Giant uterine leiomyoma: a case report with literature review

Sonali Kalyan, Sonam Sharma*

Department of Pathology, Kalpana Chawla Government Medical College, Karnal, Haryana, India

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*Correspondence:
Dr. Sonam Sharma,
E-mail: drsonamsharma@gmail.com

ABSTRACT

Uterine leiomyomas are one of the most common entities encountered in routine gynaecological practice; however, the giant uterine myomas are very rare and can often create a diagnostic dilemma and therapeutic challenge owing to their size, non-specific clinical presentation and degenerations. Here, in this article we review the literature on giant uterine leiomyomas and report one such case in a 38-year-old woman who presented with the complaints of vague abdominal lump, enlargement of abdomen, dysmenorrhea, lower abdominal and pelvic pain since last 2 years and a 6-month history of increased frequency of micturation. Physical examination and radiological investigations suggested a giant abdominopelvic mass, probably a uterine or an ovarian malignancy. An exploratory laparotomy was performed followed by total hysterectomy. Grossly, the specimen was a 15.2 kg uterine leiomyoma measuring 18x18x13 cm in size. Histologically, the benign leiomyoma showed cystic degeneration. The patient’s post-operative and follow-up period was uneventful.

Keywords: Cystic degeneration, Giant tumor, Leiomyoma, Uterus

INTRODUCTION

Uterine leiomyomas are one of the commonest benign tumors of the female genital tract, arising from the smooth muscle cells of the myometrium.1 The chances of their occurrence increases during the reproductive period and usually decreases after menopause.2 They are found in more than half of the women over the age of 35 years but can also be present in adolescents.3 Their exact pathogenesis is still debatable, however hormonal stimulation by estrogen, progesterone and other growth factors plays a pivotal role. Many risk factors have also been implicated for their occurrence like family history, nulliparity, black race, obesity, meat consumption and hypertension. Based on their location they can be categorized as intramural, submucosal and subserosal. Leiomyomas may appear as single or multiple and can be of varying sizes.4 Most of the patients have small uterine leiomyoma. However, giant ones do exist and are extremely rare.5 They are usually asymptomatic, but large tumors often produce abnormal bleeding, pelvic discomfort (pressure or pain), dysmenorrhea, infertility, frequent urination, constipation, “myomatous erythrocytosis syndrome”, Pseudo-Meigs syndrome or preterm labour.6 We herein describe an interesting case of giant uterine leiomyoma with degenerative changes along with review of the pertinent world literature so as to create awareness among the dealing clinicians about this enigmatic entity.

CASE REPORT

A 38-year-old illiterate woman from a rural background presented to gynaecological outpatient department with the complaints of vague abdominal lump, enlargement of abdomen, dysmenorrhea, lower abdominal and pelvic pain since last 2 years and increased frequency of micturation from last 6 months. There was history of recent weight gain and loss of appetite. She had 2 children and her last child birth was 5 years back. Her medical history was non-contributory; she had no major illness or any previous surgical procedures. Her family
history was not significant. On general examination, her vitals were normal except a protuberant abdomen was seen. On abdominal examination, her abdomen was circumferentially distended by a huge abdominopelvic mass, which was non-tender, firm in consistency and dull on percussion. Per speculum examination revealed normal external genitalia and uterine cervix. On bimanual examination, fornices were full and a very large, firm, mobile, central mass that filled the entire pelvis and abdomen was felt. It was difficult to specify the origin of the mass.

Abdominal ultrasonography confirmed the presence of a huge heterogeneous soft tissue mass arising from the pelvis and occupying almost the entire abdomen, the uterus was poorly visualized. Contrast-enhanced computed tomography (CECT) of the abdomen and pelvis revealed a large solid-cystic mass in the pelvis extending into the abdominal cavity. The mass appeared to be arising from the uterus. The right ovary and fallopian tube were normal, however the left sided adnexa was not visible. The lymphnodes were not enlarged. Her routine hematological, microbiological and other biochemical tests including serum CA 125 (cancer antigen 125) levels and pap smear were within normal limits.

Considering the size of the mass and suspicion of a uterine or an ovarian malignancy, the patient underwent laparotomy. On exploration, bilateral ovaries and fallopian tubes were normal and hence, a total abdominal hysterectomy was performed. The specimen was sent for histopathological examination. On gross examination, a total hysterectomy specimen was received measuring 25x19x15 cm in size. Cervical length measured 3 cm and cervical diameter measured 3.5 cm. External surface of the uterus showed few congested blood vessels. On probing, endocervical canal was patent. It weighed 15.2 kg (Figure 1).

Cut surface showed a large, grayish-white, sharply circumscribed, firm intramural fibroid measuring 18x18x13 cm in size with characteristic “raw silk” and whorled pattern. Few cystic areas were also evident. No haemorrhage or necrosis was seen. Endometrial canal was compressed, and its thickness measured 0.2-0.3cm (Figure 2).

Microscopic examination revealed features of a benign leiomyoma with cystic degeneration (Figure 3). The cervix was unremarkable. After the surgical procedure the patient was hospitalized for 10 days and the postoperative period was uneventful. Her symptoms resolved and there were no fresh complaints on 2 month follow-up.
DISCUSSION

Giant uterine leiomyomas are uncommon benign neoplasms and are those which weigh more than 11.4 kg or have a diameter which is more than 17 cm or dimension 33x28x22 cm. In literature, till date less than 100 cases of giant uterine myomas have been documented worldwide. The largest uterine fibroid ever reported weighed 63.3 kg, which was removed postmortem in 1888.

Table 1: Giant uterine leiomyoma cases reported in the literature from the year 1800 to 2010.

<table>
<thead>
<tr>
<th>First Author (Reference)</th>
<th>Clinical presentation</th>
<th>Weight/size of leiomyoma</th>
<th>Other pathological features and associated conditions</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hunter SH⁸</td>
<td>AS and pressure symptoms</td>
<td>63.6 kg</td>
<td>Simulating ovarian cyst having both solid and cystic areas</td>
<td>Died of pneumonia after 48 hours of operation</td>
</tr>
<tr>
<td>Behrend⁸</td>
<td>AS</td>
<td>60.7 kg 60x58x29 cm</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Singhabandhu B⁹</td>
<td>NA</td>
<td>45.4 kg</td>
<td>NA</td>
<td>Survived</td>
</tr>
<tr>
<td>Jonas HS⁹</td>
<td>NA</td>
<td>29.48 kg</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Evans AT¹⁰</td>
<td>NA</td>
<td>11.4 kg</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Mazzocconi G¹¹</td>
<td>NA</td>
<td>NA</td>
<td>Pedunculated fundus uteri</td>
<td>Uncomplicated</td>
</tr>
<tr>
<td>Ozsaran AA¹²</td>
<td>AM and plethora</td>
<td>14.2 kg</td>
<td>Subserous myoma associated with erythrocytosis</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Djelmis J¹³</td>
<td>NA</td>
<td>25 kg</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Oelsner G¹⁴</td>
<td>Severe pulmonary hypertension and respiratory failure</td>
<td>40 kg and 43.2 kg</td>
<td>NA</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Nguyen-Duc H¹⁵</td>
<td>AM, menorrhagia and severe anemia</td>
<td>NA</td>
<td>Giant leiomyoma in a 15-year-old teenager</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Costa BL¹⁶</td>
<td>AM, constipation and respiratory difficulty</td>
<td>12.4 kg</td>
<td>NA</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Perez M¹⁷</td>
<td>AD and WL</td>
<td>27 kg 65x54x25 cm</td>
<td>Uterine myoma with myxomatous degeneration, calcification and chondroid metaplasia</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Panayotidis C¹⁸</td>
<td>Pelvic mass and pressure symptoms</td>
<td>4.5 kg 31x26x15 cm</td>
<td>Fetal shaped fibroid uterus (multiple fibroids)</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Nappi L¹⁹</td>
<td>AD</td>
<td>27.7 kg</td>
<td>Bilobed giant myoma</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Karim T²⁰</td>
<td>AM and AD</td>
<td>3.2 kg 25x15x10 cm</td>
<td>Giant fibroid in a 16-year-old adolescent girl</td>
<td>Uneventful</td>
</tr>
</tbody>
</table>

On the other hand, the largest one ever removed from a patient who survived the procedure weighed 45.4 kg. This fibroid was reportedly the 34th weighing more than 18.2 kg since 1878.⁸

On further extensive reviewing the cases since the year 2011 till date (Table 2 and 3) it was noted that there was a surge in the number of patients being reported with giant uterine leiomyomas.

The uterine fibroids have an unremarkable potential to grow to an incredible extreme size before disabling the patient or producing appreciable symptoms. The only limit on size seems to be the capability of the host to bear it. This is owing to the relatively large volume of the abdominal cavity, the distensibility of the anterior abdominal wall, and the slow growth rate of this tumor.
Table 2: Literature review of giant uterine leiomyoma cases reported from the year 2011 to 2014.

<table>
<thead>
<tr>
<th>First Author (Reference)</th>
<th>Clinical presentation</th>
<th>Weight/size of leiomyoma</th>
<th>Other pathological features and associated conditions</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steward RG(^\text{21})</td>
<td>AE with history of uterine fibroids</td>
<td>11.618 kg 31x26x14 cm (largest)</td>
<td>Conglomerate, pedunculated leiomyomata</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Savulescu F(^6)</td>
<td>AE, WG, pressure symptoms, dysmenorrhea, dyspareunia, menorrhagia, lower abdominal and pelvic pain, frequent urination and constipation</td>
<td>18.1 kg 33x28x22 cm</td>
<td>Bilobed solid tumor with multiple subserosal fibroids</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Murtaza B(^22)</td>
<td>AM, dull pain in lumbar region, constipation, WL</td>
<td>7.4 kg 55x40x35 cm</td>
<td>Pedunculated subserosal fibroid with bilateral hydronephrosis</td>
<td>Uneventful with complete regression of bilateral hydronephrosis after 2 months of surgery.</td>
</tr>
<tr>
<td>Öndeş B(^23)</td>
<td>AD</td>
<td>25 kg 33x28x23 cm</td>
<td>Few cystic and ulcerated areas</td>
<td>Postoperative wound dehiscence</td>
</tr>
<tr>
<td>Al-Obaidi SM(^24)</td>
<td>AM, AD, constipation, difficulty in initiation of micturition</td>
<td>19.8 kg 60x45x25 cm</td>
<td>Atrophied uterus with fibroid and cystic degeneration of benign ovarian tissue</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Alam IP(^2)</td>
<td>AM, AD, occasional constipation, increased frequency of micturition</td>
<td>9.5 kg</td>
<td>Multiple fibroids, few showing necrosis and few others showing cystic and degenerative changes</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Aydin C(^25)</td>
<td>Lower abdominal pain, AD, recent WG of 25 kg</td>
<td>33x20x18 cm</td>
<td>Multiloculated, cystic, pedunculated leiomyoma with extensive cystic degeneration</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Mate S(^26)</td>
<td>Acute dyspnea and severe acute anemia (due to intratumoral bleeding)</td>
<td>13.5 kg</td>
<td>Pedunculated leiomyoma</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Gajewski M(^27)</td>
<td>AE</td>
<td>25 cm in diameter</td>
<td>Pedunculated leiomyoma with cystic degeneration</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Funaki K(^28)</td>
<td>AD and WG</td>
<td>8 kg 40x40x30 cm</td>
<td>Subserous leiomyoma with cystic degeneration</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Konard Wronski(^1)</td>
<td>Abdominal pain, AD, constipation</td>
<td>24.2x17.3 cm</td>
<td>Uterine leiomyomata with cystic degeneration</td>
<td>Uncomplicated</td>
</tr>
<tr>
<td>Ezugwu EC(^4)</td>
<td>AS, infertility, WL</td>
<td>16.8 kg (total fibroid weight) Largest: 12.4 kg, 32x24 cm</td>
<td>Large conglomerate fibroid masses attached to fundus along with multiple intramural fibroid seedlings.</td>
<td>Uneventful recovery and successful pregnancy seven months following myomectomy.</td>
</tr>
<tr>
<td>Moris D(^7)</td>
<td>Progressive constipation, increasing abdominal size, back pain, vague abdominal pressure sensations and urinary frequency</td>
<td>28.1 kg 62x39x21 cm</td>
<td>Solid and nodular</td>
<td>Uneventful</td>
</tr>
</tbody>
</table>

As a result, these patients can present in a physical condition which is unlike that of a typical woman with fibroids, but instead more like that of a much older patient with multiple comorbidities. However, the giant neoplasms frequently causes abnormal periods, pelvic pain and pressure effects on lungs, urinary bladder, ureters and other adjacent organs leading to further complications like lower-limb thrombosis as well as renal and respiratory failure.

Table 3: Giant uterine leiomyoma cases reported from the year 2015 to 2018 (till date).

<table>
<thead>
<tr>
<th>First Author (Reference)</th>
<th>Clinical presentation</th>
<th>Weight/size of leiomyoma</th>
<th>Other pathological features and associated conditions</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kalayci TO29</td>
<td>Abdominal pain and AD</td>
<td>30x25x17 cm</td>
<td>Pedunculated subserosal leiomyoma with degenerative changes</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Yavuz A30</td>
<td>Menometrorrhagia and AS</td>
<td>9.8 kg</td>
<td>Associated umbilical hernia</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Mulayim B31</td>
<td>No symptoms or complaints</td>
<td>3.98 kg 17 cm</td>
<td>Intramural, cellular leiomyoma without secondary changes</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Sharma RP32</td>
<td>Gradual WG, increasing abdominal size, vague abdominal pressure sensations, dysmenorrhoea, dyspareunia, menorrhagia, lower abdomen and pelvic pain, frequent urination and constipation</td>
<td>20 kg 36.4x28.7x23.3 cm</td>
<td>Fibroid arising from fundus region</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Reshmy JR33</td>
<td>AD and lower abdominal pain</td>
<td>28x20x10 cm</td>
<td>Leiomyoma with cystic degeneration</td>
<td>NA</td>
</tr>
<tr>
<td>Rahman H3</td>
<td>AS, vague abdominal discomfort, lower abdomen and pelvic pain and frequency of urination</td>
<td>11.6 kg 43x32x23 cm</td>
<td>Associated erythrocytosis</td>
<td>Complete recovery after hysterectomy</td>
</tr>
<tr>
<td>Carbunaru A34</td>
<td>AM with left lower limb swelling</td>
<td>4.5 kg 25x21x20 cm</td>
<td>Myoma with hyaline degeneration and associated deep vein thrombosis of left lower limb</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Gennaro Della Rossa MN35</td>
<td>AM</td>
<td>23x19x12 cm</td>
<td>Intramural fibroid with minimal nuclear atypia and low mitotic activity with interstitial edematous degeneration without signs of necrosis</td>
<td>Uterine reconstruction following myomectomy with uneventful recovery</td>
</tr>
<tr>
<td>Ramteke S36</td>
<td>AD, WL, decreased appetite, bleeding per vaginum</td>
<td>6.5 kg 26x23x18 cm</td>
<td>Intramural fibroid</td>
<td>NA</td>
</tr>
<tr>
<td>Muller R37</td>
<td>AE and WL</td>
<td>15.6 kg</td>
<td>NA</td>
<td>Uneventful</td>
</tr>
<tr>
<td>Hrgovic Z38</td>
<td>AD</td>
<td>25 cm diameter</td>
<td>Myoma spontaneously regressed in size 3 years after delivery</td>
<td>Pregnancy outcome favourable</td>
</tr>
<tr>
<td>Bartos V39</td>
<td>AE</td>
<td>8.1 kg 30x30x20 cm</td>
<td>Subserous leiomyoma with regressive and degenerative changes</td>
<td>Uneventful</td>
</tr>
</tbody>
</table>


Furthermore, on enlargement, uterine fibroids undergo different types of degeneration such as hyaline, cystic, myxoid or red degeneration and dystrophic calcification. Hyaline degeneration is most common (60% cases of leiomyomas) while cystic degeneration occurs in 4% of such cases. These degenerations arise due to the inadequate blood supply and seem to depend on the degree and rapidity of the onset of vascular insufficiency. However, researchers have found no relationship between any symptoms and the incidence of degenerative
changes. The diagnostic approach whenever a uterine leiomyoma is suspected is the pelvic examination which is the first step of evaluation in such patients, however, usually the smaller ones are not palpated and it is only the very large myomas which can be felt during examination. Therefore, imaging studies should be considered as they are helpful in determining the location, number, size and extent of the leiomyomas.

Ultrasoundography is preferred as the initial screening tool because it is least invasive and most cost-effective investigation. Computed Tomography (CT) and Magnetic Resonant Imaging (MRI) are quite helpful especially for ruling out any malignant transformation and in differential diagnosis but they have certain limitations. On CT scans, sometimes the leiomyomas are indistinguishable from healthy myometrium unless they are calcified or necrotic while MRI has limited availability and high cost restricting its use. Tumor markers have also been implicated to play an important role in its diagnosis as based on them the likelihood of malignancy can be ruled out. However, there is no single best modality to diagnose it pre-operatively and usually the giant uterine leiomyomas are diagnosed after laparotomy on histopathological examination. The differential diagnosis includes adenomyosis, hematomata, uterine cancer (carcinoma, sarcoma and carcinosarcoma), ovarian and retroperitoneal cysts or malignancy. In the present case study too, it was confused with a uterine or an ovarian malignancy both clinically and radiologically. However, it turned out to be a case of giant uterine leiomyoma with cystic degeneration on histopathology.

The treatment in cases of giant uterine leiomyomas is usually individualized as both the symptoms severity and patients desire to preserve the fertility are very important factors in determining any therapeutic intervention. The options include expectant management, medical management, surgery, interventional procedures like uterine artery embolization and ablative techniques. Expectant management with observation is done in cases of women with asymptomatic small and large fibroids, since rarely uterine leiomyomas may undergo sarcomatous transformation. Patients with giant myomas present as unusual challenge even for the most experienced gynecologists mainly due to massive blood loss caused by increased vascularity and postoperative possible complications like injury to bowel and urinary tract, infections, haematomas. Therefore, the medical management using Gonadotrophin Releasing Hormone (GnRH) analogues has been used to suppress estrogen production, thereby reducing the size of existing myomas and blood loss before the surgery. Uterine artery embolization also aims to decrease intraoperative blood loss, as it causes myoma infarction with very less adverse effects. Surgical treatment includes hysterectomy, myomectomy and myolysis. The removal of giant uterine leiomyomas by total abdominal hysterectomy with or without salpingo-oophorectomy is the traditional treatment of choice as these leiomyomas may cause infertility and in the presence of pregnancy they may affect the outcome. However, other approaches like vaginal and laparoscopic hysterectomies are associated with less postoperative complications and quicker recovery of the patient as compared to laparotomy /abdominal approach. Myomectomy is performed through laparotomy, laparoscopy or hysteroscopy and is preferred in symptomatic women who refuse hysterectomy or have a desire to retain their fertility. However, it causes significant perioperative morbidity. Myolysis including mono or bipolar cautery, Nd-YAG laser vaporisation or cryotherapy is currently experimental and is associated with risk of recurrence.

CONCLUSION

Giant uterine leiomyomas are exceedingly rare and can present with variable clinical manifestations. Therefore, this entity should always be kept in mind while dealing with patients with giant abdominopelvic tumors. Ultrasound stays as the initial screening tool for its diagnosis, but most of them are usually diagnosed on laparotomy followed by histopathological examination. Nevertheless, the treatment approach for every patient should be individualized and appropriate multidisciplinary approach with meticulous patient pre and postoperative care is essential for a better outcome in such patients.

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REFERENCES