CASE REPORT

Giant uterine leiomyomata

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Objective: To present the case of a patient with giant uterine leiomyomata and review literature pertinent to the subject.

Design: Case report and literature review.

Setting: A major university, tertiary-care hospital.

Patient(s): One patient, with said pathology, who gave informed consent for exploratory laparotomy, total abdominal hysterectomy, and bilateral salpingoophorectomy.

Intervention(s): Exploratory laparotomy, total abdominal hysterectomy, bilateral salpingoophorectomy, reoperation with abdominal washout, and hemostasis for hemoperitoneum.

Main Outcome Measure(s): Not applicable.

Result(s): Not applicable.

Conclusion(s): Those with giant uterine leiomyomata are a very unique and tiny subset of the millions of women with fibroids. They should be treated similarly to older, more critically ill patients. Their optimal surgical management requires the careful attention to considerations and techniques not common to the typical myomectomy or hysterectomy. (Fertil Steril 2010; ■■■■-■■. ©2010 by American Society for Reproductive Medicine.)

Key Words: Giant, uterine, leiomyoma, myoma, fibroid, tumor

Uterine leiomyomata are benign neoplasms arising from uterine smooth muscle. Although their pathogenesis remains unclear, they are the most common tumor of the female reproductive tract, occurring in as many as half of women older than 35 years. By age 50 years, 80% of African American women and nearly 70% of Caucasian women have fibroids (1). Forty percent of abdominal hysterectomies and 17% of vaginal hysterectomies contain fibroids (2). Although they may cause menorrhagia, dysmenorrhea, and symptoms related to pressure and mass effect, half of women with fibroids are asymptomatic (2). Most of these patients have small fibroids. On rare occasions, however, fibroids can grow extremely large. It is in these cases that treatment is not standardized because proper management of patients with very large fibroids is complex and requires exceptional skill.

Although uterine leiomyomas are the most common tumor of the female reproductive tract, giant myomas (11.4 kg or more) are exceedingly rare. In 1977, Jonas and Masterson described one such case, stating it to be the 56th ever reported and only the eighth in the preceding 35 years (3). A PubMed search using the terms “giant,” “uterine,” “leiomyoma,” “myoma,” “fibroid,” and “tumor” returned only five similar cases since 1977.

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CASE REPORT

A nulligravid 33-year-old African American woman presented for an annual examination. She reported a history of uterine fibroids and a 10–15-year history of a slowly enlarging abdomen. She had not sought medical attention because of fear of surgery. Her menses were regular and moderate in volume. Past medical history was significant for anemia. Her body mass index was 20.0 kg/m². Physical examination revealed a cachectic woman with distended jugular veins and a massively protuberant abdomen containing a firm mass. The mass extended to the xiphoid and bilateral flanks and measured 50 cm above the pubic symphysis (Fig. 1). A transabdominal ultrasound scan showed multiple large masses resembling fibroids. Magnetic resonance imaging from the previous year showed a markedly enlarged and lobular uterus measuring 21.5 × 15.2 × 12 cm, containing intramural and pedunculated fibroids, the largest measuring 28 × 18.9 cm. The liver, diaphragm, bowel, and urinary system were compressed by the masses. Complete blood count revealed a hemoglobin level of 6.2 g/dL, hematocrit 21.8%, platelets 444,000/μL, and mean corpuscular volume of 60 fL. Tumors makers (CA-125, carcinoembryonic antigen) were within normal limits. The albumin/globulin ratio was 1.0.

After 1 month of iron supplementation, her hemoglobin level rose to 10.5 g/dL. The patient then underwent total abdominal hysterectomy and bilateral salpingoophorectomy because the fibroid encompassed both adnexae. A central venous catheter and an arterial line were placed to aid with hemodynamic monitoring. Only sequential compression devices were used for deep vein thrombosis prophylaxis. Intraoperative findings included almost absent subcutaneous tissue, 1 L of ascites, and extensive collateral vasculature involving the anterior abdominal wall, rectus muscle, and omentum. Dense
adhesions between bowel, omentum, and uterus were noted. Several extremely large, conglomerate, pedunculated fibroids were immediately encountered, the most anterior of which were adhered to the right pelvic side wall and anterior abdominal wall. After the adhesions were lysed, the masses were clamped and transected in two large pieces (Fig. 2). A supracervical hysterectomy was performed first, followed by trachelectomy. A hand-held vessel-sealing device was used throughout the case to expedite removal of the specimen and help control bleeding. Excellent hemostasis was noted before closure. Estimated blood loss was 2 L. The patient received IV fluids (total crystalloid and colloid, 4.3 L), packed red blood cells (PRBC, 4 U), and fresh frozen plasma (FFP, 2 U). Urine output was 450 mL. Pathology confirmed a myomatous uterus with conglomerate, pedunculated leiomyomata, the largest of which measured 31 × 26 × 14 cm, with an aggregate weight 11.618 kg.

Approximately 3 hours postoperatively, the patient developed hypotension (blood pressure 70/46), tachycardia (128 beats per minute), tachypnea (32 beats per minute), and low oxygen saturation (87%). Significant findings included cold extremities, a distended abdomen with an obvious fluid wave, and severe anemia despite 2 additional units of PRBC (hemoglobin and hematocrit 5.4 g/dL and 16.5%, respectively). The patient was immediately taken back to the operating room, where 3.5 L of blood were found in the abdomen, with diffuse oozing from raw peritoneal surfaces, areas of previous adhesion, the vaginal cuff, and bladder pillars. Disseminated intravascular coagulation was confirmed by an international normalized ratio of 2.4. After fluid resuscitation (1 L of crystalloid and 500 mL of colloid), blood product transfusion (12 U each of PRBC and FFP, three jumbo-packs of platelets, and 1 U of cryoprecipitate), and multiple hemostatic interventions (cauterity, hemostatic agents, and bilateral hypogastric artery ligation), the bleeding was controlled. Estimated blood loss was 5 L. After a 2-day stay in the shock–trauma intensive care unit, the remainder of her second postoperative course was uneventful, and she was discharged home on postoperative day 4.

DISCUSSION

The largest uterine fibroid ever reported weighed 63.3 kg; it was removed postmortem in 1888 (4). A 60.7-kg myoma was removed from a patient in 1930, but she died of pneumonia 48 hours later (3). The largest uterine tumor ever removed from a patient who survived the procedure weighed 45.4 kg (4). This fibroid was reportedly the 34th weighing more than 18.2 kg since 1878 (5). Two leiomyomata, weighing 40 kg and 43.2 kg, respectively, were removed from two separate patients without complication in 2003.

The differential diagnosis of leiomyomata includes adenomyosis, hematometra, and uterine cancer. The latter includes three main types: carcinomas, sarcomas, and carcinosarcomas. Carcinomas are by far the most common. Sarcomas (leiomyosarcomas and endometrial stromal tumors) and carcinosarcomas (malignant mixed müllerian tumors) are rare and clinically aggressive. They grow quickly, lymphatic or hematologic spread occurs early, and prognosis is generally poor (6). Otherwise, presenting signs and symptoms are often similar to those for benign myomas. Also like myomas, multiple theories of pathogenesis exist. Routine workup includes endometrial sampling and pelvic imaging. Treatment most typically consists of surgery with or without adjuvant radiation or chemotherapy.

The potential for uterine leiomyomas to grow to extreme size before causing symptoms is quite remarkable. This is likely due to the relatively large volume of the abdominal cavity, the distensibility of the abdominal wall, and the slow growth rate of these tumors. Indeed, the process may take decades. As a result, these patients can present in a physical condition not typical of most women with fibroids, but instead more like that of a much older patient with multiple comorbidities. Thus, they must be treated as such. Combined pre- and postoperative mortality rates are reportedly as high as 16% (4).

Imaging studies (ultrasound, CT, MRI) and tumor markers are helpful to define the extent of the mass and the likelihood of malignancy, respectively. Preoperative pelvic angiography with or without arterial embolization may help prevent blood loss related to neovascularization (7, 8). Correction of anemia, if present, is of paramount importance because blood loss is expected during the surgery. Decreased renal and respiratory function and an increased chance of pelvic and lower extremity venous thrombosis (due to pressure effects) should be managed accordingly (9). Optimization of nutritional status is helpful in light of the “parasitic effect” of the fibroid. Preoperative mechanical bowel preparation may decrease the risk of bowel injury and aid visualization.
Intraoperatively, central venous and arterial pressure lines help closely monitor volume status and vital signs. The patient should be positioned to allow adequate ventilation and reduce vena cava compression. The skin incision should allow both easy manipulation of the mass without causing avulsion of any pedunculated fibroids, as well as exploration of the upper abdomen. En-bloc removal decreases bleeding. Potential sources of hemorrhage, including collateral circulation, arterial–venous shunting, and sheer tumor size, must be addressed because these patients are more vulnerable to hypovolemic shock due to anemia, poor nutritional status, and decreased reserve (9). Hand-held vessel-sealing devices may prove superior to traditional techniques by allowing easier dissection in tight spaces and by shortening the time to specimen removal, thereby decreasing overall blood loss. Prophylactic use of agents for venous hemostasis, such as thrombin spray, should be utilized, especially in light of the high possibility of deperitonealized surfaces. Trachelectomy need not be performed in the patient with benign cervical and uterine pathology if operative time and/or blood loss are already an issue.

Close monitoring of hemodynamic and respiratory status in the postoperative period may discover early evidence of hemorrhage. If fluid and blood product resuscitation fail to quickly stabilize the patient, immediate surgical re-exploration is necessary. Signs and symptoms of occult bleeding are often less obvious. Changes in vital signs and on physical examination can be subtle and easily overlooked, especially with loss of less than 15% of the total blood volume (10). Intraoperative blood loss is often underestimated, and hematocrit values typically lag behind true losses (10). Abdominal and pelvic ultrasound and/or CT may be helpful in diagnosing hemoperitoneum or hematoma formation. Arterial embolization may be considered in the stable patient before reoperation.

Only the most experienced gynecologic surgeons should attempt such an operation. Even then, intraoperative consultations from gynecologic oncology, general, colorectal, urologic, and/or plastic surgeons may be helpful. Postoperative aid from physical and occupational therapists will also likely be beneficial.

Patients with giant myomas present an unusual challenge even for the most experienced gynecologist. Even though these patients are relatively young, they require care more typical of older, chronically ill patients. Conscientious perioperative management and multidisciplinary patient care are essential to prevent morbidity and mortality and speed recovery. Several specific points from this case are historically underemphasized (or altogether unmentioned) and deserve special attention: [1] preoperative optimization of nutritional and hematologic status, [2] careful attention to potentially massive collateral vessels (with possible benefit from preoperative angiography, with or without arterial embolization), [3] the use of hand-held vessel-sealing devices and prophylactic venous hemostatic agents, [4] the avoidance of trachelectomy and anticoagulative drugs, if possible, and [5] close postoperative follow-up with appropriate consultation may all be keys to good outcomes.

REFERENCES