CASE REPORT

Extrauterine fibroids: a diagnostic challenge and a long-term battle

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SUMMARY

Extrauterine fibroids often present a diagnostic challenge due to the unusual locations they arise from. We present a series of rare extrauterine fibroids. In recent years, these fibroids have been associated with previous morcellated hysterectomies or myomectomies. Our series of six patients were found to have extrauterine fibroids (confirmed through histology) and underwent open hysterectomy and open or laparoscopic myomectomy. Four had undergone previous laparoscopic myomectomies while the other two had no previous intra-abdominal surgeries. Postsurgical occurrence may be caused by incomplete removal of morcellated fibroid tissue. Spontaneous occurrence can be associated with congenital Müllerian system defects. Extrapolating from this hypothesis, we recommend physicians to make sure that counselling for extrauterine seeding and dissemination of unexpected malignancy is undertaken in cases of minimally invasive surgeries where morcellation is expected. Long-term tumour surveillance is thus essential in such instances.

BACKGROUND

Extrauterine fibroids are extremely rare. They are a diagnostic challenge due to the unusual locations that they arise from. Their differential diagnoses include parasitic leiomyoma (PL), disseminated peritoneal leiomyomatosis, uterine-like mass lesions, adenomyoma, endomyometriosis and leiomyoma with entrapped benign endometrial tissue.

In recent years, extrauterine myomas have been associated with previous morcellated hysterectomies or myomectomies.^{1–3} A larger case series by Kho and Nezhat¹ reported 12 patients with parasitic myomas between 2000 and 2008. Ten of 12 patients had previous abdominal surgery, and 8 had history of morcellation procedures (six laparoscopically and two by laparotomy), highlighting previous morcellation as a high-risk factor for iatrogenic parasitic myomas.

KK Women's and Children's Hospital is a tertiary centre that performs more than 1500 laparoscopic operations a year. At least 300 laparoscopic myomectomies and 200 laparoscopic hysterectomies are performed each year. We reported our first case of extrauterine fibroid-leiomyomatosis peritonealis disseminata in 2009.⁴ Now we present a series of these unusual fibroids that were managed in our centre in the year 2013. We describe these six clinical cases and discuss the possible aetiology, risk factors and management of extrauterine myomas (table 1).

CASE PRESENTATION

Case series of six individuals with extrauterine myomas.

Case 1

A 42-year-old Chinese woman with a history of asthma and hyperlipidaemia presented with an abdominopelvic mass. Physical examination revealed a 16-week-size uterus. Ultrasound of the pelvis revealed multiple uterine fibroids ranging from 2.7 to 8.7 cm. An open hysterectomy was performed. Intraoperatively, the patient was noted to have a mesenteric fibroid of 5 cm. Histology confirmed it to be an extrauterine adenomyoma (EA). The patient recovered well postoperatively.

Case 2

A 67-year-old Chinese woman with a history of hypertension, type 2 diabetes mellitus and hyperlipidaemia presented with postmenopausal bleeding. Ultrasound of the pelvis showed a large echogenic structure measuring 5.2×4.6×4.0 cm distending the endometrial cavity with an abnormally increased vascularity. It was suspicious for a neoplastic lesion. Subsequent uterine curetting revealed it to be endometrial adenocarcinoma. CT of the abdomen and pelvis showed a distended endometrial cavity with a heterogeneous mass within. There were no other organ involvements or lymphadenopathies seen. A separate heterogeneous mass with foci of calcifications at the left adnexal region was noted with left ovary being seen separately. This likely represented a subserosal fibroid. A staging laparotomy was performed. Intraoperatively, the aforementioned subserosal fibroid was found to be on the bladder serosa. Histology confirmed it to be a leiomyoma.

Case 3

A 41-year-old Malay woman presented with an asymptomatic abdominopelvic mass. A significant history included a laparoscopic myomectomy 5 years earlier. Ultrasound of the pelvis showed a right adnexal mass measuring 9.9×6.1×7.4 cm and a lobulated 13.5×10.4×7.7 cm mass in the left abdomen, at the level of the umbilicus. In view of the inability to demonstrate their connection to the uterus, MRI of the abdomen and pelvis (figure 1) was performed. It showed two large lobulated peritoneal masses with similar signal and enhancement characteristics. Disseminated leiomyomatosis (DL) was the clinical diagnosis. An exploratory laparotomy revealed a 12 cm fibroid that was adherent to the anterior abdominal wall at the umbilicus and a 10 cm free-hanging fibroid that was vascularised by

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Table 1 Characteristics of extrauterine myomas

Patient	Presenting symptoms	Previous intra-abdominal surgery	Location of the fibroids	Histology	Final diagnosis
1	Abdopelvic mass	Nil	Mesentery	Adenomyoma	Extrauterine adenomyoma
2	Postmenopausal bleeding	Nil	Bladder	Leiomyoma	Parasitic leiomyoma
3	Abdopelvic mass	Laparoscopic myomectomy	Omentum and anterior abdominal wall	Leiomyoma	Disseminated leiomyomatosis
4	Abdopelvic mass	Laparoscopic myomectomy	Rectosigmoid mesentery	Leiomyoma	Parasitic leiomyoma
5	Menorrhagia	Laparoscopic myomectomy	Ovaries and anterior abdominal wall	Leiomyoma, fibroma	Disseminated leiomyomatosis
6	Menorrhagia	Laparoscopic myomectomy, laparoscopic cystectomy	Anterior abdominal wall, Sigmoid mesentery	Leiomyoma	Disseminated leiomyomatosis

the omental vessels (figure 2). Histology confirmed both to be leiomyomas.

Case 4

A healthy 42-year-old Chinese woman presented with an asymptomatic abdopelvic mass. She had a laparoscopic myomectomy 7 years ago. Clinically she had an 18-week-size uterus. Ultrasound pelvis showed three fibroids measuring 9.1, 6.2 and 4.3 cm, respectively. An open myomectomy was performed. Two 4 cm fibroids, one arising from the left round ligament and the other from the right infundibulopelvic ligament were observed. A 9 cm fibroid was found attaching to the rectosigmoid colon and it was perfused by vessels from the rectal mesentery (figure 3). The patient also had three fundal fibroids. Histology confirmed them to be leiomyomas.

Case 5

An otherwise well 34-year-old Chinese woman reported menorrhagia of 1-year duration. She had undergone a laparoscopic myomectomy 5 years earlier. Ultrasound of the pelvis showed multiple fibroids with the three largest measuring 5.5, 4.7 and 4.3 cm, respectively. She underwent laparoscopic myomectomy. Intraoperatively, she was noted to have a 6 cm anterior abdominal wall fibroid at previous right port site (figure 4), a 4 cm fibroid attached to the right ovary and a 1 cm left ovarian fibroma. In addition, she had two uterine fibroids measuring 4 cm each. Histology confirmed them to be leiomyomas and left ovarian fibroma.

Case 6

A 34-year-old Malay woman presented with menorrhagia. A significant history included a laparoscopic myomectomy 7 years

prior, a laparoscopic cystectomy for endometriotic cyst 2 years prior and excision of infraumbilical leiomyoma 1 year prior. Ultrasound of the pelvis showed a left endometriotic cyst measuring 2.7 cm and multiple fibroids ranging from 0.8 to 3.6 cm. The patient's endometrial thickness was 12 mm. A laparoscopic myomectomy and left cystectomy were performed. Intraoperatively she was noted to have a total of 14 fibroids arising from the uterus, pelvic side wall and large bowel mesentery (figure 5). Histology confirmed them to be leiomyomas and endometriotic cysts.

Unfortunately, the patient's postoperative recovery was complicated by a sigmoid colon perforation from the excision of the myoma. A Hartmann's procedure was performed. She recovered uneventfully thereafter with the successful reversal of the Hartmann's procedure 4 months later.

DISCUSSION

As shown in table 1, most of the patients in our case series were found to have leiomyomas through histology. PL develop when they are pedunculated off the uterine serosa or when fragments of a fibroid detach and implant and grow within the peritoneal cavity. They recruit additional blood supply from nearby organs, but do not directly affect endometrial blood flow. This phenomenon was recently reported to be more commonly associated with morcellated hysterectomies or myomectomies.^{1–3} DL is characterised by the presence of multiple smooth muscle growths in the peritoneal cavity. The possible causes could be either hormonal, subperitoneal mesenchymal stem cells metaplasia, genetic, or iatrogenic after morcellation of myoma.^{5 6}

Four of our cases, that is, cases 3–6 had previous laparoscopic myomectomies, which predisposed them to developing

Figure 1 MRI of a myoma in upper abdomen.

Figure 2 A myoma that gains blood supply from omentum.

Figure 3 A myoma that grew in rectosigmoid mesentery.

extrauterine fibroids. With the advent of the power morcellator, the number and the size of fibroids that can be removed through laparoscopy increased exponentially. A power morcellator is a hollow cylindrical instrument that penetrates the abdominal wall, ending with sharp cutting blades, through which a grasper can be inserted to pull the myoma into the cylinder to cut out extractable pieces. Morcellation is associated with spreading of cellular materials of the morcellated tissue. These loose fibroid fragments may become infarcted, necrotic or even parasitic and disseminated if they are left behind.³ In addition, there is a risk of disseminating unexpected malignancy with an increase in mortality. The rate of unexpected sarcoma after morcellation is 0.09%. Disseminated disease occurred in more than half of the cases.⁷ We note a previous report of leiomyoma growing in an abdominal wall incision after laparoscopic retrieval suggesting implantation of the fibroid at the port site during removal.⁸ Case 5 had a similar parasitic fibroid at the previous laparoscopic port site. We emphasise the importance of careful and diligent removal of every single morcellated fibroid specimen. Special attention should be paid to falling pieces during morcellation and removal via the port site. Reverse Trendelenburg position after morcellation and copious peritoneal lavage followed by a thorough inspection is recommended to aid the removal of remnant myoma pieces.³

The other two cases, however, did not have previous intra-abdominal surgery. In case 2, it is likely that the fibroid

Figure 4 A myoma that grew at previous port site.

Figure 5 Laparoscopic view of a myoma that grew in sigmoid mesentery.

pedunculated off the uterine serosa and developed angiogenesis from the bladder serosa in view of its close proximity to the uterus.

The only exception in our case series was case 1, which revealed EA on histology diagnosis. Adenomyomas are benign tumours composed of smooth muscles, endometrial glands and endometrial stroma. They are differentiated from leiomyomas by the presence of intrinsic endometrial glandular and stromal elements.⁹

EA is linked to four possible aetiologies, (1) Müllerian duct fusion defect, (2) subcoelomic mesenchyme transformation, (3) Müllerianosis and (4) endomyometriosis.^{9–11}

1. The first aetiology is due to an abnormal development of the female genital tract. Each fetus has two pairs of genital ducts: Wolffian or mesonephric in the male and Müllerian or paramesonephric in the female. The Müllerian duct is formed by craniocaudal invagination of thickened coelomic epithelium on the anterolateral surface of urogenital ridge. Failure of fusion of Müllerian ducts results in duplications or atresia of the uterus. This has been suggested to be the cause of uterine-like masses.¹²
2. The subcoelomic mesenchyme or 'secondary Müllerian system' is a layer of tissue that lies underneath the mesothelial surface of the peritoneum. In adults, it lies underneath the subserosal stroma of uterus, ovaries, tubes and uterine ligaments.^{10 11}
3. Müllerianosis was first described by Batt as heterotopic Müllerian rests incorporated into other organs during organogenesis.^{13 14} This theory, as well as the first two theories, explains that the lesions arising from the multipotent cells of the Müllerian system may possibly proliferate in response to hormones.
4. Endomyometriosis is essentially endometriosis with smooth muscle hyperplasia or metaplasia.^{15 16}

Case 1 did not have a history of endometriosis; nor did she have any other urogenital abnormalities. Her lesion was within the lower abdominal cavity. Hence, we believe the cause of her EA was most likely due to subcoelomic mesenchymal stem cells metaplasia.

Prior diagnosis of extrauterine fibroids is often difficult owing to non-specific clinical and radiological findings. Half of the individuals in our series presented with abdominopelvic masses. The fibroid from case 2 was an incidental finding during her staging laparotomy whereas cases 5 and 6 presented with menorrhagia. Preoperative pelvic ultrasound mostly reported uterine fibroids

except in case 3, whose MRI eventually diagnosed her with DL due to the unusual upper abdominal location of one of the fibroids. However, the larger sizes and smaller numbers of the lesions are atypical of the usual DL. In case 5, the anterior abdominal wall fibroid was not even reported preoperatively.

Preoperative imaging including MRI can be used as a guide for operative planning. The use of MRI is especially useful when fibroids are growing rapidly, have poor delineation on ultrasound and where malignancy is suspected. That said, we note that MRI may not always represent the anatomy accurately. In a case report by Sewell and Russo,¹⁷ preoperative MRI was performed but failed to report the location of a retroperitoneal leiomyoma. The exact anatomical location of the fibroid is often ascertained intraoperatively. Gynaecologists should be prepared for fibroids in unusual locations. Albeit rare, malignant transformation should always be borne in mind.

All our patients were managed surgically, four by laparotomy and two by laparoscopy. Complete surgical excision is considered the mainstay and definitive treatment for extrauterine myomas.^{1 17} Recently, the Food and Drug Administration of USA discouraged the use of laparoscopic power morcellation during hysterectomy or myomectomy for the treatment of women with uterine fibroids in their safety communication notice. One of the largest suppliers of the device has subsequently suspended the device's sale until the medical community redefines the role of morcellation. Despite decades of experiences, the understanding of short-term and long-term sequelae of morcellation is limited. During preoperative counselling, especially for patients who are undergoing laparoscopic myomectomy and hysterectomy where morcellation is anticipated, it is imperative to include the possible risk of recurrence at extrauterine locations despite the surgeon's best efforts and possible dissemination of unexpected malignancy with associated increase in mortality.

Long-term follow-up of patients with extrauterine fibroids is recommended. As they seem to be hormone sensitive, reduction

of oestrogen exposure should result in regression.⁶ Carinelli *et al*¹² reported two cases of EA and uterine-like masses, one of whom had a relapse after 6 months. Long-term therapy with monthly gonadotropin-releasing hormone agonist in both patients prevented recurrence, at 4 and 10 years, respectively. Hence hormonal treatment and its side effects should be considered in a bid to prevent recurrence. Repeat surgeries for recurrent extrauterine fibroids usually entail greater surgical difficulty and risks including visceral injuries. A detailed discussion of the pros and cons of observation versus treatment should be carried out with such patients.

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Learning points

- It is difficult to confirm diagnosis by clinical judgement and radio-imaging modalities alone. Most often than not, definitive diagnosis is only made intraoperatively.
- Spontaneous occurrence is associated with Müllerian system defects or endometriosis.
- Increasing incidence is expected in patients with previous morcellated hysterectomies and myomectomies.
- Preoperative counselling of recurrence at extrauterine sites and possible dissemination of unexpected malignancy is imperative.
- Since these lesions are hormone sensitive, long-term tumour surveillance is essential.

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