Case report

A 30-year-old unmarried woman, who had undergone total abdominal hysterectomy (TAH) and bilateral salpingo-oophorectomy (BSO) for multiple uterine fibroids and a right ovarian cyst 5 years earlier, presented with symptoms of irregular bleeding per vagina, pain and heaviness of the lower abdomen in September 2009. Eight years prior to TAH and BSO, she had had laparotomies at which myomectomies were carried out on two occasions. She was started on oestrogen replacement therapy following her last surgery and had been doing well for 5 years. The oestrogen replacement therapy was discontinued following her complaints and she was started on progestogens, whereupon the vaginal bleeding ceased.

She was found to have 0.15 m x 0.15 m abdominopelvic mass that was firm, non-tender and had restricted mobility. Ultrasonography (USG) evaluation elsewhere showed a 0.15 m x 0.10 m solid pelvic mass, which appeared to be causing bilateral hydronephrosis. She was advised to undergo another laparotomy to remove the mass. Routine preoperative chest radiography, however, revealed multiple, bilateral pulmonary nodules that were also confirmed by computed tomography (CT). With a diagnosis of advanced malignancy with pulmonary metastases, she was referred to the Christian Medical College and Hospital for further management in February 2010. The CT images were reviewed and the findings confirmed (Fig a). A USG-guided biopsy of the pelvic mass was reported as showing benign leiomyoma.

At laparotomy, there were two leiomyomas, measuring 0.10 m x 0.10 m and 0.08 m x 0.08 m, bunched up together and were densely adherent to posterior bladder wall and sigmoid colon. The bladder was injured inadvertently during surgery and repaired with a urologist’s assistance. The patient developed consumptive coagulopathy, associated with}

Benign metastasising leiomyomatosis is a rare condition affecting women in the reproductive age-group with a history of uterine fibroids, who have undergone treatment by myomectomy or hysterectomy. It is characterised by development of multiple, indolent, smooth muscle tumours outside the uterus, most commonly in the lungs, and manifests several years after the uterine surgery. We describe the case of a young woman, who had undergone total abdominal hysterectomy and bilateral salpingo-oophorectomy for multiple fibroids and a right ovarian cyst. After 5 years of being on oestrogen replacement therapy, she was detected to have benign metastasising leiomyoma, for which an additional laparotomy was performed. At laparotomy, removal of the pelvic mass was associated with several complications. The metastatic lesions in the lung responded well to progestogens (megestrol acetate) alone as evidenced by regression of the lesions detected at follow-up after 6 months and 1 year.

Key words
Leiomyomatosis; Lung Neoplasms; Megestrol acetate; Progestins

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FIG. (a) Multiple, bilateral discrete pulmonary nodules on chest computed tomography. (b) Histopathology of the pelvic mass: closely packed bundles of smooth muscle cells with no cellular atypia or mitotic activity (H&E, x 20)
Benign metastasising leiomyomatosis (BML) was first described by Steiner in 1939. It is now used to describe the presence of cytologically bland, mitotically inactive smooth muscle tumours in extrauterine sites, which occur in conjunction with similarly appearing or previously removed leiomyomas (at myomectomy or hysterectomy). Although they are most commonly found in the lungs, they have also been detected in lymph nodes, skin, bones, the retroperitoneum, heart, and the brain. Much controversy surrounds the pathogenesis of BML and several mechanisms have been proposed. These include lymphovascular embolisation; seeding from ruptured leiomyoma; metaplasia from mesothelial mesenchyme and true metastases from low-grade leiomyosarcomas of the uterus.

Most commonly patients present with pulmonary lesions, and the history reveals that years earlier the women had undergone myomectomy or hysterectomy for uterine fibroids. Moreover, like our patient, they were often started on hormone replacement therapy following TAH and BSO at a young age. Benign metastasising leiomyomatosis has also been described following uterine artery embolisation. Late recurrences (even after 20 years) have been reported. Usually the patients are asymptomatic, and the lesions are often discovered incidentally. Symptoms, if present, include mild cough, dyspnoea, and flu-like illnesses. Rarely do patients progress to respiratory failure. The differential diagnosis includes primary or metastatic leiomyosarcoma, pulmonary hamartomas, primary pulmonary leiomyomas, lymphangiomyomatosis and inflammatory pseudotumours.

The pulmonary lesions tend to be bilateral, multiple, nodular, and well-circumscribed and do not display contrast enhancement on CT. The nodules show no significant metabolic activity on positron emission tomography scans. The pathology of pulmonary nodules retrieved by thoracoscopy-guided biopsy appears consistent with leiomyomas. Immunohistochemical staining is positive for oestrogen and progesterone receptors, thereby confirming their uterine origin. In the present clinical scenario, the pulmonary lesions co-existed with the pelvic mass. An USG-guided biopsy of the mass revealed a benign leiomyoma, thereby obviating the need for a thoracoscopy-guided biopsy of lung lesions.

The optimal management of BML is still unclear, and several management options have been considered (Table). 'Wait-and-watch' strategy with close surveillance has been described by some, but can lead to late complications after several years. Local excision of the lung nodules along with TAH and BSO has been described by many authors, but may not be feasible or desirable in certain situations, owing to the associated morbidity.

The growth of BML was long thought to be oestrogen-dependent, as there were reports of spontaneous regression following pregnancy or menopause. Both surgical and medical castration have been described as effective, including treatment with selective oestrogen receptor modulators (eg raloxifene), aromatase P-450 inhibitors (eg anastrozole), and gonadotropin-releasing hormone agonists. Some authors have described success with the use of luteinising-hormone releasing hormone analogues for treating BML.

Varied results have been reported following treatment of BML using progestogens. Cohen and Robins reported progressive worsening of lesions with their use and complete regression on...
TABLE. Management options of benign metastasising leiomyomatosis in the literature

<table>
<thead>
<tr>
<th>Author(s)</th>
<th>No. of case</th>
<th>Prior surgery</th>
<th>Age at presentation (years)</th>
<th>Site of metastasis</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hoetzenecker et al., 2009</td>
<td>1</td>
<td>TAH</td>
<td>56</td>
<td>Lung</td>
<td>Expectant</td>
<td>Increased tumour size</td>
</tr>
<tr>
<td>Rivera et al., 2004</td>
<td>1</td>
<td>TAH</td>
<td>37</td>
<td>Lung</td>
<td>Raloxifene + anastrozole</td>
<td>Regression</td>
</tr>
<tr>
<td>Hague et al., 1986</td>
<td>1</td>
<td>TAH</td>
<td>NA</td>
<td>Lung</td>
<td>LHRH analogue</td>
<td>Regression</td>
</tr>
<tr>
<td>Cohen and Robins, 1993</td>
<td>1</td>
<td>TAH</td>
<td>NA</td>
<td>Lung</td>
<td>Progestins (MPA followed by megestrol)</td>
<td>Progression</td>
</tr>
<tr>
<td>Wentling et al., 2005</td>
<td>1</td>
<td>TAH + USO</td>
<td>37</td>
<td>Lung</td>
<td>Thoracotomy + progestins (megestrol)</td>
<td>Regression</td>
</tr>
<tr>
<td>Motegi et al., 1993</td>
<td>1</td>
<td>TAH + USO</td>
<td>47</td>
<td>Lung</td>
<td>Progestins (MPA)</td>
<td>Regression</td>
</tr>
</tbody>
</table>

* TAH denotes total abdominal hysterectomy, NA not available, LHRH luteinising-hormone releasing hormone, MPA medroxyprogesterone acetate, and USO unilateral salpingo-oophorectomy.

Our report describes a patient whose BML occurring following multiple surgeries for uterine leiomyomata. Occurrence following TAH and BSO has been reported sparsely in literature and hormone replacement therapy may also play a role. At laparotomy for removal of the tumour, potential life-threatening complications ensued. We found megestrol acetate to be a simple and effective drug for treating hormone receptor–positive BML, thereby obviating the need for additional procedures like thoracotomy which are associated with increased morbidity.

References