Uterine multiple leiomyomas complicated by extensive mucoid degeneration: case report

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Summary

Uterine leiomyomas are the most common form of gynaecological tumours, and are exclusively benign. Only a few are associated with sarcomatous change. It is therefore important for the radiologist to be familiar with their range of appearances on magnetic resonance imaging (MRI) scans to distinguish them from other significant uterine pathologies, such as ovarian neoplasms, that require different management strategies. Here, the authors present the case of a 37-year-old Han woman, gravida 2, para 1 (cesarian section in 1996), who presented with a two-month history of lumbosacral swelling and pain. Physical examination revealed a pelvic mass and she was admitted with the presumptive diagnosis of an ovarian neoplasm. Laparotomy revealed multiple degenerated neoplasms that were benign in appearance, which was pathologically confirmed. A literature review was conducted to explore the natural history of uterine leiomyomas and their underlying etiopathogenesis. The optimal imaging modalities are also defined in the report, which enable the correct preoperative diagnosis to be made in order to optimize the care of women by multiple uterine leiomyomas.

Key words: Uterine leiomyomas; Gynaecological tumour; Aetiopathogenesis.

Introduction

Uterine leiomyomas are common, benign smooth muscle tumours of the uterus. They are found in nearly half of women over the age of 40 years and infrequently cause complications. Uterine leiomyomas, also colloquially known as fibroids, tend to grow under the influence of estrogen, and regress when estrogen levels are reduced. Thus, growth frequently occurs during pregnancy, followed by regression after delivery. Most uterine fibroids are asymptomatic, but some women develop heavy menstrual flow (menorrhagia), which often cause anemia, bleeding between periods, pain, infertility or subinfertility, pelvic pressure, stress urinary incontinence, and ureteral obstruction. The diagnosis of uterine leiomyoma is usually based on the clinical findings of an enlarged, irregularly shaped, firm uterus, which may or may not be tender. Sometimes the diagnosis is unclear, and diagnostic tests are used to delineate fibroids and exclude other problems. Diagnostic techniques include ultrasound, magnetic resonance imaging (MRI), and computed tomography (CT) scanning, laparoscopy, and histological examination.

A variety of degenerative changes may occur in leiomyomas. The larger the leiomyoma, the more likely it will be that a degenerative component will be present. Several mechanisms are likely to contribute to this phenomenon, including ischemia and hormonal effects. More than one pattern of degeneration may be observed in the same leiomyoma. These changes include hyaline, cystic, red, calcific, and fatty degenerations. The most common of these is hyaline degeneration, whereby expanded septa lose their fibrillary structures and assume a uniform, pale, eosinophilic, translucent appearance resembling ground-glass. Degenerative changes may be localized or affect extensive areas of the tumour, and occasionally even its entirety. Surviving muscle cells may orient themselves into lacework patterns that accompany degenerative changes in leiomyomas.

The terms mucoid and myxoid degeneration are used to describe changes that are similar to hyaline changes, with or without cystic formation. In mucoid degeneration, the matrix typically appears to be mucinous in nature. There is no difference in practical terms between mucoid and myxoid forms of degeneration, thus the two terms are often used interchangeably. However, extensive mucoid degeneration is rare among these changes. Here, the authors report a case of uterine multiple leiomyomas that were complicated by mucoid degeneration.

Case Report

A 37-year-old Han woman, gravida 2, para 1, with a history of one cesarean section in 1996, was admitted after presenting with lumbosacral swelling and pain for more than two months. These symptoms had worsened over the previous week. Gynaecological examination revealed a non-tender anteverted uterus, which was enlarged to the size of a two-month pregnancy, with moderate texture and mobility. A mass that was four cm in diameter was discovered in the left adnexal area, and another mass, six cm in diameter, was discovered in the right adnexal area. The two masses were moderate in texture with no clear borders but had infiltrated the uterus.

B-mode ultrasound examination revealed an anteverted uterus that measured 6.4 × 4.6 × 5.0 cm, with a regular morphology. An intrauterine device was observed in the correct location. One enhanced echo image showed a mass that measured 4.8 × 3.1 cm in the left adnexal area, without a clear border separating it from the left ovary. One cystic dark area, measuring 6.0 × 4.6 cm, was detected in the right adnexal region. The wall of this cyst appeared

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Fig. 1

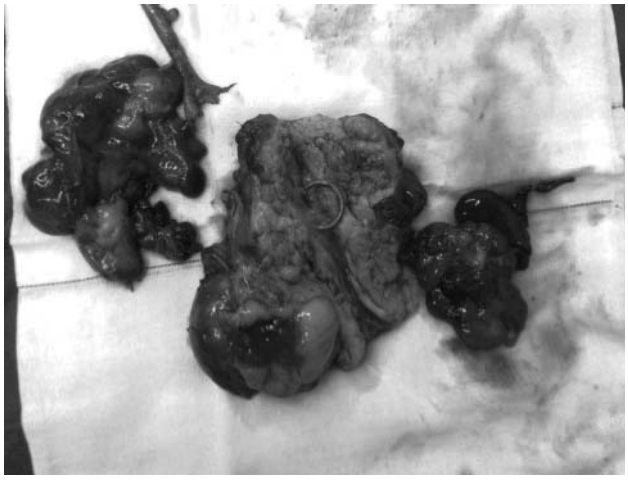

Fig. 2

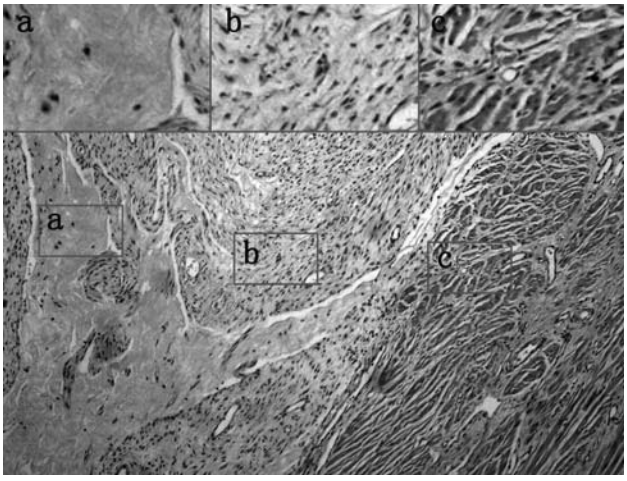

Figure 1. — Macroscopic appearance of the lesion showing the longitudinal section of the uterus. The tissue samples show scattered intramural myoma tissue without a capsule, as well as myomas in the adnexa bilaterally, and the broad ligament.
Figure 2. — Microscopic features of the degenerated leiomyoma: a) mucoid degeneration; b) myoma tissue; and c) normal myometrial tissue. There is a clear boundary between the tumour cells and the normal myometrial tissue. The spindle-shaped tumour cells are arranged in a staggered pattern, and the mucoid degeneration is obvious. The atypia of the tumour cells is not remarkable.

to be thick and ill-defined, and a dense reflection to the right could be detected from the cystic wall. A fluid dark area, measuring 1.1 cm in diameter, was observed in the pouch of Douglas. The results of the following examinations and tests were within the normal range: routine blood tests; coagulation function; hepatic and renal functions; blood biochemistry; blood glucose; carcinoma antigen 125 (CA 125); alpha fetal protein (AFP); carcinoma embryonic antigen (CEA); and electrocardiography. No positive findings were detected with an ultrasound examination of the liver, gallbladder, pancreas or spleen. A barium enema check also yielded negative results.

The patient was admitted to hospital with the presumptive diagnosis of an ovarian neoplasm in November 2009. A laparotomy was performed under general anaesthesia. At laparotomy, the uterus was found to be slightly enlarged, and two soft masses, measuring three cm in diameter, were found in the bilateral uterine horns (Figure 1). There were multiple dark red bubbles, ranging in size from one to three cm, attached to the anterior uterine wall. A four-cm cyst was found in the left mesosalpinx, and another six-cm cyst was found in the right mesosalpinx. Both were soft in texture and were composed of multiple cysts with gelatinous contents. Similar neoplasms were also detected in the bilateral broad ligaments of the uterus. The neoplasm in the left mesosalpinx was stripped and sent for frozen section examination. The findings suggested the diagnosis of a benign soft tissue neoplasm. Both ovaries appeared to be normal in both morphology and size.

When the masses from the uterine horns were incised, soft and gelatinous tissue with indistinct boundaries was observed. Pathological examination suggested the diagnosis of an endometrial mesenchymal neoplasm, which was likely to be benign (Figure 2). Consequently, a total hysterectomy and bilateral salpingectomy were performed, and the neoplasms in the bilateral broad ligaments were stripped using blunt dissection. The features of these neoplasms were the same as those from the mesosalpinx. There was no obvious abnormality in the endometrium upon dissection. Gelatinous tissue with no observable envelope was scattered within the muscular layer of the uterus. Examination of the pathology of paraffin-embedded sections yielded a diagnosis of multiple uterine leiomyomas with conspicuous mucoid degeneration. The patient was discharged from hospital after rehabilitation. No recurrence was observed after a 12-month follow-up period.

Discussion

Molecular biologists have begun to probe the etiology of uterine leiomyomas, exploiting DNA methylation differences between polymorphic loci on both active and inactive X chromosomes to confirm that each leiomyoma is derived from a single transformation event [1]. Most importantly, these studies also suggest that each tumour is a distinct clone, which reinforces the notion that smooth muscle tumourigenesis is exceedingly common. The genetic mechanisms that initiate and promote the growth of leiomyomas must occur frequently, but are not fully understood. However, cytogenetic analysis of these benign smooth muscle tumours has already revealed some important clues. Almost half of leiomyomas have chromosomal rearrangements that are large enough to be seen in G-banded karyotypes. These chromosomal rearrangements are generally simple, which is in sharp contrast to the aberrations seen in leiomyosarcomas. To date, recurrent aberrations have allowed the definition of seven cytogenetic subgroups: t(12;14)(q14-15;q23-24); del(7)(q22q32); rearrangements of 6p21 and 10q22; trisomy 12; and deletions of 3q and 1p. Of these, the translocation between chromosomes 12 and 14 and the rearrangements involving chromosome 6 are perhaps best understood. Both rearrangements involve genes for two closely related non-histone chromatin proteins: *HMGA1* at 6p21 and *HMGA2* at 12q15 [2, 3]. There are few reports that describe the various mechanisms by which uterine leiomyomas degenerate.

In the present patient, the degeneration of multiple uterine leiomyomas led to the formation of variously-sized,

hollow masses filled with neoplastic tissue, which were liquefied due to the lack of a blood supply. The mucoid contents in the cavities modified the texture of the leiomyomas to soft masses. Given the soft texture of the tissue, malignant ovarian tumour and/or malignant uterine tumours had to be considered in the differential diagnosis. Intraoperative pathologic diagnosis was very helpful in the differential diagnosis.

Degenerated leiomyomas, especially those with larger volumes, often bring difficulties in differential diagnosis and corresponding clinical decision-making. A MRI scan is the most accurate technique for detecting and localizing leiomyomas. Degenerated leiomyomas have variable appearances on T2-weighted images and contrast-enhanced images. The common types of degeneration are hyaline (> 60% of cases), cystic (approximately four percent), myxoid, and red. Edema is not a phenomenon of degeneration, but is a common histopathological finding (approximately 50% of cases). Hemorrhage, necrosis, and calcification (approximately four percent of cases) may also be observed. Specific types of unusual leiomyomas include lipoleiomyoma and myxoid leiomyoma, which may have MRI features that are sufficiently characteristic to allow differentiation from other gynaecological and non-gynaecological diseases. Intravenous leiomyomatosis, metastatic leiomyoma, diffuse leiomyomatosis, and peritoneal disseminated leiomyomatosis represent unusual growth patterns. Other unusual growth patterns are retroperitoneal growth, parasitic growth, and a pattern that may occur in cervical leiomyoma [4].

On T2-weighted MRI images, non-degenerated leiomyomas appear as well-circumscribed masses of decreased signal intensity; however, cellular leiomyomas can have relatively higher signal intensities on T2-weighted images and demonstrate enhancement on contrast material-enhanced images. The differential diagnosis of leiomyomas includes adenomyosis, solid adnexal mass, focal myometrial contraction, and uterine leiomyosarcoma [5]. For patients who are symptomatic, medical or surgical treatment may be indicated. MRI also has a role in treatment of leiomyomas by assisting in surgical planning and monitoring response to medical therapy. The use of 18F-FDG positron emission tomography/CT (PET/CT) may also play a role in the diagnosis of uterine leiomyoma and can sometimes be helpful in the evaluation of related degeneration [6].

As leiomyomas are the most common gynaecological tumours, and are almost exclusively benign, it is important to be familiar with the variety of MRI appearances of uterine leiomyomas in order to distinguish them from other significant diseases.

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