

Umbilical Metastasis in Ovarian Cancer: An Erroneous Sign of Advance Malignancy

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Introduction

Umbilical metastasis in ovarian cancer carries ominous clinical importance generally heralding the demise of the patient over relatively short period of time despite all attempts at controlling disease. Sister Mary Joseph's nodule (SMJN) is defined as an irregular lump on the umbilicus, ranging from 0.5 to 2 cm, reaching up to 10 cm as a result of metastasis from visceral malignancies. It can be smooth, non-ulcerated or ulcerated necrotic mass with or without blood, mucinous, serous or purulent discharge [1,2]. Umbilical metastasis from intra-abdominal visceral malignancies is a form of SMJN. The term "Sister Mary Joseph's nodule" was projected by Sir Hamilton Bailey for the umbilical metastasis of an abdominal malignancy in 1948. It can be a presenting symptom or sign of undiagnosed malignancy. Here, we have presented a classical example of SMJN with a diagnosis of umbilical metastasis in ovarian cancer as a primary presentation.

Case Presentation

A 52-years old post-menopausal woman presented with a two month history of swelling over the umbilicus without any other abdominal symptoms. She had history of open tubal ligation 17 years back. On general physical examination, no abnormality found. On systemic examination, an approximately 3 × 3 cm bluish and dark brown swelling seen over the umbilicus with multiple small tiny ulcers over the swelling, without any other visible lump or sinus (Figure 1). On abdominal palpation, there was a hard, tender, umbilical swelling arising from anterior abdominal wall. There was no organomegaly, succussion splash or fluid thrill in the abdomen. The rest of the systemic examination and digital per rectal examination did not reveal any abnormality. Per vaginal examination revealed only post-menopausal mucosal changes. CECT scan abdomen and pelvis revealed a heterogenous mass lesion in anterior abdominal wall with minimal free fluid in peritoneal cavity. There was a heterogenous mass lesion in left ovary of size 5 × 4 × 4 cm. Serum CA-125 level was 174. Tissue biopsy of umbilical lesion was metastatic papillary adenocarcinoma. In view of heterogenous ovarian mass and raised serum CA-125 level she was diagnosed as umbilical metastasis of ovarian origin. Palliative systemic chemotherapy (carboplatin+paclitaxel) was started to assess the response later.

Discussion

On autopsy reports, the incidence of cutaneous metastasis ranges in from 1 to 9%. Among all malignant umbilical lesions,

88% are metastatic lesions, the rest are primary skin tumours [2]. The overall incidence of umbilical metastasis of intra-abdominal malignancies, including gastrointestinal and genitourinary cancers is reported in 1-3% of patients [3]. In a patient with history of known malignancy, umbilical metastasis is a common presentation as recurrence or dissemination. The initial presentation of the

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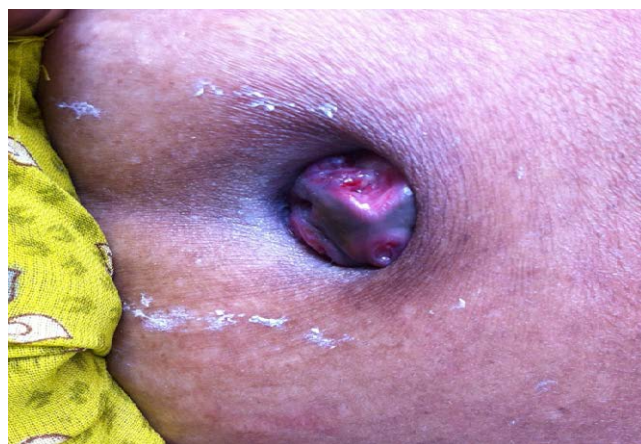


Figure 1 An umbilical nodule of size 3×3 cm with bluish discoloured skin and superficial ulceration

primary malignancy is reported in up to 30% of the cases [2]. The common primary sources of umbilical metastasis are the gastrointestinal (35-65%), and genitourinary (12-35%) tract. Rarely haematological malignancies, lung or breast cancers may be the primary tumour in 3-6% of these cases. After an extensive search for the primary source, 15% to 30% of patients remain with unknown primary lesions [4]. Ovarian cancer is being the most common (34% of the cases) primary cancer to metastasize to umbilicus among all gynaecological cancers [5]. Adenocarcinoma is the most frequent histology in umbilical metastatic lesions followed by squamous cell carcinoma, melanoma or sarcoma [6].

The mechanism of tumor spread to the umbilicus is yet to be vindicated. A perfect understanding of the mechanism whereby the tumor spreads to the umbilicus remains unclear. Numerous postulates have been suggested, including continuous spread of the tumour cells over the surface of anterior peritoneum and invasion to umbilical tissue, which is one of the most common proposed mechanisms. The other possible mechanisms are lymphatic spread via the axillary, inguinal, para-aortic, internal mammary and external iliac lymph nodes; hematogenous spread and spread along with embryological remnants [7,8]. The ultimate deposition of the tumor cells to the umbilicus is favoured by rich arterial supply, an anastomotic venous network which is well connected cranially and caudally with lymphatic draining chains including pelvic and para-aortic lymph nodes. History of recent trauma to the abdominal wall, for example, diagnostic laparoscopy or operation for sterilization are also known to increase the dissemination of tumor cells to the anterior abdominal wall as in present case. The clinical presentation of these lesions is relatively analogous. The appearance of these lesions could be as primary presentation, during or after the diagnosis of the primary tumor. The umbilical nodule might be neoplastic or non-neoplastic lesion. Among non-neoplastic conditions, the differential diagnosis includes paget's disease, angioma, umbilical adenoma (raspberry tumor), umbilical hernia, endometriosis, hypertrophic scar, umbilical granuloma, pilonidal sinus, mycosis psoriasis, and eczema [9]. Fine-needle aspiration biopsy is satisfactory to establish the diagnosis [10].

The presence of umbilical metastasis generally signposts

advanced cancer with widespread metastases, which leads to a poor prognosis [11]. Patients with good performance status, multimodality therapy including surgery and adjuvant therapy can aid in survival. Rarely, the isolated umbilical metastatic is curable with only surgical resection with negative margins with or without reconstruction of abdominal wall defect, but chemotherapy is usually mainstay of the treatment.

Umbilical metastasis usually presents with disseminated disease which is unlikely to get cured. However, these patients should be considered for combination regimens such as carboplatin plus paclitaxel based chemotherapy that gives good response in ovarian cancers. Overall, the umbilical metastases carry a poor prognosis, and median survival is reduced to only about one year [11]. It is documented that the time interval from the diagnosis of ovarian cancer to appearance of umbilical metastasis is an important prognostic factor [12]. However, the umbilical metastasis is as a primary presentation in ovarian cancer is an erroneous sign of advance malignancy.

Conclusion

Sister Mary Joseph nodule might be an unusual indicator of occult visceral and other malignancies. The consciousness of this rare clinical presentation is a must for the physicians so that they can promptly diagnose the primary cancer or its progression or recurrence so as to offer the best treatment with multimodality therapy. However, SMJN is a thumbprint of disseminated advanced disease with poor prognosis, which requires aggressive combined treatment in every individual instance.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author's Contribution

SK, KD and CS initially examined, planned the work up, executed the investigative profile and wrote the manuscript. KD and JG collected the patient's details and reference material. All authors have read and approved the content of final manuscript.

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