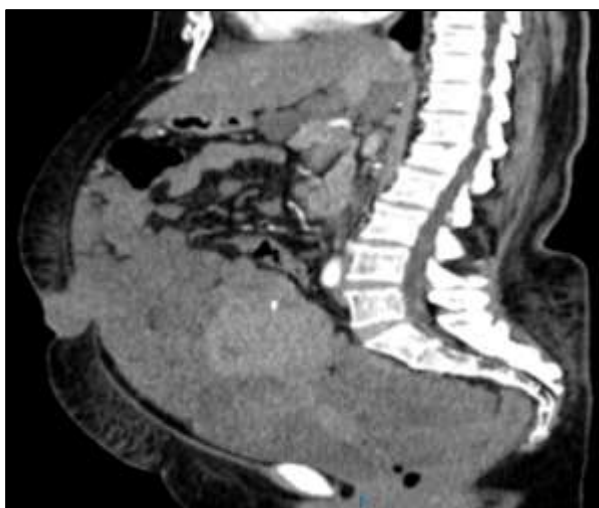


Figure 3: Sagittal view. CT scan showed umbilical nodule with homogeneous enhancement and 33 mm wall defect in the umbilical scar, which protrudes material with soft tissue density associated to peritoneal implants.

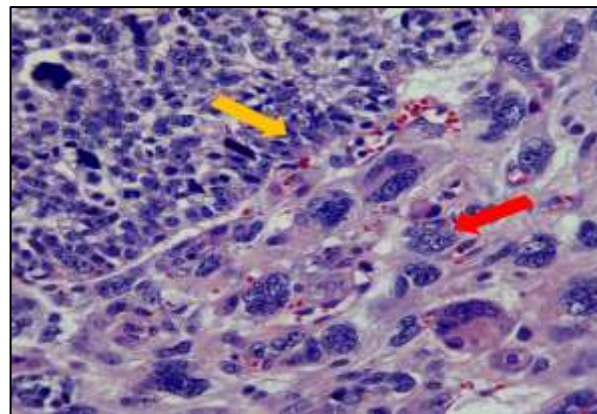


Figure 4: Histology. Biopsy of the mass showed the presence of epithelial tissue component (yellow arrow) and sarcomatoid tissue component (red arrow). Biopsy showed Malignant Mullerian Tumor.

DISCUSSION

The Sister Mary Joseph nodule represents an umbilical metastatic lesion that typically originates from gastrointestinal and gynecologic malignancies. The presence of the Sister Mary Joseph nodule is a sign of widespread abdominal or pelvic malignancy and usually indicates a poor prognosis.⁴ With an incidence of only 5% of uterine malignancies, uterine carcinosarcoma is rare, but it is an aggressive neoplasm.⁸ The presence of an umbilical nodule should raise suspicion of an underlying malignant neoplasm.¹⁰ Surgical resection is the standard management. A combined chemo-radiotherapy can act as an adjuvant treatment.⁹

CONCLUSION

Sister Mary Joseph nodule is a clinical sign that emphasizes the importance of a proper physical examination, especially in elderly patients. It is the first sign of cancer in 0.8%, represents advanced malignancy as in our patient and is associated with a poor prognosis.

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