An unusual case of primary extra-uterine endometrial stromal sarcoma involving the gastrointestinal tract and review of literature

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Introduction

Extrauterine endometrial stromal sarcoma (ESS) is a rare and is usually typified by a delayed recurrence of a primary ESS. However, primary extrauterine ESS has been described in literature in the absence of a uterine pathology and have been found to occur in the ovary, fallopian tube, pelvic cavity, abdominal cavity as well as the retroperitoneum. We report an interesting case of extra-uterine ESS in a 44 year old lady who presented with a large abdominal mass involving the gastrointestinal tract.

Case Presentation

The patient is a 44 year old lady who presented with upper abdominal pain and was found to have a large upper abdominal mass in March 2009. CT scan showed a large mass extending from the greater curvature of the stomach to the distal segment of the transverse colon, likely a gastric carcinoma. There were liver metastases as well. She was also found to have a fibroid uterus but did not experience any menstrual problem. She underwent an exploratory laparotomy and was found to have a large tumor involving the distal stomach and extending to the transverse colon. A subtotal gastrectomy and an extended right hemicolectomy. Final histology revealed that the patient had a low grade endometrial stromal sarcoma involving the stomach and colon. The resection margins were clear.

However, as a metastatic uterine ESS was more common than a primary extra-pelvic ESS, the patient was advised to have a hysterectomy. In the meantime the patient was started on Megace (Megestrol) as she had liver metastases and she was also undecided on a hysterectomy. She finally underwent a hysterectomy in October 2009 but the histology was negative for malignancy hence it was concluded that she had a primary extrautine ESS of the gastrointestinal tract. She was continued on hormonal treatment and was asymptomatic with stable liver metastases. In 2012, she underwent a complete resection of the liver metastases and histology was compatible with ESS. She was started on letrozole after the surgery and has been disease free for the past 3 years.

Discussion

The mean age of ESS presentation is about 50 years, and an estimated half are pre-menopausal. The main complaint at presentation is abnormal vaginal bleeding; other symptoms and signs include abdominal discomfort, an enlarged uterus and visible mass or polyp on pelvic examination. The presentation of a primary extra-uterine ESS would depend on the site of occurrence and in this case the patient had upper abdominal pain. There are several hypotheses surrounding the origins of extra-uterine ESS, including ectopic endometrial stroma associated with endometriosis which was found in this patient during her hysterectomy.

References


There are only two previous reports of primary gastric ESS. The first article reported a lady who presented with epigastric pain and was found to have a gastric antral mass. She had a wedge resection of the stomach, and subsequent histologic analysis of the mass was consistent with ESS. In the second article, 2 cases were described. Both cases underwent endoscopic ultrasound (EUS)-guided fine needle aspiration (FNA) biopsy, but the final diagnosis of ESS was only made after resection of the tumour. In all 3 cases, the resected specimens showed spindle cells which were positive for CD10 staining, and negative for CD117 (c-kit), CD34 and desmin staining. Staining for these biomarkers can help in identifying ESS, while excluding other gastric malignancies such as gastro-intestinal stromal tumours (GIST). The fundamental management of all localised ESS is the surgical resection of primary tumour. ESS tends to strongly express oestrogen and progesterone receptors, hence bilateral salpingo-oophorectomy is usually recommended. Hormonal therapy has also been widely used in the treatment of ESS for advanced disease, recurrence, and as an adjuvant in early disease. As most ESS are found to have a high rate of steroidal receptor expression, these tumours tend to be hormone-responsive. The agents used include progestogens and aromatase inhibitors, and these serve to modify the hormonal profile such that it becomes unfavourable for tumour growth. In this case, our patient was kept on a maintenance hormonal therapy with megestrol acetate and subsequently letrozole following her operations, and she has not had any disease progression or relapse for the past 5 years.

Primary extrauterine ESS involving the gastrointestinal tract is uncommon. However, long term remission can be achieved when all the disease has been resected as demonstrated in this case.