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Case report

Fungating sister mary joseph's nodule as an initial manifestation of transverse colon adenocarcinoma

M.A. Tunio[a,*], R. Akasha[b], M. AlAsiri[c], K. Riaz[d]

[a]*Assistant Consultant, Radiation Oncology, Comprehensive Cancer Center, King Fahad Medical City, Riyadh-59046, Saudi Arabia.*

[b]*Resident, Radiation Oncology, Comprehensive Cancer Center, King Fahad Medical City, Riyadh-59046, Saudi Arabia.*

[c]*Consultant, Radiation Oncology, Comprehensive Cancer Center, King Fahad Medical City, Riyadh-59046, Saudi Arabia.*

[d]*Resident, Radiation Oncology, Comprehensive Cancer Center, King Fahad Medical City, Riyadh-59046, Saudi Arabia.*

[*]Corresponding author; Assistant Consultant, Radiation Oncology, Comprehensive Cancer Center, King Fahad Medical City, Riyadh-59046, Saudi Arabia.

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ABSTRACT

Sister Mary Joseph's nodule refers to a palpable umbilical metastasis secondary to underlying abdominopelvic malignancies (gastric, colonic, pancreatic, ovarian and uterine cancer). The clinical appearance varies from small nodule to a large fungating or ulcerated lesion.Proposedhypothesis for spread is through direct trans-peritoneal extension via lymphatic running along the obliterated umbilical vein, hematogenous spread or via median umbilical ligament or a remnant of the umbilical duct. Sister Mary Joseph nodule is associated with dismal prognosis. Here-in we present a case of 76 year old Saudi woman who presented with initial manifestation of Sister Marry Joseph's nodule which subsequently diagnosed as case of underlying metastatic adenocarcinoma of transverse colon and was treated with systemic chemotherapy and radiotherapy to Sister Marry Joseph's nodule.

1. Introduction

Sister Mary Joseph's nodule is referred to rare umbilical metastatic nodule which manifests secondary to underlying primary colonic, pancreatic, gastric, gallbladder, biliary ducts, ovarian and uterine malignancies and lymphomas (Gabriele R et al, 2005). Sister Mary Joseph's nodule as an initial manifestation of colon adenocarcinoma is rarest and only few cases have been reported (Cosentini T et al 2003). The term itself was coined by Sir Hamilton Baily in his surgery textbook "Demonstrations of Physical Signs in Clinical Surgery" in 1949after the name of a long serving head surgical nurse Sister Mary Joseph who pointed out an umbilical nodule of a patient during preoperative care to Dr William James Mayo in 1928 (Tan ML and Padhy AK, 2011).

Here-in we present a case of 76 year old Saudi woman who presented with initial manifestation of Sister Mary Joseph's nodule which subsequently diagnosed as case of underlying metastatic adenocarcinoma of transverse colon and was treated withsystemic chemotherapy and radiotherapy to Sister Mary Joseph's nodule.

2. Case presentation

A 76 year sold Saudi woman presented umbilical fungating lesion and abdominal pain. She had noticed this lesion for 6 months and it was initially within the umbilicus; since then it had been rapidly increasingin size over a month causing discomfort and abdominal pain. Medical history revealed bronchial asthma for which was taking fluticasone 250 micrograms (mcg) inhaler two puffs twice daily and salbutamol 100mcg inhaler one puff 6 hourly.

On physical examination, she was wheel-chair bound with Eastern Cooperative Oncology Group (ECOG) performance scale-3 and was mildly icteric. Per abdominal examination, a hard fungating lesion in the umbilicus and peri-umbilical area of size 6 x 4 cm was seen along with an underlying palpable mass (Figue 1). The rest of examination was found normal.

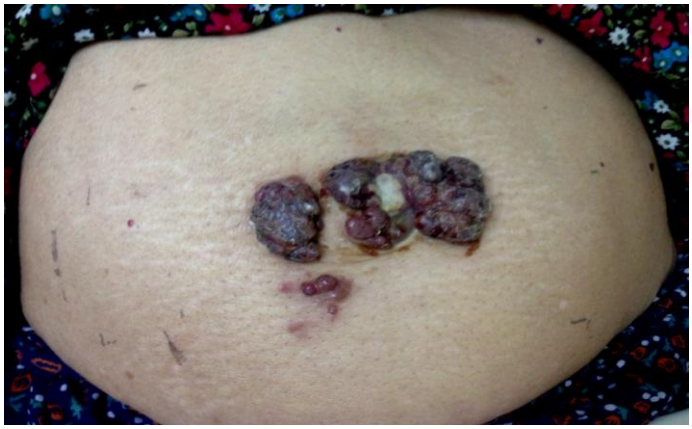

Fig. 1. Per abdominal examination showing a hard fungating lesion in the umbilicus and peri-umbilical area of size 6 x 4 cm.

Hematological tests were within normal limits. Serum LDH was 308 U/L ↑, fasting blood glucose 7.6 mmol/L↑, alkaline phosphatase 170 U/L ↑ and Gamma Glutamyl Transferase (GGT) 69 U/L↑. The rest of hepatic and renal function tests and serum biochemistry was within normal limits. Baseline Carcinoembryonic antigen (CEA) level was 186 mcg/L. Serum CA125 and CA19.9 were not raised. Differential diagnosis was underlying colonic cancer, gastric and primary peritoneal cancer.

Computed tomography (CT) scan of her abdomen was performed which revealed a mass of 4.9 x 5.2 cm size within umbilicus and adjacent rectus abdominis muscle (Fig. 2A). Also there was an 8 cm long asymmetrical mass in transverse colon (Fig. 2B). Multiple hepatic metastatic lesions were also seen in segments II, IVA and VIII (Fig. 2C). The bone scan andCT chest revealed no other distant lesions. Colonoscopy revealed a fungating, partially obstructing 8 cm mass in the transverse colon and biopsies were taken which confirmed the diagnosis of moderately differentiated adenocarcinoma.

After discussing her case in the multi-disciplinary board meeting, she was staged as T3N0M1 and was started on XELOX (capecitabine + Oxaliplatin). After one cycle of chemotherapy, she was referred to radiation oncology

department for palliative radiotherapy to umbilical lesion for symptom relief. Total 30 Gy in 10 fractions were given using RapidArc IMRT (Fig. 3). The course of treatment was tolerated without any radiation side effects. At the time of publication, she was alive and was receiving chemotherapy.

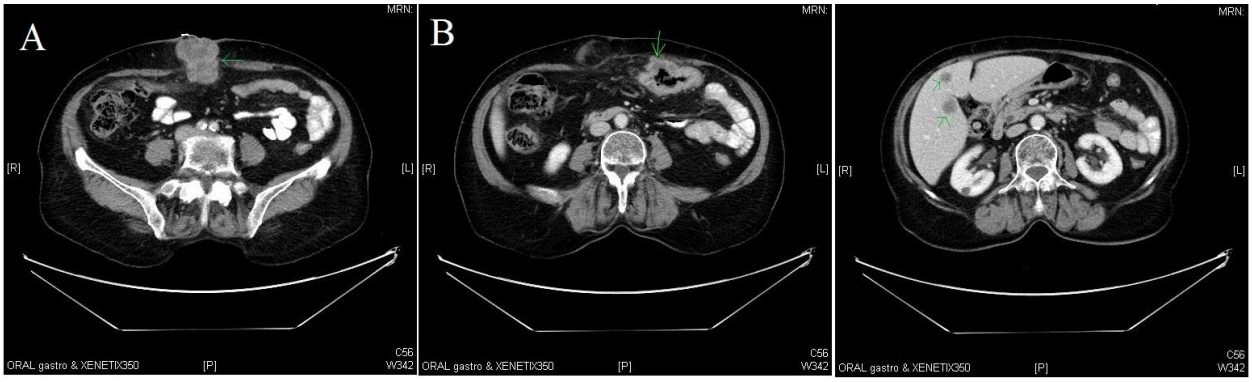

Fig. 2. Computed tomography (axial images) of abdomen showing (A) a mass of 4.9 x 5.2 cm size within umbilicus and adjacent; rectus abdominis muscle, (B) asymmetrical mass in transverse colon of size 8 cm and (C) hepatic metastasis in II, IV and VIII segm.

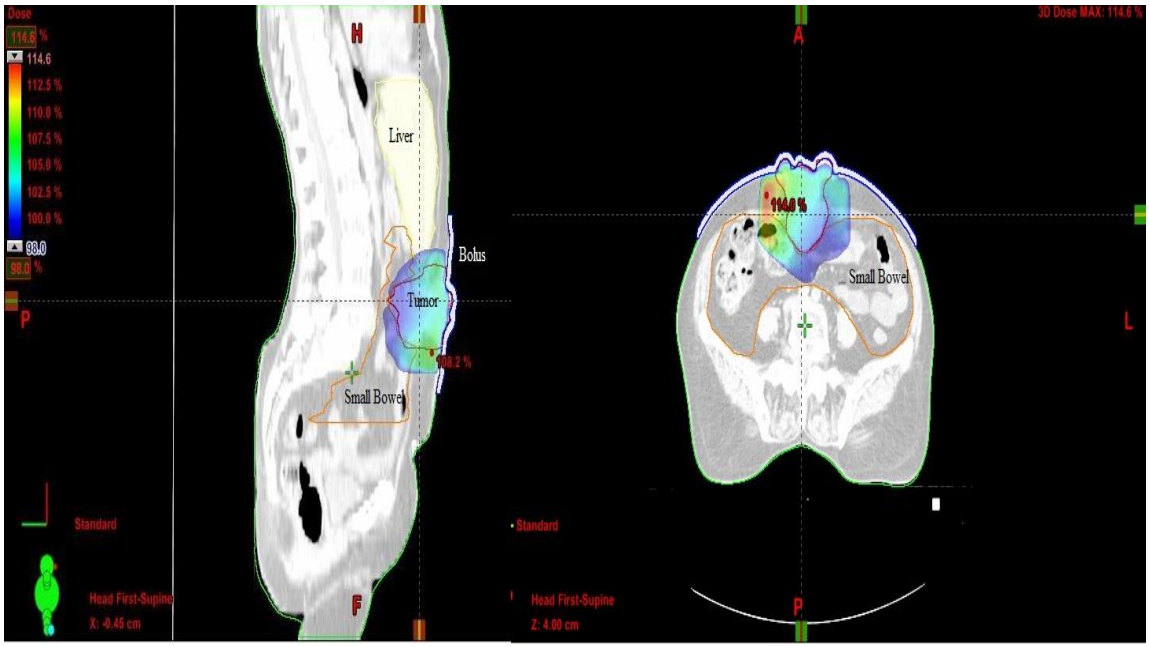

Fig. 3. RapidArc intensity modulated radiotherapy (IMRT) showing better tumor coverage and minimal dose to small bowel.

3. Discussion

The occurrence of Sister Mary Joseph's nodule (umbilical metastasis) is a very rare manifestation and accounts for 1-2% as an initial manifestation of colon adenocarcinoma (Nakamura M et al 2009). The clinical appearance varies from small nodule to a large fungating or ulcerated lesion as seen in our case and female gender has greater preponderance (Ioannidis O et al). The possible routes through which the primary colon adenocarcinoma spread to the umbilicus could be through direct peritoneal extension along ligaments (urachus, vitello-intestinal duct remnant and the obliterated vitelline artery), hematogenous and lymphatic (Papalas JA et al, 2011).

Diagnosis is primarily clinical; however for very small umbilical nodules CT and positron emission tomography (PET) are useful diagnostic tools (Liu Y, 2011). Appearance of Sister Mary Joseph's nodule is well known to be usually associated with a grave prognosis with median survival of 10 months (2-13); however recent advances in chemotherapy may further enhance the survival rates (Kar JK and Kar M, 2012). Other modalities (surgery and radiotherapy) are usually meant for palliation and are used to relieve obstructive symptoms and pain relief, as radiotherapy was used in our patient.

Interestingly, role of radiotherapy for Sister Mary Joseph's is scanty mainly because of proximity of tumor to the small bowel thus high risk of bowel toxicity especially in the conventional radiotherapy era, however in last two decades, novel radiotherapy techniques like intensity modulated radiation therapy (IMRT) has resolved these concerns and has shown that abdominal lesions can be treated with high median doses (55-64 Gy) with 70-80% symptoms relief in the absence of major acute and late sequelae (Dubreuil A et al 1998; Otto K 2003), as our patient received 30 Gy without any bowel toxicity. We believe that, IMRT can be feasible option for treating fungating ulcerated Sister Mary Joseph's nodule.

In conclusion, Sister Mary Joseph's nodule is rare initial manifestation on colonic cancer with varying size and is associated with dismal prognosis. However novel radiotherapy techniques can be used for ulcerated and fungating nodules for symptoms relief.

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