Case of Intravenous Lipoleiomyomatosis in a Postmenopausal Woman

ABSTRACT

Leiomyomas are benign proliferations of smooth muscle in the uterus. Intravenous lipoleiomyomatosis is a benign neoplasm composed of adipose tissue and smooth muscle with extension into surrounding pelvic vessels. It is an extremely rare phenomena with 6 case reports existing in English literature to date. We report the case of a post-menopausal African American woman who was incidentally found to have IV lipoleiomyomatosis by pathology after hysterectomy with bilateral salpingo-oophorectomy for a suspected fibroid uterus with pelvic pain.

INTRODUCTION

Leiomyomata are benign monoclonal proliferations of myometrial smooth muscle and are the most common benign gynecological disorder [1]. On the other hand, intravenous lipoleiomyomatosis is a rare diagnosis, with very few case reports in the literature [2-7]. Prior case reports demonstrate that although histopathologically benign, intravenous lipoleiomyomatosis exhibits behavioral characteristics of a malignant tumor, in some cases extending into the IVC with intracardiac extension [2-4, 8]. We report the case of a post-menopausal African American female with chronic pelvic pain, hypertension, and morbid obesity, with pathologic diagnosis of intravenous lipoleiomyomatosis after total abdominal hysterectomy with bilateral salpingo-oophorectomy.

CASE

A 55 year old post-menopausal African American female with a history of known 20-week-size fibroid uterus and leiomyomas presented to the gynecology clinic for evaluation of chronic pelvic pain and pressure. Relevant comorbidities included hypertension, morbid obesity (BMI: 40.6), IBS (diarrheal type) and depression. The diagnosis of uterine leiomyoma was initially confirmed two years prior to this evaluation, during hospitalization for acute-onset LLQ and suprapubic pain attributed to likely degenerating fibroid. CT at that time was significant for an enlarged uterus measuring approximately 19cm x 13cm x 11cm with heterogeneous attenuation consistent with prior history of uterine leiomyomas. Ultrasound at that time showed two fibroids sized 9.6 cm x 6.9 cm x 9.9 cm and 6.4 cm x 6.4 cm x 6.6 cm. Patient was evaluated by the gynecologic oncology team, but was lost to follow up for two years prior to current evaluation. At present, she reported that her sensation of pelvic fullness, “similar to feelings of labor”, had persisted and was associated with lumbar back pain. She denied any urinary hesitancy or vaginal bleeding since her last menses three years ago. Prior to menopause, she had menometrorrhagia. Family history was negative for breast and gynecological malignancies. The patient was not sexually active with recent pap smear within normal limits. Social history is significant for ½ ppd tobacco use and rare alcohol use. Patient had no prior surgeries. Repeat US at this time showed the two fibroids similar in size to evaluation two years prior. Patient elected to proceed with total abdominal hysterectomy with bilateral salpingo-oophorectomy for relief of symptoms and definitive diagnosis.

Pre-op physical examination revealed blood pressure 126/62 mmHg, pulse rate of 95 bpm, BMI 40.6. The patient was well appearing and in no acute distress. Cardiovascular exam was benign with normal rate and rhythm without murmur, rubs or gallops. Her abdominal exam was limited by body habitus, but was with benign findings without distention or tenderness, with

normal bowel sounds. Gynecologic exam was deferred. Transvaginal and transabdominal ultrasounds revealed two large uterine fibroids consistent in size with US two years prior. Preoperative laboratory evaluation was unremarkable.

The operative and postoperative course was without complication and biopsies of endometrium, fibroids, and adnexa were sent for pathologic analysis. Final pathologic report was significant for intravenous lipoleiomyomatosis of uterine fibroid and cervix. Immunohistochemistry staining was positive for SMA, negative for HMB45, PanMel. Post-operatively, the patient was referred for consultation with Gynecologic Oncology at our institution. They ordered a baseline echocardiogram which did not show any lesions in the heart. She also had a baseline CT of the abdomen/pelvis with contrast which showed no pelvic lesions consistent with remaining intravenous lipoleiomyomatosis. She is currently eight months post-op with plan for CT imaging every three months for the first year after surgery with further follow-up pending findings.

DISCUSSION

Intravenous lipoleiomyoma (intravenous LPL) is a benign mixed neoplasm composed of adipose tissue and smooth muscle with extension into surrounding pelvic vessels. The benign nature of this tumor is defined by a lack of cellular mitotic atypia that is characteristic of its malignant sarcoma counterpart; however, its extension into surrounding vasculature poses unique diagnostic and therapeutic challenges. Theories regarding the pathogenesis of intravenous LPL suggest either intravenous extension of uterine leiomyomas or proliferation of the smooth muscle in the veins of the myometrium [9].

As a subtype of the more common lipoleiomyoma, intravenous lipoleiomyomatosis has potential to be overlooked in pathologic analysis when excised early in its clinical course, thus making the true incidence of the condition difficult to assess [8]. Intravenous leiomyomatosis (IVL) can be differentiated from intravenous LPL by the lack of defining adipose tissue by pathological analysis. Although the incidence of lipoleiomyoma is believed to fall in the range of .03% - 0.2% [10], IVL is estimated to occur in approximately 0.097% of all smooth muscle tumors of the genitourinary tract according to a single-center analysis by Du et al [11]. The incidence of intravenous LPL is thus likely much less, with only 6 cases of true intravenous lipoleiomyoma (LPL) in English literature to date [2-7].

Although benign, intravenous lipoleiomyoma has potential to cause significant morbidity and mortality secondary to its intravascular nature. Patients tend to present with symptoms similar to that of leiomyomas - pain, vaginal bleeding, pelvic pressure and dysmenorrhea [3]. However, several cases of intracardiac IVL have been documented, representing a wide variety of presenting symptoms. Cardiac symptoms have been reported in patients with significant intravascular tumor burden, ranging from cardiac insufficiency to pulmonary tumor embolism and sudden cardiac death [12].

Although intravenous LPL represents a rare histologic variant of IVL, it is believed to follow a similar clinical course. Vural describes the case of a woman who presented with chest pain and paroxysmal nocturnal dyspnea and was found to have intravenous lipoleiomyomatosis with tumor involvement of right atrium, IVC, right ovarian vein and right adnexa. This

demonstrates the ability of intravenous LPL to mobilize in a similar fashion to its more commonly reported counterpart [4]. Due to the rarity of myxomas originating in the right atrium, cardiac extension of IVL and intravenous LPL should be considered in the differential for all women [3, 5, 13-15].

As previously discussed, the majority of patients presenting with intravenous LPL present with symptoms typical of leiomyoma, thus posing a diagnostic challenge outside of careful histopathological analysis. Due to the rarity of the condition, potential for intravenous spread of the neoplasm, the potential for significant cardiac complications, and lack of clinical guidelines on how to manage patients with intravenous LPL, our patient is currently followed by the gynecology oncology team at our institution. She has had a negative baseline echocardiogram and does not have any cardiopulmonary symptoms. She has had serial CT imaging of the abdomen and pelvis without evidence of recurrent intravenous LPL, and is now eight months post-operative from her initial surgery. Our management of this patient was decided due to the concern for extension of intravenous LPL into the abdominal veins, and risk of travel to the heart. There are no clear guidelines for how to manage patients with intravenous LPL given the rarity of the condition; however, previous case reports have demonstrated this potential for cardiac involvement and subsequent compromise. Hence, we thought it prudent to obtain this baseline imaging.

Although rare, intravenous lipoleiomyomatosis represents an important cause of significant morbidity and mortality for patients due to its tendency to extend to distant sites. Patients may present with a variety of symptoms ranging from pelvic pain to signs of cardiac insufficiency, making accurate histopathological diagnosis prudent to guide treatment plans and postoperative surveillance.