

Case Report-IV

Sister Mary Joseph's Nodule

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ABSTRACT

Umbilical nodule, universally known as Sister Mary Joseph's Nodule, mostly is a metastatic manifestation of an underlying advanced malignant disease and is associated with a grave prognosis. Umbilical swelling due to any benign disease is called Pseudo Sister Mary Joseph's nodule. A 41-year-old woman presented with a gradually increasing peri-umbilical nodule. Fine needle aspiration cytology of the nodule proved to be metastatic adenocarcinoma and search for the primary revealed an advanced gastric malignancy. An apparently insignificant umbilical nodule, may be the only manifestation of an underlying advanced malignant disease and therefore clinicians should be aware of such nodule. Histological/cytological examination of such a nodule not only confirms the diagnosis but also helps the clinician to search for the possible primary site.

INTRODUCTION

Umbilical metastasis in the form of a hard nodule or mass which can be seen or felt in the peri-umbilical region is universally referred to as Sister Mary Joseph's Nodule (SMJN). Historically, Sister Mary Joseph (1856 – 1939) was the nursing superintendent and surgical assistant of Dr. William Mayo at St Mary's

Hospital in Rochester, Minnesota. She first noticed the connection between umbilical nodule and intra-abdominal malignancies. In 1949, Sir Hamilton Bailey first used the term 'Sister Mary Joseph's Nodule' to describe this particular type of metastasis in his book 'Demonstration of Physical Signs in Clinical Surgery'.¹

Umbilical nodules are usually malignant and the most common primary site is an abdomino-pelvic tumour. It is estimated that 1% to 3% of the abdomino-pelvic malignancies metastasize to the umbilicus.² In men, the commonest primary site is the gastro-intestinal tract of which stomach is the single most common entity where as gynecological malignancies, particularly epithelial ovarian tumours are the most common primary sites in women. In about 11% cases of Sister Mary Joseph's Nodule, the primary site may never be found.^{3, 4}

Sometimes, umbilical nodules may be benign e.g. endometriosis, fibroma, epithelial inclusion cysts, foreign body granuloma, keloid, myxoma etc and rarely it may be a primary malignant tumour of the umbilicus, like, melanoma, squamous or basal cell carcinoma, sarcoma etc.⁵ Benign umbilical nodules are called Pseudo Sister Mary Joseph's nodule.⁶ Setty⁷ et al described a laparoscopic scar of the umbilicus, which was positive on Positron Emission Tomography but biopsy proved the real diagnosis. Therefore, a histological or cytological study of the umbilical is not only mandatory, but it also guides the clinician to search for the potential primary site.

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We here report a case of Sister Mary Joseph's Nodule in a woman whose primary site was not the ovary but the stomach.

CASE: A 41 year old woman of rural Bengal attended the Radiotherapy department of Bankura Sammilani Medical College with a progressively increasing mass around her umbilicus. She first noticed the mass about one month back and was treated by a local doctor. She had a vague history of dyspepsia and fullness of abdomen of about three months duration. On examination, the mass was peri-umbilical in location, 6 x 4cm in size and reddish in color (Fig- 1).



Fig1: photograph showing umbilical nodule

On palpation, the mass was a conglomeration of nodules with irregular borders, none more than 2 cm in diameter, fixed to the anterior abdominal wall and non tender. Abdominal ultrasonography revealed a larger mass of about 8 cm diameter in the subcutaneous plane, fixed to the anterior abdominal wall, enlarged para-gastric and celiac group of lymph nodes, moderate ascites and mild right sided pleural effusion. Gynecological examination and trans - vaginal ultrasonography showed no adenexal mass. X-ray chest P-A view was unremarkable except obliteration of right costo-phrenic angle. Barium meal examination of the G-I tract revealed two large ulcer craters in the stomach. Upper G-I tract endoscopy showed extensive diffuse growth in the stomach but no obstruction. Both fine needle aspiration cytology of the umbilical growth (Fig - 2) and

endoscopic biopsy of the gastric lesion (Fig- 3) proved to be adenocarcinoma. Routine

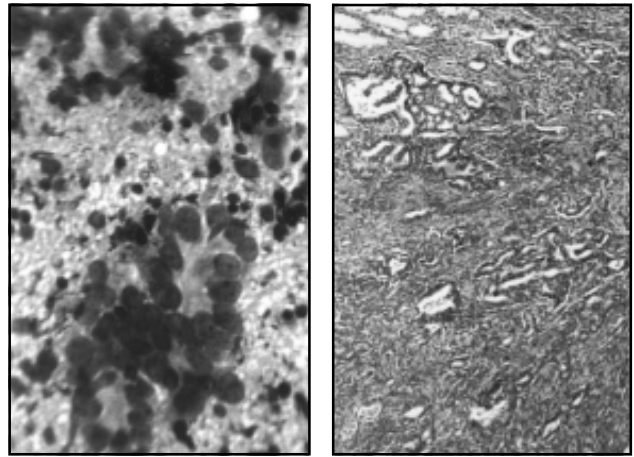


Fig 2: Fine needle aspiration cytology showing adenocarcinoma

Fig 3: Endoscopic guided biopsy from gastric lesion showing adenocarcinoma

hematological examinations, kidney and liver function tests were within normal limits except, moderate anemia (Hb 8.9 gm %), mild jaundice (serum bilirubin: 1.6 gm %) and raised serum alkaline phosphatase level (251 IU / L). Patient received palliative chemotherapy with Inj. Cisplatin - 60 mg / m² IV on day-1, Inj. Doxorubicin - 40 mg / m² IV on day-1 and Inj. 5 FU - 300 mg / m² from day-1 to day-5. The cycle was repeated every five weeks. She gradually improved, peri-umbilical nodule regressed partially, and there was evidence of diminution of size of the retroperitoneal lymph nodes as well as ascitic fluid volume on ultrasonography. Subsequent, she was lost to follow up.

DISCUSSION

Metastasis to umbilicus is very uncommon.⁸ When occur, the primary site is usually gastrointestinal tract in men and gynecological organs in women. Rarely liver, gall bladder, lung, breast, kidney and prostate malignancies may metastasize in the umbilicus.⁴ In most cases, umbilical metastasis accompanies the signs and symptoms of the primary malignancy but rarely it is the only manifestation of the underlying disease. Steck et al⁹ observed that in 45% of their patients, umbilical nodule was the only clinical sign of cancer. Shetty et al⁸ reviewed all

cases of malignant disease of the umbilicus from 1830 to 1989 and found that 42% originated either from the abdomen or from the pelvis and classified the primaries as stomach – 17%, pancreas – 06%, large bowel – 06%, gall bladder – 03% and small bowel – 01%. 9% of primaries originated from the female genital tract. Primary sites like lung, fallopian tube, cervix etc are rare and represented only 01% of his patients. 85 out of 265 published cases were from unknown primary site. However as part of his data was before the advent of modern radiological techniques, number of unknown primaries were a bit high and therefore should be analyzed cautiously. Powel et al³ made an extensive literature search about umbilical nodules and found 32% of the cases were benign neoplasm. Therefore, histological / cytological evaluations of all umbilical lesions are mandatory, not only to determine it's nature but also to guide the clinician to search for the possible primary source, if it is a metastatic one. Though ovary is the commonest primary site in women, other possible sites should not be overlooked, as in this case the umbilical nodule has originated from the stomach and not from the ovary. A case report describing SMJN originating from prostate has been recently published.¹⁰ Sometimes intra abdominal desmoplastic small round cell tumours and intra abdominal neuroendocrine tumours may be associated with umbilical nodules^{11, 12}. In very rare instances, a hard black mass mimicking malignant melanoma but consisting stratified corneocytes may be found in the umbilicus and is called 'omphalolith'.¹³

A variation in vascularity and embryological development makes the umbilicus an easy target of metastasis, particularly from abdomino-pelvic tumours. Several modes of spread have been proposed, but none is conclusive. Metastasis may occur due to proximity of the tumour by contiguity e.g. from peritoneal site. Venous communication between lateral thoracic vein and internal

mammary vein with the portal circulation is another possible route. The retrograde flux from the superficial and deep lymphatic systems originated at para-aortic, inguinal and axillary lymph nodes may lead to umbilical involvement. Malignant cells from the portal circulation may reach the umbilicus through the patent Ductus Venosus particularly in the presence of portal hypertension and the resulting porta-systemic shunting of blood.¹⁴

Fine needle aspiration cytology of the umbilical nodule is very easy due to its favorable location. Edoute et al¹⁵ analyzed the cytological material of 14 patients using fine needle aspiration. Only one case was diagnosed as 'false negative' since 'an inflammatory cell containing aspiration' was obtained. The usual evaluation with Hematoxylin – Eosin may differentiate the primary from metastatic tumours. Moreover, an immuno-histochemical analysis defines the cellular origin in 72% of cases of unknown primary.¹⁶ The use of electron microscopy and cytogenetic analysis enhances the diagnostic sensitivity and specificity.

Sister Mary Joseph's Nodule usually represents an advanced stage of the primary disease and is associated with a grave prognosis. Consequently, palliative management to improve the quality of life is important. In a considerable number of patients, it may be the only presenting sign of a hitherto undiagnosed cancer. Depending upon the site and nature of the primary neoplasm and patients' general condition, combined surgical and medical management may significantly improve survival (from 2.3 to 17.6 months).¹⁷ Long term survival is uncommon.⁴

CONCLUSION

Umbilical metastasis, also known as Sister Mary Joseph's nodule, is a rare phenomena and whose presence usually signifies an advanced malignancy. Clinicians must be aware of this rare manifestation of malignancy which otherwise looks like an innocuous one.

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