Case Report

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Sister Mary Joseph Nodule mimics Umbilical Hernia: A report of exceedingly rare case

Volume 14 Issue 1 - 2022

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Received: November 06, 2020 | **Published:** January 31, 2022

Introduction

Ovarian cancer is a very common primary malignancy of 50-70 years postmenopausal women. Metastasis usually takes place through blood and lymphatic system. Skin metastasis in (of) ovarian cancer is rare. The frequency of umbilical metastasis for abdominopelvic cancers is nearly 1% to 3%.[1] Umbilical metastasis in ovarian cancer though not very common carries a high prognostic value. An irregular lump at umbilicus (0.5cm-2cm) due to metastasis from abdominal malignancy is termed as Sister Mary Joseph's nodule (SMJN). SMJN signposts advanced cancer with poor prognosis[2] with median survival as less as one year in ovarian cancer despite all attempts of disease control. The duration of diagnosis of primary cancer to diagnosis of SMJN reveals utmost prognostic importance.[3]

Sometimes SMJN mimics umbilical hernia and the diagnosis is missed by the clinician. This clinical dilemma is more confusing when the patient has undergone an abdominal surgery recently. Thus a missed diagnosis of SMJN may interpret the prognosis of the primary disease wrongly; thereby may lack urgent comprehensive treatment. We report a case of SMJN in a patient suffering from ovarian cancer and has misdiagnosed as umbilical hernia both by clinical examination and in abdominal CT scan or CT scan of abdomen.

Case presentation

A 64 years old post-menoposal woman who was suffering from ovarian cancer and received four cycles of chemotherapy (Paclitaxel and carboplatin), presented to our institution with a complaint of umblical hernia and abdominal swelling. Two months back patient was diagnosed as ovarian cancer and undergone minilaparotomy followed by biopsy which was reported histopathologically as a papillary adenocarcinoma of ovary.

Laboratory investigation showed increased serum CA125 levels of 245 U/L.

During clinical examination; umbilical swelling was moderately hard in palpation and diagnosed as post laparotomy incisional umbilical hernia. On examination, there was a mass of 2 cm at umbilical and another one 1.5 cm at infraumbilical region without any pain, skin ulceration or fistula.

Abdominal CT scan, plain and with contrast shows ovarian mass (10.9x8.7cm2 on right ovary; 8.7x7.0 cm2 on left ovary) with multiple enlarged lymph nodes. CECT whole abdomen was advised and also reported that there is abdominal wall hernia at umbilicus with hernia orifice of 2 cm and bilateral ovarian tumour (Figure 1).

This patient was posted for interval debulking surgery for ovarian tumour under general anaesthesia. During operation it was found that there were two metastatic deposits of 3cmx4cm and 1.5cmx2cm at umbilical and infraumbilical region without any mesenteric herniation (Figure 2). It was excised and debulking surgery was performed. Next plan of treatment was to put the patient for next cycle of chemotherapy.

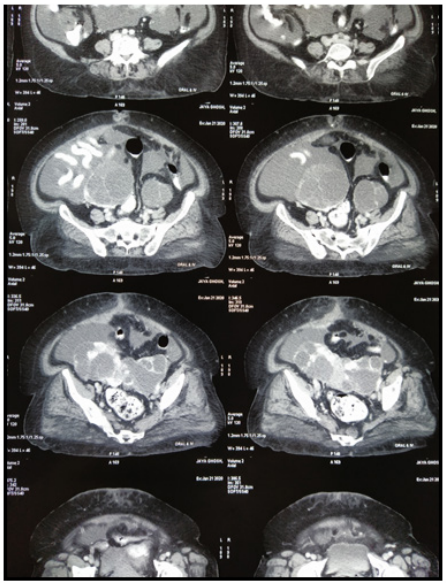

Figure 1 CT scan of SMJN mimics umbilical hernia.

Figure 2 Hard whitish deposits of malignant ovarian cells at umbilicus (SMJN) intraoperative findings.

Figure 3 Sister Mary Joseph.

Discussion

Sister Mary Joseph's nodule was first coined in 1949 by Sir Hamilton Bailey, an English surgeon, for honoring the Sister Mary Joseph (1856-1939) (Figure 3), superintendent nurse who first observed the nodule at the St. Mary's Hospital (now Mayo Clinic) of Rochester, Minnesota.[4]

SMJN is an unusual umbilical cutaneous manifestation of metastatic deposits of abdominopelvic malignancies. SMJN may be the first symptom of an underlying cancer or when recurrence occurs in a patient with a previous visceral cancer.[5-7]

There are so many hypotheses for the spread of tumour cells over the anterior peritoneum and umbilical tissue; among which lymphatic spread through axillary, paraaortic, external iliac, inguinal is the most accepted pathway. However (,) hematogenous spread along the embryological remnants is also evident [8,9] due to rich arterial supply and venous network at umbilicus.

SMJN usually is 0.5 cm to 2 cm in size; however, some nodules may be as large as 10 cm. Umbilical metastasis is indirect diagnosis of advanced neoplastic diseases, with very poor prognosis with 2 years survival of only 15% or less.[6,10] Umbilical lesions are benign only in 47% cases and rests are malignant.[9] Among all malignant umbilical lesions, 88% of cases are metastasis and few are primary skin tumours.[11] SMJN is most common in gastrointestinal malignancy (35-65%). Genitourinary malignancy contributes 12-35%. Primary tumour is only in 3-6% cases.[12] Among the gynaecological malignancies ovarian cancer is the most common primary cancer to metastasize to umbilicus (34% of the cases).[13] Considering the types of ovarian cancer, adenocarcinoma is the most common for SMJN, followed by squamous cell carcinoma, melanoma or sarcoma.[14]

The common age group of diagnosis of SMJN is 50years with range of 18years to 87 years and females are most commonly affected.[15] Umbilical swelling is either benign or malignant. Umbilical hernia, pyogenic abscess, granuloma, mycosis, eczema are common benign diseases. Malignant umbilical lesion is most commonly metastatic rather primary malignancy.[16,17] The diagnosis of SMJN is often misleading due to its variable presentations. It may present with variable range from a hard irregular mass to a soft painful swelling with or without skin erythema, ulceration or fistula.[18,19]

The gross variability in presentation along with variability in timings of diagnosis of primary malignancy results misdiagnosis of SMJN at first clinical examination. This confusion in diagnosis may lead to delay in prompt active intensive treatment of primary malignancy resulting decrease in survival. SMJN is prognostically very important to diagnose accurately at first presentation due to its direct relation to duration of survival. Although FNAC is the diagnostic tool for this lesion; a solid hypoechoic mass with irregularity is the diagnostic radiological feature in ultrasonoghaphy for SMJN.[19] It is more suspicious when there is a history of abdominopelvic malignancy. However; (,) history of recent abdominal surgery may mislead this kind of lesion as umbilical hernia, especially when it is painless and there are no skin changes at umbilicus as in our case. Thus SMJN can wrongly be diagnosed as umbilical hernia both clinically and radiologically resulting negative impact in terms of survival due to wrong assessment of prognosis and lack of imposing all attempts of disease control.

Conclusion

Although unusual, Sister Mary Joseph nodule is an important prognostic indicator of primary abdomino-pelvic malignancies. Physicians must always be aware of this rare clinical presentation so that they can promptly diagnose the primary cancer or its progression or recurrence so as to offer the best treatment with multimodal therapeutic approach. However, SMJN is a thumbprint of disseminated advanced disease with poor prognosis, which requires aggressive combined treatment in each cancer patient without any misdiagnosis.

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Citation: Das D, Mandal A. Sister Mary Joseph Nodule mimics Umbilical Hernia: A report of exceedingly rare case. *J Anesth Crit Care Open Access.* 2022;14(1):24–26. DOI: 10.15406/jaccoa.2022.14.00501

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