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Liver tumours due to endometriosis and endometrial stromal sarcoma

AW Khan, M Craig, M Jarmulowicz and BR Davidson

Department of Surgery, Royal Free Hospital, London, UK

Background

Endometriosis can occur in unusual sites, liver involvement being first described in 1986. Extra-uterine malignant transformation in endometriosis has been reported, occurring mainly in the ovary. Liver involvement with endometrial stromal sarcoma (ESS) has not been previously reported.

Case outline

Two patients presenting with symptomatic liver masses related to endometriosis, who successfully underwent surgical intervention, are presented.

Case 1

A 31-year-old woman previously had been treated with hysterectomy and bilateral salpingooophorectomy for severe pelvic endometriosis. Six years later, she presented with malaise from bilobar liver involvement with endometrial deposits. She proceeded acutely to hilar obstruction with obstructive jaundice and portal vein thrombosis.

Case 2

A 59-year-old post-menopausal woman had earlier presented acutely from a ruptured mesenteric cyst, which showed features of endometrial stromal sarcoma (ESS). Two years later, she presented with symptoms from a large ESS occupying the right lobe.

Discussion

Endometriosis *per se*, as well as malignant transformation into ESS can involve the liver. These should be considered in women with hepatic space occupying lesions of unknown aetiology.

Keywords

Endometriosis; endometrial stromal saracoma; liver tumour; liver resection.

Introduction

Gynaecological tumours involving the liver are extremely rare. Endometriosis has been known to involve unusual sites [1], and five cases involving the liver, all the left lobe, have been reported to date [2–6]. We report the first case of bilobar hepatic endometriosis. The radiological features have previously been described [7]. Malignant transformation in endometriosis is also rare, occurring most commonly in the ovary [8]. An endometrial stromal sarcoma, presenting as a large space-occupying lesion in the liver, has not been previously reported.

Case reports

Case no. 1

A 31-year-old woman was admitted for investigation of a three-month history of malaise, abdominal distension and abnormal liver function tests. She had a history of severe pelvic endometriosis and had undergone previous hysterectomy (for a double uterus) and bilateral oophorectomy. She had been on 6-monthly oestrogen and testosterone implants, as hormone replacement therapy, for the preceding three years. Clinical examination on this admission revealed ascites and mild jaundice. Ultrasound scan showed a large mass in the right and a small one in the left lobe, and percutaneous biopsy (left lobe) obtained endometrial tissue. Ct scan confirmed a large non-enhancing lobulated mass in the right lobe liver parenchyma and extending into the right subhepatic region. Another mass was noted in the left lobe (Figures 1 and 2). The jaundice rapidly worsened (bilirubin 713umol/l) and she developed tense ascites. Angiography revealed portal vein thrombosis. At operation, the large right lobe liver mass, partly cystic and partly solid, together with its retroperitoneal extensions, was resected *en bloc* while the small left lobe mass was left *in situ*. The patient developed a bile leak postoperatively and recovered slowly over six weeks.

The patient was readmitted 18 months later, when CT showed two low attenuation masses in the left lobe with peripheral rim enhancement. Frank pus was aspirated with

Correspondence to: *BR Davidson, Professor of Surgery, Royal Free Hospital, Royal Free and UCL Medical School, Royal Free Campus, Rowland Hill Street, London NW3 2PF, UK*

Figure 1. *CT showing multi-lobular mass lesions involving right and left lobes of the liver (case 1).*

Figure 2. *CT showing inferior extension of the endometrioma to the right subhepatic space (case 1).*

resolution of pyrexia. Six weeks later, a follow-up CT scan showed complete disappearance of the left lobe disease. She remains well four years postoperatively on intranasal goserelin (anti-oestrogen).

Case no. 2

A 61-year-old woman presented with right upper quadrant abdominal pain, and hepatomegaly was noted on clinical examination. Two years previously she had undergone a laparotomy for acute abdomen. A ruptured cyst in the jejunal mesentery was found, which was removed along with a second cyst in the mesentery of the descending colon. Histological examination of both lesions showed low-grade endometrial stromal sarcoma (ESS). Subsequent hysterectomy and bilateral salpingooophorectomy was performed but these organs were free of tumour on histopathological examination.

Contrast CT scan of the abdomen (Figure 3) showed a large mass lesion occupying the entire right lobe of the liver. Serum tumour markers and liver function tests were normal. The patient proceeded to a right hepatectomy. No extrahepatic disease was noted. Histopathological examination confirmed a low-grade ESS. There has been no recurrence on four year follow-up.

Discussion

Two patients presenting with symptomatic liver masses related to endometriosis are presented. Endometriosis is considered a benign disease, but occasionally becomes severe and progressive. Spread beyond the pelvis is rare [9]. Similarities with the previously reported cases involving the liver include presentation with pain, a history of endometriosis and a requirement for surgical intervention [2–6]. Unique features of the present case are bilobar disease, associated portal vein thrombosis and rapid progression of jaundice. These features suggesting particularly aggressive disease may relate to the previous hormone implants [10] and high circulating oestrogen levels. Nevertheless, a good outcome was achieved with surgical resection. Genital tract anomalies predispose to early-onset endometriosis, possibly due to retrograde menstruation, so the present patient with genital tract duplication is quite typical [1]. The mechanism of spread of endometriosis to the liver is unknown. The proximity of reported lesions to the falciform ligament has suggested a route via vessels in the median umbilical and falciform ligaments [2]. Hepatic abcess within the remaining endometrioma in the left lobe may have developed as a consequence of low-grade biliary

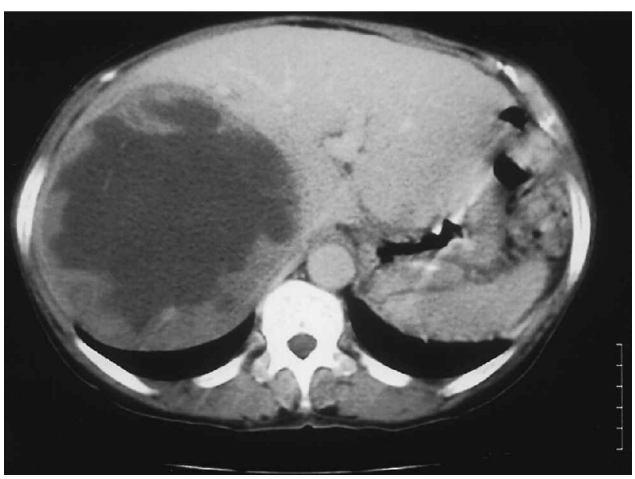

Figure 3. *CT showing replacement of the right lobe of liver by the tumour (case 2).*

sepsis postoperatively or from an unrelated bacteraemia after full recovery.

Endometrial stromal sarcoma (ESS) can occur in extrauterine sites in the absence of a primary uterine lesion [11]. Tumours may originate from endometriosis or from metaplasia of the pelvic peritoneal surface [12]. In this case, biopsy from the unruptured mesenteric cyst had shown foci of inactive endometriosis (of the type seen after the menopause) supporting the former hypothesis. The prognosis of ESS has been related to the mitotic index (1–10) of the resected tumour [12]. In the present case the mitotic index was 1, and the outcome with no evidence of disease four years following presentation with a symptomatic liver tumour would support resection of the lesion.

Endometriosis and ESS should be considered in women with liver tumours of unknown aetiological cause. Surgical resection would appear to have an important role in treatment.

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