Metachronous small bowel metastasis from a mixed Müllerian mesodermal tumour

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ABSTRACT

A mixed Müllerian mesodermal tumour (MMMT) is a rare aggressive carcinosarcoma. Metastatic progression is uncommon, and occurs via haematological, lymphatic and intraperitoneal spread. Although the latter is seen most frequently, the small intestine seems to be relatively preserved from disease progression with only one reported case of synchronous involvement. We report a case of metachronous MMMT involvement of the small bowel presenting with subacute obstruction that was successfully resected at operation.

KEYWORDS

Mixed Müllerian tumour – Mesodermal – Malignant – Carcinosarcoma – Small bowel metastasis

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A mixed Müllerian mesodermal tumour (MMMT) is a rare aggressive carcinosarcoma that accounts for 2–5% of all uterine malignancies.¹ Metastatic progression is uncommon, and occurs via haematological, lymphatic and intraperitoneal spread.² Although the latter is seen most frequently, the small intestine seems to be relatively preserved from disease progression with only one reported case of synchronous involvement.⁵ We report a case of metachronous MMMT involvement of the small bowel presenting with subacute obstruction that was successfully resected at operation.

Case history

A 71-year-old woman (para 3) presented with a 2-week history of nausea, bilious vomiting, anorexia and abdominal distension but no constipation. She had undergone a total abdominal hysterectomy (TAH) and bilateral salpingo-oophorectomy (BSO) two years previously for a stage Ib uterine MMMT. She had completed four cycles of adjuvant chemotherapy in addition to pelvic radiotherapy (45Gy in 25 fractions) and intravaginal brachytherapy. She had a metastatic lung lesion resected with video assisted thoracic surgery 12 months prior to admission and was under surveillance for a right adrenal metastasis. Her past medical history also included breast cancer (wide local excision and lymph node clearance 12 years previously, with adjuvant chemoradiotherapy and anastrozole), atrial fibrillation (receiving warfarin), hypertension, hypothyroidism, type 2 diabetes mellitus and bilateral lower limb idiopathic lymphoedema.

The patient was being investigated for a six-week history of iron deficiency anaemia; both oesophagogastrroduodenoscopy and colonoscopy were unremarkable but subsequent computed tomography (CT) had revealed a proximal jejunal tumour measuring 8cm × 6cm although there were no signs of intestinal obstruction or other metastatic disease. At the oncogynaecological multidisciplinary team meeting, it was felt that the lesion represented a new primary malignancy and the patient was awaiting diagnostic laparoscopy for further assessment prior to her presentation.

On examination, the patient was obese (body mass index 35kg/m²) and dehydrated with a mildly tender, distended abdomen. Urgent CT noted enlargement of the left-sided proximal small bowel mass (10.5cm × 8.5cm) and detected a new jejunal lesion with associated lymphadenopathy as well as signs of subacute obstruction (Figs 1 and 2).

Given the progression of the disease and the threatened intestinal obstruction, the patient underwent operative intervention. At laparoscopy, two jejunal lesions were identified with invasion of the transverse colon; these lesions were removed via *en bloc* open resection of the jejunum and a concurrent extended right hemicolectomy with continuity restored by primary anastomoses. Histological assessment of the specimen revealed a metastatic carcinosarcoma (MMMT) with infiltration of all layers of the small bowel serosa and mucosa into the adherent transverse colon. There was no evidence of residual disease. Although further chemotherapy was not considered owing to the patient's co-morbidities, she remains well ten months later with no progression of the disease on repeat imaging.

Figure 1 Transverse computed tomography showing jejunal metastatic disease deposit

Discussion

MMMT is a rare aggressive tumour with an incidence of 2 in 100,000.⁴ These tumours occur in postmenopausal women, usually present at an advanced stage and account for 3–5% of all uterine malignancies.² Tumours arise from the Müllerian system (the precursor of the female genital tract) and are biphasic in nature, comprising both epithelial (carcinoma) and mesenchymal (sarcoma) elements; which component accounts for its aggressive behaviour is uncertain.⁵ Most tumours develop in the uterus although they can form throughout the genital tract as well as extra-genitally (including the peritoneum, retroperitoneum, colon, intestinal mesentery and spleen).^{6–8}

Risk factors for MMT include radiation, excessive oestrogen exposure, obesity and nulliparity.^{9,10} The only risk factor in our case was obesity although the patient's history of previous breast cancer may implicate oestrogen exposure. The two-year survival rate for stage I disease is 53%, which decreases to 8.5% in stages II and III.¹

Tumour metastases are uncommon and occur most frequently via transcoelomic intraperitoneal spread but haematological and lymphatic spread has also been reported.^{11,12} Tumour markers can be used to assist in diagnosis and monitor disease progression with treatment.⁵ Radiology provides the mainstay of diagnosis although CT and magnetic resonance imaging results may not be characteristic for MMT metastasis.¹¹ Additionally, the judicious use of small bowel meals, enteroclysis or enteroscopy (push, double balloon or capsule) could further characterise the nature of any small bowel lesions.¹³

Isolated small bowel metastatic involvement is rare, with only one previous case report published in the literature; this paper stated that a concurrent small bowel wedge resection was performed with a TAH and BSO.³ No cases of metachronous small bowel metastases have been reported. The reason for the relative preservation of the small bowel from metastatic spread is not fully understood but may reflect the overall resilience of the small intestine to tumour development.¹³ Our experience and the limited

Figure 2 Coronal computed tomography showing jejunal metastatic deposit with adherent transverse colon

literature would advocate resection of isolated intestinal metastases as this seems to confer a decrease in tumour burden and an increase in disease free survival.³ Antioestrogen therapy can be considered for patients in whom operative resection is not feasible.¹⁰

Conclusions

Our patient developed a metachronous small bowel metastasis from MMT two years following curative TAH and BSO resection. Preoperative diagnosis based on radiological imaging was uncertain and was only confirmed on pathological assessment. Resection of the metastatic disease seems to be curative, with no evidence of disease recurrence or further intestinal obstruction. We would advocate consideration of metastatic disease in any patient with a previous history of MMT and recommend operative resection.

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