

Case Report

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A rare case of Sister Mary Joseph nodule as the only sign of unknown primary malignancy

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ABSTRACT

Sister Mary Joseph nodule or Sister Mary Joseph Sign refers to a palpable nodule bulging into the umbilicus as a result of metastasis of a malignant cancer in the pelvis or abdomen. We report a rare case of umbilical mass as the only presenting sign in an old age woman confirmed by CT scan and true cut biopsy to be malignancy. The primary tumor for the umbilical mass or Sister Mary Joseph nodule was not found in this case, although possibility of gynaecological tumor is very high.

Keywords: Umbilical Mass, Sister Mary Joseph nodule

INTRODUCTION

Umbilical tumors are rare and can be classified as benign or malignant.¹ Sister Mary Joseph nodule or Sister Mary Joseph Sign refers to a palpable nodule bulging into the umbilicus as a result of metastasis of a malignant cancer in the pelvis or abdomen. Gastrointestinal malignancies account for about half of the underlying sources (gastric, colonic, pancreatic cancer), gynecologic (ovarian, uterine cancer), unknown primary tumors and rarely bladder or respiratory malignancies cause umbilical metastasis.²

Mechanism of spread of cancer to the umbilicus is unknown but proposed mechanisms include direct transperitoneal spread via lymphatic running along the obliterated umbilical vein, hematogenous spread or via remnant structures like the falciform ligament, median umbilical ligament or a remnant of the umbilical duct.³ Sister Mary Joseph nodule is associated with multiple peritoneal metastases and signifies a poor prognosis.

CASE REPORT

A 60 year old female came with the complaints of umbilical mass since 4 months which was initially of size 2×2 cm and at the time of presentation was of size 8×7 cm. She was referred to our institute as a case of irreducible umbilical hernia. Her bladder and bowel habits were regular. The skin over the swelling was congested and ulcerated at 2 places, the size of the ulcers being 1×1 cm. The patient had no other complaints. The patient was a known case of diabetes mellitus, ischemic heart disease and calcified valve with 25% ejection fraction. Her general examination was completely normal. Abdominal examination revealed hepatomegaly of around 2 cm below entire right costal margin. No fluid thrill or shifting dullness was noted. An umbilical mass of size 8×7 cm with skin excoriation and two small ulcers was noted. The umbilical mass was hard in consistency, immobile and irreducible. Our initial diagnosis was Sister Mary Joseph nodule under evaluation. Complete blood picture was normal, kidney function test and liver function test were also normal. Coagulation profile was normal. CEA, CA 19.9 and AFP were in normal range.

CA 125 was increased up to 18 times the reference range. CT scan was suggestive of hepatomegaly with 3 well defined hypodense lesions involving segments six and seven, enhancing splenic nodular lesion in anterior pole, multiple heterogeneously enhancing soft tissue nodules in the abdominal cavity and anterior abdominal wall in the region of umbilicus, multiple lymph nodes throughout abdominal cavity. FNAC was suggestive of metastatic adenocarcinoma from both umbilical mass and liver. True cut biopsy was suggestive of poorly differentiated epithelial malignancy. A diagnosis of Sister Mary Joseph nodule was made but primary could not be located. The patient was referred to oncologist for further management.



Figure 1: Sister Mary Joseph nodule.



Figure 2: Lateral view of the nodule.



Figure 3: Sister Mary Joseph Nodule front view.

DISCUSSION

Umbilical tumors are rare and can be classified as benign or malignant.¹ Malignant umbilical tumors can be primary or metastatic (secondary) tumors. Metastatic tumors arise from a variety of primary malignant tumors. The term “Sister Mary Joseph nodule” is used to describe a malignant umbilical tumor usually associated with advanced metastasizing intra-abdominal malignancy and generally indicating a poor prognosis. This sign was first identified by Sister Mary Joseph (1856-1939) who as a surgical assistant to Dr. William James Mayo drew attention to the presence of a hard umbilical nodule in a patient being prepared for surgery in 1928.

Sir Hamilton Bailey coined the term “Sister Mary Joseph nodule” in the 11th edition of his textbook “Demonstrations of Physical Signs in Clinical Surgery” in 1949. The nodule usually presents as a firm to hard, indurate often vascular swelling which may be fissured or ulcerated over a smooth surface. It may have serous, mucinous, purulent or bloody discharge. It is usually irregular in shape, generally painless when palpated except if the overlying skin has ulcerated. It is typically less than 5 cm in diameter averaging 1-1.5 cm in diameter but occasionally enlarges up to 10 cm to form a protruding tumor.²

The nodule has been described as white, bluish violet or brownish red and is occasionally pruritic. Patients with Sister Mary Joseph nodule present with a number of clinical symptoms consistent with intra-abdominal cancer including epigastric pain, abdominal distension, weight loss, nausea, ascites and bleeding per rectum.³ The sign of Sister Mary Joseph nodule has been extensively described in literature. Its occurrence is uncommon and as a first sign of malignancy is rare. The evaluation of an umbilical mass should be directed by suspicion of its being a metastatic deposit keeping in mind its potential to be either a primary malignant umbilical lesion or a

benign disease. In a patient with a known malignancy an umbilical mass represents a spread or seeding of the primary tumor and thus can influence therapeutic decision making.⁴ The incidence of Sister Mary Joseph Nodule is very low being only 1-3% of all intra-abdominal and pelvic malignancies.⁵

The most common origins of Sister Mary Joseph nodule are gastrointestinal (52%), gynecologic (28%), stomach (23%) and ovarian (16%) carcinomas. About 15-29% of all cases have an unknown origin (as depicted by the presented case) and 3% originate from the thoracic cavity. Primary tumors in many other sites like gall bladder, uterus, liver, endometrium, small intestine, fallopian tube, appendix, cervix, penis, prostate, urinary bladder, breast, lung and kidneys have also been reported to cause Sister Mary Joseph nodules. Histology of the metastatic umbilical tumor usually reveals adenocarcinoma but rare reports of umbilical metastasis from sarcomas, mesotheliomas and melanomas have also been seen.²⁻⁶

CT Scan (abdomen and chest) and Fine Needle Aspiration Cytology (FNAC) of the tumor are invaluable in the diagnosis of Sister Mary Joseph nodule and help to exclude a primary benign umbilical neoplasm. In 14-33% of cases, umbilical metastases lead to the diagnosis of previously occult neoplasms.⁷ In 40% of patients with a known neoplasm the nodule was an early sign of relapse.⁸ Spread of metastatic carcinoma to the umbilical region has been hypothesized to occur by either 1) contiguous spread of peritoneal cancer, 2) hematogenous spread through arterial and venous systems, 3) lymphatic spread (mainly pancreatic carcinoma) or 4) extension along ligaments of embryonic origin (round ligament of liver, urachus, vitello intestinal duct remnant and the obliterated vitelline artery).^{8,9} The presence of a Sister Mary Joseph nodule verified histological signifies advanced metastatic carcinoma and a poor prognosis with a survival time of 10 months (range 2-17 months) and inoperability. Treatment of established Sister Mary Joseph nodule is palliative as wide excision, surgery and radiotherapy have all proved ineffective. Nevertheless a Sister Mary Joseph nodule is a time tested, honoured clinical sign emphasizing the importance of a careful physical examination of the abdomen. It still remains an interesting and useful diagnostic tool in modern medicine.

CONCLUSION

The diagnosis of any umbilical mass or tumor is not complete without considering the possibility of Sister Mary Joseph nodule. Once proven that the umbilical tumor is malignant or metastatic carries a poor prognosis for the patient and also changes the mode of therapy. Thorough investigations and a high degree of suspicion is a must in any case of umbilical mass or tumor.

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