Teaching Cases

Intravenous lipoleiomyomatosis of uterus with cardiac extension: A case report

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ARTICLE INFO

Article history:
Received 25 July 2010
Received in revised form 13 October 2010
Accepted 18 October 2010

Keywords:
Lipoleiomyomatosis
Intravenous
Intracardiac
Leiomyoma
Uterus

ABSTRACT

A case of intravenous leiomyomatosis (IVL) with histological features of a lipoleiomyoma (intravenous lipoleiomyoma) in a 48-year-old woman is reported. The patient, with the tumor located in the uterus and extended up on the right side of the heart through the inferior vena cava, was diagnosed as having a cardiac mass. She displayed symptoms of dyspnea, chest pain due to the cardiac mass, as well as pelvic pain and dysmenorrhea. She underwent cardiac surgery because of a right atrial mass, and the histopathological diagnosis was leiomyoma without the knowledge of a uterine mass. Afterwards, a right adnexal mass was detected in the pelvis, and a total hysterectomy–bilateral salpingo-oophorectomy was performed for the adnexial mass, which was also diagnosed as leiomyoma. In this case report, we describe an intravenous lipoleiomyomatosis of the uterus which, at the initial clinical presentation, showed cardiovascular symptoms. We emphasize the histopathological features and the differential diagnosis of this rare tumor in the light of the literature.

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Introduction

Intravenous leiomyomatosis (IVL) is a rarely encountered smooth muscle cell tumor that grows within venous channels but does not invade the tissues [1,2,8,9,13,15]. The tumor primarily extends through the uterine veins, sometimes as far as the inferior vena cava (IVC), reaching the right-side cardiac chambers and the main pulmonary artery, even resulting in death [1,2,11]. The lesions were first described by Birsh-Hirshfeld in 1896, and the first case report in the English literature was published by Marshall and Morris in 1959 [2,4,8,9,13]. More than 150 cases have been reported in the English literature so far [2,7].

Cellular, atypical (leiomyoma with bizarre nuclei), epithelioid, and lipoleiomyomatous histological variants have been described. These have the same behavior and prognosis as ordinary intravenous leiomyomatosis [8,15,17]. Here, we present a case of intravenous lipoleiomyomatosis of uterus with cardiac extension. The patient developed a critical condition with respiratory failure and was finally cured by surgical removal of the tumor. We also review the histopathological features of this rare tumor in the light of the literature.

Case report

A 48-year-old multiparous woman was admitted to the Department of Thoracic Surgery of Başkent University Hospital with dyspnea, chest pain extending to shoulders and arms, orthopnea, and paroxysmal nocturnal dyspnea of several months’ duration. A tumoral mass in the right atrium was detected, and the patient underwent cardiac surgery. At the time of surgery, a sausage-shaped mass measuring 9.5 cm in length and 2.5 cm in diameter was removed. The tumor occupied the right atrium, was not actually attached to the heart at any point, and extended down the inferior vena cava (IVC) as much as it could be palpated. Histological examination showed mixed hypo and hypercellular areas, composed of bundles of isolated spindle cells with oval nuclei and long eosinophilic cytoplasmic processes (Fig. 1a and 1b). There were small and large hyalinized vessels. No cytological atypia, tumor cell necrosis, or mitosis was seen. An immunohistochemical panel including the antibodies for SMA (smooth muscle actin), desmin, S-100, and CD 34 was performed. The tumor cells expressed SMA.
pose tissue, and small and large blood vessels with hypocellular and fibrous regions. Necrosis and cytologic atypia were absent, and there was less than one mitotic figure per 10 high-power fields. The fat cells showed mature appearance with unilocular cytoplasm. Nuclear pleomorphism and immature lipoblasts were not observed (Fig. 4). This lipomatous component was noticed more frequently in intravenous regions (right adnexial region) than in the other locations. A prominent infiltration of mast cells was observed. Finally, a diagnosis of a lipomatous, histological variant of intravenous leiomyomatosis was made.

Discussion

The term lipoleiomyoma (LPL) of uterus refers to a rare benign lesion consisting of smooth muscle and fat, whereas intravenous leiomyomatosis (IVL) of the uterus is a lesion capable of local intravascular extension, and is slightly more common[16]. Uterine lipoleiomyoma and intravenous leiomyoma are already different uncommon tumors of the uterus, and their combination is even rarer [5].

Intravenous leiomyomatosis is a benign smooth-muscle tumor arising either from a uterine leiomyoma or from the walls of a uterine vessel and extending into venous channels [1,2,8,9,13,15]. Although histologically benign, IVL might behave in a “malignant” fashion [7]. The tumor has the potential to grow inside the veins of broad ligament, extending to the IVC through different routes (i.e., iliac veins and ovarian veins), reaching the right heart chambers and occasionally the pulmonary artery, resulting in death [1,2,11]. Durck reported the first case of intracardiac extension of IVL in 1907 [3]. Because cardiac involvement is present in up to 10% of cases,
Fig. 4. Lipoleiomyomatous region of tumors with smooth muscle cells, adipose tissue, and small-large blood vessels.

It may be misdiagnosed as a primary cardiac tumor or a venous thrombus-in-transit [4,9,11].
There are two theories about IVL pathogenesis. The first theory claims that tumor arises from smooth muscle cells in uterine veins. According to the other theory, it develops by intravascular projection into an adjacent venous channel from a leiomyoma [2,12,17,18].

IVL has been reported in women between 23 and 80 years of age, with most patients being middle-aged (median: 44 years). The majority of patients present symptoms of typical uterine leiomyoma; i.e., pain, vaginal bleeding, pelvic pressure, and dysmenorrhea [18]. Even with extensive intravenous tumor extension, patients may remain asymptomatic until intracardiac growth leads to cardiac insufficiency, to pulmonary embolism, or even to sudden cardiac death [1,12,15]. Rarely are initial symptoms of the patients related to cardiac involvement such as congestive heart failure, dyspnea, syncope, orthopnea, pleuritic chest pain, pedal edema, or abdominal distension [4,8,9,12,18]. The correct preoperative diagnosis of IVL is difficult, and it is usually made by intraoperative findings or postoperative pathological examination [2,8].

Grossly, intravenous leiomyomatosis is a coiled or nodular growth within the myometrium with worm-like extensions into the uterine veins in the broad ligament or into other pelvic veins [4,9,14,17]. The growth extends into the vena cava in more than 10% of patients, and in some it reaches as far as the heart [17]. It grows into the vascular tree and occasionally metastasizes to the lungs without obvious invasion [15]. The tumor can be entirely free-floating within the vessel lumen or, less commonly, it can have attachments to the vessel or atrial walls [13].

Microscopically, the tumor is found within venous channels lined by endothelium [15,17]. Arteries are not involved. The tumor resembles a typical leiomyoma, or it may show the histological features of variants, such as cellular, atypical (leiomyoma with bizarre nuclei), epithelioid, myxoid and lipoleiomyoma. Smooth muscle cells may be inconspicuous and difficult to identify when the tumor contains extensive fibrosis and hyalinization. The intravenous growth itself is highly vascular, and in some cases it contains so many small and large blood vessels that it can mimic a vascular tumor [17]. Most cases are cytologically bland, with either mild or no nuclear atypia and with low mitotic index [15].

Intravenous lipoleiomyomatosis (LPL) is a very rare variant. The histogenesis of these tumors is controversial, and the proposed sources of origin include lipoblasts entrapped during embryogenesis, metaplasia of existing smooth muscle, lipogenic cells at the entrance of blood vessels into the uterine corpus, primitive embryonic mesoderm, and multipotent fibroblasts [6,10,16,18,19].

Cytogenetic study showed that they have similar chromosomal abnormalities with ordinary leiomyomas [19]. There are some case reports in the literature which suggest a relationship between menopause, obesity, or cholecystolithiasis and the occurrence of lipoleiomyoma, but there is no correlation between their occurrence and hyperlipemia or abnormal lipid metabolism [5]. The prognosis is as favorable as that in common leiomyoma. Macroscopically, the majority of tumors are located in the myometrium of the uterine corpus [5]. Histologically, these tumors are composed of an intimate admixture of spindle-shaped smooth muscle cells, hyalinized blood vessels, adipocytes, and inflammatory cells, especially mast cells. The spindle-shaped smooth muscle cells usually predominate. When an abdominopelvic intravenous mass is demonstrated, differential diagnostic considerations should include tumor thrombus of renal and adrenal tumors, bland thrombus, or leiomyosarcoma of IVC. Although the differential diagnosis for IVL and intravenous LPL is lengthy, several factors can help make the diagnosis. Demonstration of a direct connection between the intravenous mass and the uterus or adnexa, and absence of renal or adrenal masses support the diagnosis. In addition, if a female patient is diagnosed as having right atrial myxoma (usually based on echocardiography), then IVL or intravenous LPL should also be considered given the relative rarity of myxoma originating in the right atrium. Therefore, histopathological evaluation must be performed to distinguish atrial myxoma from intravenous leiomyomatosis [18].

The histopathological differential diagnosis of IVL includes low-grade endometrial stromal sarcoma (LGESS) and leiomyosarcoma with vascular invasion. In contrast to IVL, LGESS typically exhibits endometrial involvement, as well as extensive extravascular myometrial permeation, in both gross and microscopic examination. In addition, vascular invasion in LGESS is usually not as conspicuous in gross examination as it is in IVL. An extension of LGESS into the inferior vena cava and right side of the heart has been reported only once [14]. In histological examination, LGESS contains a diffuse network of small arterioles and typically lacks the large thick-walled blood vessels characteristic of IVL. The neoplastic cells of ESS grow in a diffuse pattern and resemble the cells of a proliferative phase endometrium, with round to oval nuclei. In contrast, cellular IVL is characterized by fascicular arrangement of spindle cells with blunt-ended elongated nuclei and the presence of thick walled-blood vessels rather than the spiral arterioles typical of LGESS. Cellular IVL with unusual degrees of mitotic activity should be distinguished from leiomyosarcoma. Leiomyosarcoma almost never shows grossly visible vascular involvement and is characterized by tumor cell necrosis, severe cytological atypia, and high mitotic rates that usually exceed 10MF/10HPFs [14,16,18]. Intravenous lipoleiomyomatosis is distinguishable from the very rare uterine liposarcoma by the mitotic inactivity and bland nuclear features of the neoplastic cells, as well as the admixture of smooth muscle [14].

In conclusion, intravenous lipoleiomyomatosis is a rare condition, and in the absence of unusual histological features (such as high mitotic counts or necrosis of an uncertain type), its behavior is equivalent to an ordinary intravenous leiomyomatosis. In addition, IVL should be kept in mind regarding young women with cardiac symptoms, who have a right atrial mass and a pelvic mass, since the only clinical manifestation could be cardiovascular symptoms. Histopathologically, their resemblance to malignant uterine tumors, such as ESS and leiomyosarcoma, can make the diagnosis challenging. These neoplasms have different biological behaviors, so it is important to be able to distinguish them.

Please cite this article in press as: Ç. Vural, et al., Intravenous lipoleiomyomatosis of uterus with cardiac extension: A case report, Pathol. – Res. Pract. (2010), doi:10.1016/j.prp.2010.10.004
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