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An unexpected diagnosis of primary omental endometrial stromal sarcoma in a patient with acute right abdominal pain: A case report and review of literature

Vered Buchholz*,[1], George Kiroff, Markus Trochsler, Harsh Kanhere

Upper Gastorintestinal and Hepatobiliary Unit, Department of Surgery, The Queen Elizabeth Hospital, Adelaide, Australia

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ABSTRACT

INTRODUCTION: Extrauterine Endometrial Stromal Sarcoma (EESS) is an extremely rare mesenchymal tumour that simulates other pathologies, and therefore poses a diagnostic challenge. This report outlines a case of EESS arising from the greater omentum mimicking a colonic tumour, with review of literature.
PRESENTATION OF CASE: A 47-year-old woman, with history of hysterectomy for menorrhagia and hormone replacement therapy (HRT), presented with right sided abdominal pain and localized peritonism. On exploratory laparoscopy an omental tumour, suspected to arise from the transverse colon was identified and biopsied. The histological features suggested an EESS. Colonoscopy ruled out colonic lesion. A laparoscopic tumour resection and bilateral salpingo-oophorectomy (BSO) was performed. Immunohistochemistry confirmed the diagnosis. No additional lesions or associated endometriosis were found. Resection was followed by adjuvant medroxyprogesterone-acetate therapy.
DISCUSSION: We reviewed 20 cases of EESS originating from extragenital abdominopelvic organs reported since 1990. Acute presentation is rare, as well as upper abdominal occurrence. Isolated omental involvement was previously reported in only one case. Endometriosis is a risk factor for development of EESS and history and/or histological evidence for endometriosis is usually present. HRT is another acknowledged risk factor, mostly on the background of endometriosis. To our knowledge, this is the only report of EESS occurring in a woman on HRT treatment without background of endometriosis.
CONCLUSION: EESS can occur without endometriosis and HRT may be an aetiological factor. The condition can mimic a chronic or acute abdominal pathology and laparoscopic core biopsy is the best way to achieve a diagnosis and formulate management.

1. Introduction

Endometrial stromal sarcoma (ESS) is a rare mesenchymal tumour of the uterus. It may also arise as a primary extrauterine endometrial sarcoma (EESS), mainly on the background of endometriosis, with predominant gonadal involvement as a result. Very rarely does EESS arise primarily in the gastrointestinal or extra-gastrointestinal tract organs outside the pelvis, or evolve without preceding endometriosis [1]. In these cases, the non-specific presentation and the unexpected location pose a diagnostic challenge [2,3], and often another abdominal pathology is first suspected. A case of primary EESS arising from omentum mimicking a colonic primary is presented, with review of the literature. This report is line with the SCARE criteria [4].

2. Presentation of case

A 47-year-old female, presented to the Emergency Department with right sided and central abdominal pain. Her medical history included hysterectomy 10 years earlier for menorrhagia, and HRT with oestradiol patch. She reported vague, generalized abdominal pain, mainly central, which later migrated to the right lower abdomen. On examination she was tender along the right and upper central abdomen with localized peritonism. WBC and CRP were elevated (12.1×10^9/L, 16 mg/L respectively). Sonography demonstrated a hypoechoic, ovoid, irregular lesion with blind end, and sonographic probe induced tenderness above it. Given the acute presentation, and the high suspicion for an inflammatory lesion,

* Corresponding author at: Upper Gastrointestinal and Hepatobiliary Unit, Department of Surgery, The Queen Elizabeth Hospital, 28 Woodville Road, Woodville South, SA 5011, Australia.
E-mail addresses: Vered.Buchholz@sa.gov.au, veredbuch@gmail.com (V. Buchholz).
[1] Present address: Upper Gastrointestinal and Hepatobiliary Unit, Department of Surgery, The Lyell McEwin Hospital, Haydown Rd, Elizabeth Vale, SA 5112, Australia.

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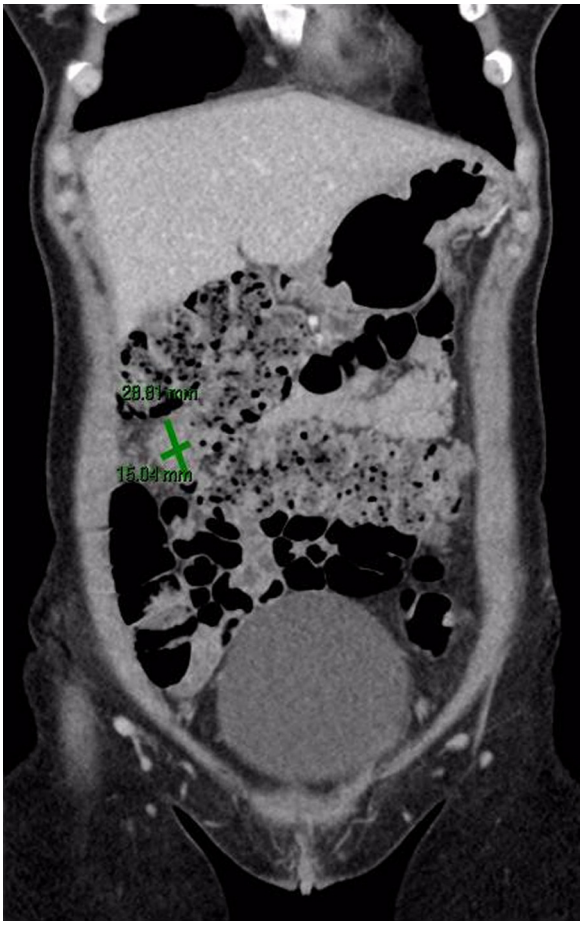

Fig. 1. (Greyscale and colour). Contrast enhancement coronal computed tomography showing showing ovoid mass in proximity to transverse colon.

the patient was taken for an exploratory laparoscopy. At surgery, a firm irregular omental mass was found, located above the proximal transverse colon, near the hepatic flexure. The lesion seemed to extend from the colon. The Appendix was normal. Prior hysterectomy was noted, right ovary and tube were normal, however optimal view of the left adnexa was impossible due to pelvic small bowel adhesions. No other pathology was found. A partially necrotic omental nodule adjacent to the larger eccentric mass was biopsied. The specimen consisted of an 18 × 15 × 12 mm nodule, with extensive haemorrhagic necrosis. The residual tissue was comprised of monomorphic ovoid to spindle-shaped cells with high nuclear to cytoplasmic ratio. The morphology suggested a mesenchymal-type lesion, but immunohistochemistry with a panel of antibodies, raised the possibility of endometrial stromal sarcoma.

Given the rarity of this diagnosis without previous ESS and endometriosis, the past hysterectomy slides were reviewed and confirmed as benign without any evidence of ESS. The current and previous slides were sent for a second external review which supported the diagnosis of primary EESS. A systemic workup was then undertaken. Chest, abdomen and pelvis computed tomography demonstrated a 28 × 15 mm ovoid mass arising from the transverse colon, without evidence for distant metastasis or nodal disease (Fig. 1). Tumour markers (CA19.9, CA-125, CEA, AFP) were negative. Colonoscopy excluded primary or secondary colon involvement. The patient was discussed by the Surgical and Gynae-Oncology multidisciplinary teams. The decision was to proceed with laparoscopic excision of the tumour together with BSO by joint surgical and gynaecology teams. A two centimetre omental lesion was found next to the hepatic flexure and resected. Pelvic small bowel loops were adhesiolysed and normal looking ovaries and tubes were resected. No peritoneal spread or other metastatic lesions were identified. The post-operative recovery was uneventful and the patient was discharged on day one post surgery.

The omental lesion contained a firm nodule measuring 21 mm in maximal dimension. The mass was characterized by multiple foci of ovoid to spindle–shaped cells forming irregular tongues dissecting through markedly fibroblastic stroma (Fig. 2a). The cells were uniform with rare mitotic figures. Small capillary-sized blood vessels were found within the proliferative cells (Fig. 2b). The tumour cells stained strongly for CD10 (Fig. 2c), oestrogen receptor, progesterone receptor, vimentin, WT-1 and Bcl-2. There was no reaction for c-Kit, CD34, desmin, or smooth muscle actin. Both ovaries and fallopian tubes were normal. The morphology and immunoprofile were compatible with the diagnosis of primary extrauterine low grade ESS originating from the omentum. HRT was ceased and the patient was put on adjuvant hormonal therapy with medroxyprogesterone acetate and was scheduled for regular follow-up.

3. Discussion

Extrauterine endometrial sarcoma involving an extra-genital site is an extremely rare condition. It is even more rare to find it in the upper abdomen and without any clinical evidence of endometriosis. Even fewer cases of upper abdominal ESS without clinical evidence of endometriosis have been described in the literature. All 20 reported cases (including the present case) reported since 1990 were reviewed. These are summarized in Table 1.

Patient ages ranges between 38–80 years (median, 47.5 years). More than half of the patients were in their fifth decade or early menopause years (11/20) as previously reported [3]. The clinical presentation was ambiguous in most instances. The commonest complaint was abdominal pain (9 cases). Other complaints included

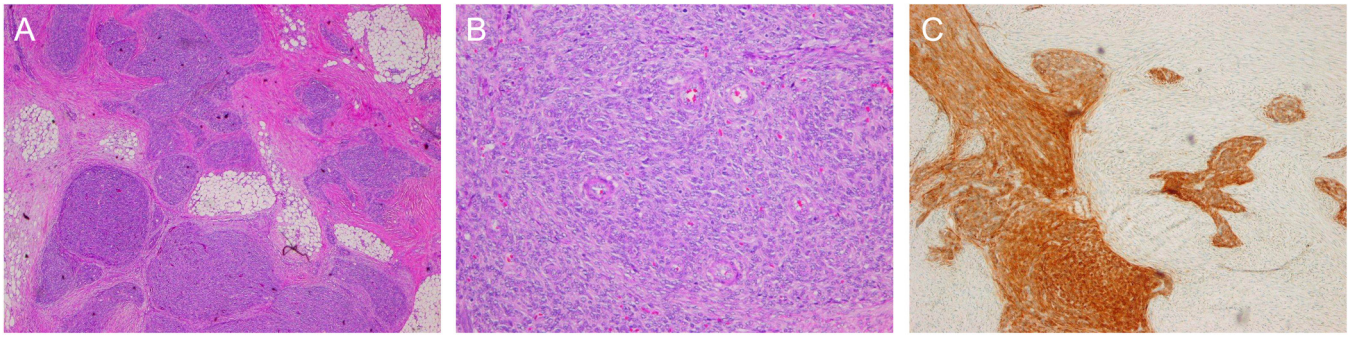

Fig. 2. (Colour image). (A) Islands of spindle-shaped tumour cells infiltrating into omental fat. (B) Uniform population of spindle small with embedded prominent capillary-sized vessels/or arterioles. (C) Strong immunoreaction of cells for CD10.