

Case Report

Diagnostic dilemma in massive cellular leiomyoma with cystic degeneration: a case report

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Abstract: Massive cellular leiomyoma with cystic degeneration is a rare and often misdiagnosed type of uterine leiomyoma. We report findings of such pathology in a 29-year-old asymptomatic, nulliparous Chinese woman. MRI revealed the 22.0 × 20.0 × 16.0 cm mass to be thick-walled and predominantly cystic. A diagnosis of ovarian tumor was initially suspected and a laparotomy was arranged. Her increscent uterus measured 20 × 18 × 16 cm. About 5000 ml of straw colored fluid was aspirated from the cystic cavity. The massive, cystic cavity arising on the left side of the uterus did not share a clear boundary with the normal myometrium. The patient underwent myomectomy and subsequent histopathological examination confirmed the diagnosis of a cellular leiomyoma with cystic degeneration.

Keywords: Cellular leiomyoma, cystic degeneration, diagnosis

Introduction

Uterine leiomyoma, which arises from uterine smooth muscle, is the most common benign gynecologic tumor. Typical cases of leiomyomata are commonly diagnosed in approximately 25% of reproductive-age women *via* imaging [1]. However, diagnostic dilemmas arise when leiomyomata are altered by degenerative changes. A growing number of leiomyoma cases clinically diagnosed as ovarian tumors have been reported [2-4]. In this study, we detail a case of an asymptomatic and unaware young woman who was diagnosed with a massive intramural leiomyoma.

Materials and methods

Case presentation

A 29-year-old Chinese woman, gravida 2, para 0, was referred to the gynecology department of Zhejiang Provincial People's Hospital to address her complaints regarding increased abdominal girth over the past three months. She had no other symptoms and her vital signs were all within normal limits. The patient under-

went menarche at age 13. She reported regular menstruation 30 days in duration, a menstrual period of seven days, menorrhagia and no dysmenorrhea. Prior medical and surgical history was unremarkable.

Abdominal examination revealed a large pelvi-abdominal mass extending above the xiphisternum (**Figure 1**). Subsequent gynecological examination revealed a large mass in the left pelvic region. The external genitalia, cervix and fornices of the vagina appeared normal. It was difficult to discern the uterus from the mass. There was no palpable lymphadenopathy and the patient had no abdominal tenderness.

Abdominal ultrasonography revealed a large cystic mass occupying the abdominal and pelvic cavities. A magnetic resonance imaging (MRI) scan revealed the mass to be predominantly cystic and possessing thick walls. The mass measured 22.0 × 20.0 × 16.0 cm in anteroposterior, transverse and craniocaudal dimensions (**Figure 2**). As there were no signs of ascites, the mass was initially suspected to be an adnexal tumor. Routine laboratory test

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Figure 1. Abdominal examination revealed a huge pelvi-abdominal extended above the xiphisternum.

results were normal, apart from the hemoglobin level, which was 72 g/L. Various tumor markers were within normal limits.

Procedure

A laparotomy was arranged for both diagnostic and therapeutic purposes. A vertical, midline incision was made from the umbilicus to the pubic symphysis. The patient's increscent uterus measured 20 × 18 × 16 cm in size. Approximately 5000 ml of straw-colored fluid was aspirated from the cystic cavity during the operation. A massive, cystic cavity arising from left side of the uterus was observed to lack a distinct boundary with the myometrium. The uterine cavity and surrounding structures (ovaries, fallopian tubes, bladder, intestines, liver and diaphragm) were observed to be normal on probe imaging. Para aortic lymph nodes were not palpable. Tissue from the cystic cavity wall was biopsied and subsequent frozen section examination revealed a diagnosis of cellular leiomyoma (CL). Myomectomy was then performed, separating the mass along its boundaries with normal myometrial tissue. After re-establishing homeostasis, the abdominal cavity was closed in layers (**Figure 3**).

Further histopathologic analysis confirmed the diagnosis of CL with cystic degeneration. Immunohistochemistry revealed positivity to SMA, Bcl-2, ER, PR, WT1, and Ki67 (+3%) (**Figure 4**). The patient's postoperative course was uneventful and she was discharged on the third postoperative day. She remained disease-free as determined both clinically and on imaging examinations after a follow-up period of more than six months.

Discussion

Uterine leiomyomata can undergo degeneration, which is detectable in approximately 65% of cases: hyaline degeneration (63%), myxoid changes (19%), calcification (8%), cystic changes (4%), fatty metamorphosis (3%), and red degeneration (3%) [2, 5]. In the present case, cystic degeneration with clear and yellow fluids was noted, similar to other previous reports [6-8]. Most patients with leiomyomata are asymptomatic and are diagnosed incidentally, but among symptomatic patients, pelvic pain, palpable masses or abnormal bleeding are the most common presenting symptoms. The asymptomatic patient in our report underwent laparotomy with a preoperative diagnosis of an adnexal mass, initially considered to be an ovarian tumor. Although MRI is a superior modality for evaluating patients with pelvic masses, our MRI findings seemed consistent with those of an ovarian tumor. With cystic degeneration, alteration of the normal leiomyoma architecture undoubtedly increases the difficulties in definitive and differential diagnosis [9]. It is challenging to distinguish between large leiomyomata with cystic degeneration and ovarian cysts.

Uterine leiomyomata are well known for their benign variants, including cellular, mitotically active and atypical leiomyomata. CL is a markedly cellular variant of leiomyoma, which itself represents a subgroup of variants as defined by the World Health Organization. CL is rare and accounts for <5% of leiomyomata [10-12]. CL often presents with an irregular border and marked cellularity (an increased number of cells per unit area compared with surrounding myometrium). CL can be recognized based on its characteristic morphologic features, which include the presence of (1) fascicular areas characteristic of smooth muscle neoplasia (at least focally, in most cases), (2) large thick-walled blood vessels, and (3) cleft like spaces [13]. In our report, the cellular leiomyoma found intraoperatively was a thick-walled cystic cavity sharing an unclear boundary with the normal myometrium. All these findings typified substantiated the characteristic features of CL.

Immunohistochemistry is helpful in the differential diagnosis of CL from other malignant smooth muscle tumors [14]. CL exhibits positivity for markers of smooth muscle differentia-

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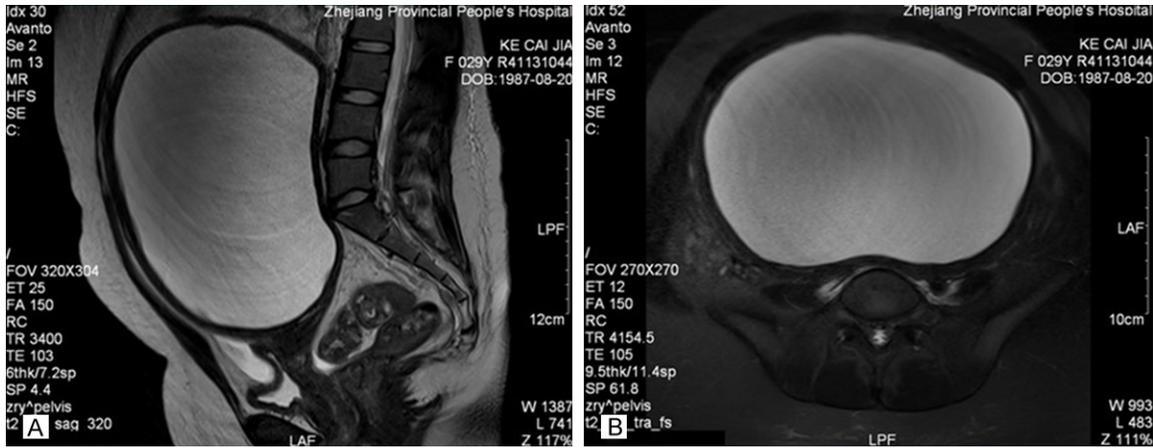


Figure 2. Magnetic resonance imaging (MRI) scan showed the mass of predominantly cystic with thick cyst wall and it measured 22.0 × 20.0 × 16.0 cm in anteroposterior, transverse and craniocaudal dimensions (A: Vertical plane; B: Coronal plane).

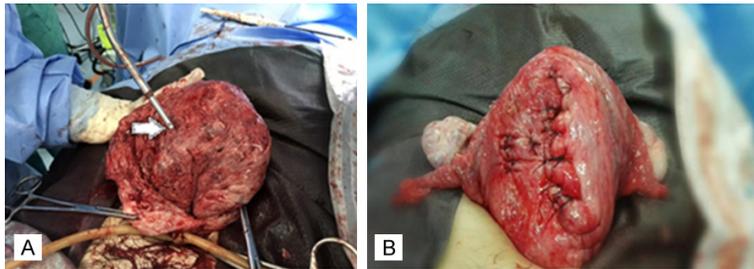


Figure 3. Intraoperative findings revealed a huge and cystic cavity from left side of uterus with unclear boundary of the normal myometrium (A). The uterine cavity is normal (behind the arrow). After Myomectomy, postoperative findings revealed the uterus, ovarian and fallopian tube (B).

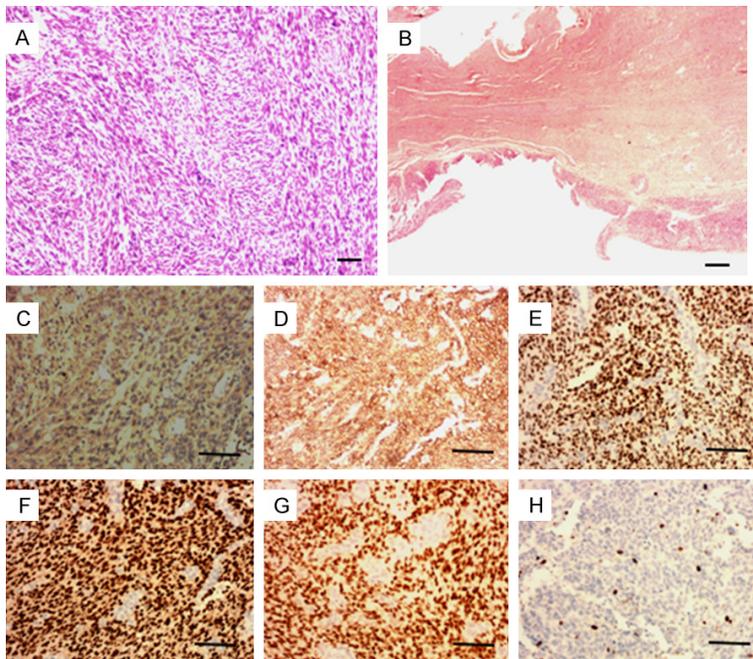


Figure 4. Light microscopy of cellular leiomyoma (A: hematoxylin and eosin staining 10×), Areas of cystic degeneration were seen (B: hematoxylin and eosin staining 2×), Immunohistochemically, SMA was positive (C: 20×), bcl-2 was positive (D: 20×), ER was positive (E: 20×), PR was positive (F: 20×), WT-1 was positive (G: 20×), only 3% of the cells were Ki-67 positive (H: 20×). Bar=100 μm (A), 20 μm (B), 200 μm (C-H).

tion (SMA), thus distinguishing it from malignant smooth muscle tumors [13]. Analysis of our sample indeed revealed positivity to SMA, indicating that the mass derived from the myometrium. Bcl-2 has been reported to be strongly expressed in benign leiomyomata as well as playing a good prognostic role [15]. Our case report also revealed similar findings, as bcl-2 was positive. Similarly, the PR receptor marker may be useful marker in distinguishing malignant smooth muscle tumors from benign growths when histological features are ambiguous. Our analysis revealed strong positivity for both PR and ER receptors, further sup-

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porting the diagnosis of benign uterine leiomyoma [16]. Uterine leiomyomata positive for WT1 confer an excellent prognosis, whereas typical leiomyosarcomas usually remain WT1-negative and variably exhibit a malignant course [17]. WT-1 positivity was also noted in our analysis. Mayerhofer K et al reported that the Ki-67 antigen was expressed at significantly higher levels in leiomyosarcomas. This marker is therefore also helpful in differentiating leiomyomata from malignant smooth muscle tumors. Ki-67 proliferation in our case report was found to be very low (3%), implying negligible mitotic activity. Necrosis and nuclear atypia were observed. Our findings correlated well with those of benign smooth muscle tumors [18]. All aforementioned marker positivity suggested that the mass derived from uterine smooth muscles and represented evidence of a benign intrauterine tumor conferring a good prognosis.

Massive CL with cystic degeneration may present difficulties in diagnostic differentiation from ovarian tumors, even on MRI. The combination of surgical management and histopathological examination, as effective adjuvant methods, however, will improve the diagnostic accuracy of atypical uterine leiomyomata.

Disclosure of conflict of interest

None.

Abbreviations

CL, cellular leiomyoma; MRI, magnetic resonance imaging; bcl-2, B-cell lymphoma 2; ER, Estrogen; PR, Progesterone; SMA, Smooth muscle actin; Ki-67, Antigen identified by monoclonal antibody Ki-67; WT-1, Wilms tumor protein.

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