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Hypovolemic Shock and Hemoperitoneum from Spontaneous Avulsion of a Large Pedunculated Uterine Leiomyoma

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ABSTRACT

Hemoperitoneum with hypovolemic shock from avulsion of a pedunculated leiomyoma is a rare but highly fatal condition that can occur spontaneously or as a result of trauma. We report a case of hemoperitoneum and hypovolemic shock secondary to a bleeding leiomyoma detected via computed tomography (CT) scan in a 39 year old premenopausal, gravida 0 female that presented with abdominal pain and became hemodynamically unstable in the emergency department. A preoperative bimanual exam revealed a mass consistent with a 20 week gestational uterus. Following fluid resuscitation, the patient underwent emergent myomectomy and ligation of the right uterine artery and was discharged home in good condition.

CASE REPORT

CASE REPORT

A 39 year old premenopausal, gravida 0 Caucasian female with a past medical history of a myomatous uterus presented to the emergency room with sudden onset abdominal cramping followed by syncope. The abdominal physical exam was positive for mild tenderness in the right upper quadrant and vital signs on admission were normal. The pelvic exam was positive for a pelvic mass. A urine pregnancy test was negative and serum hemoglobin level was 12.5 g/dL (12.5 - 15.0 g/dL). The patient was admitted for observation and over several hours developed worsening abdominal pain. Upon re-examination, the physical exam was remarkable for a rigid abdomen and new tachycardia, hypotension and tachypnea. Repeat lab values revealed a drop in serum hemoglobin of 5 g/dL over 8 hours. An emergent bedside focused assessment with sonography for trauma (FAST) exam, performed by the emergency staff revealed fluid in the abdomen. The patient was immediately hemodynamically stabilized with fluid resuscitation.

An immediate CT scan of the abdomen and pelvis was obtained with intravenous contrast. There was a large amount of perihepatic free fluid measuring 30-40 Hounsfield units in the right paracolic gutter (Fig. 1). High density layering material compatible with blood products were noted in the dependent right paracolic gutter and superior to the bladder inferior to a heterogeneous mass(Fig. 1). There was a 10 cm heterogeneous mass contiguous with the right superior aspect of the uterine fundus (Fig. 1-2). At the inferior aspect of the mass was an arterial blush arising from a right uterine artery (Fig. 1-2). A prior ultrasound (US) from 2 years ago which was unavailable at the time of interpretation showed an 8 cm pedunculated right fundal uterine leiomyoma (Fig 3a) with a vascular pedicle (Fig. 3b).

A preoperative bimanual exam revealed a mass consistent with a 20 week gestational uterus. The patient underwent emergent exploratory laparotomy, myomectomy and ligation of a right uterine artery. Intraoperative findings revealed an actively bleeding subserosal mass (Fig. 4) and 1500 mL of hemoperitoneum. A fertility sparing procedure was chosen

since the patient was a young, gravida 0 female and the uterus had a normal intra-operative appearance. The patient tolerated the procedure well and was transfused one unit of packed red blood cells at the beginning of the procedure due to pre-operative anemia and hypotension. Following the surgery the patient was transferred to the recovery unit in stable condition. The patient had an uncomplicated postoperative course and was discharged to home two days after the surgery in good condition.

Macroscopic and histologic examination of the surgical specimens revealed the mass to be a uterine leiomyoma.

DISCUSSION

In 1949 Hasskarl collected 58 cases of spontaneous bleeding occurring from dilated veins over the surface of leiomyomata[1]. Three cases of spontaneous avulsion of leiomyomata causing intra-abdominal hemorrhage have been reported in the literature. In 1933 Shelley reported a case of a detached fibroid in the abdomen with intra-abdominal hemorrhage seen on laparotomy in a patient that presented with acute onset abdominal pain[1]. In 1935 Cattolorda presented a case of a detached fibroid in the abdomen adherent to a gangrenous appendix and intra-abdominal hemorrhage[1]. In 2005 Dasari reported a case of spontaneous intra-leiomyoma hemorrhage, perforation and hemoperitoneum in a postmenopausal woman[2]. In this case report the diagnosis of spontaneous avulsion was made preoperatively which facilitated and expedited surgical management. However, this case is unique in that there are very few reported cases of spontaneous avulsion of leiomyoma. In addition, the arterial rupture of the vascular pedicle has not been reported.

Etiology & Demographics:

Uterine leiomyomata (commonly referred to as fibroids) are the most common pelvic tumors affecting females of reproductive age [3-6]. They are benign tumors arising from smooth muscle cells of the myometrium [6]. The cumulative incidence by age 50 is greater than 80% in black women and approximately 70% for white women [7]. Risk factors for developing leiomyomata are black race, increased estrogen exposure, obesity, alcohol consumption, Vitamin D deficiency, diets high in red meat, hypertension, and underlying genetic predispositions [7-14].

The typical appearance of uterine leiomyomata on radiographs can be coarse or popcorn calcifications. On CT they can appear as soft tissue masses that can have calcifications and show heterogeneous enhancement. On US they may appear as solid masses with a whorled appearance with similar echogenicity to the myometrium and with color Doppler flow. MRI will show low to intermediate signal (compared to myometrium) on T1 weighted sequences and low signal on T2 weighted sequences with variable enhancement[15]. Uterine leiomyoma usually show mild Fluorodeoxyglucose (FDG) uptake on Positron Emission Tomography (PET), but sometimes can show intense uptake [16].

Spontaneous hemoperitoneum due to leiomyoma rupture is a rare entity with less than 100 cases reported in the literature [3-4]. Massive bleeding leading to hypovolemic shock is extremely rare [17]. Larger leiomyomata usually degenerate as they outgrow their blood supply [2]. Previous cases have shown hemoperitoneum related to leiomyomata is likely due to veins coursing over the surface of leiomyomata with resultant hypovolemic shock [1,17]. This case is unique in that there was rupture of the arterial supply of the vascular pedicle seen on imaging and confirmed surgically.

Clinical & Imaging findings:

Patients with acute torsion of a uterine leiomyoma with hemoperitoneum will present with acute onset severe abdominal pain. Depending on the degree of hemoperitoneum, the patient may be tachycardic and hypotensive with a rigid and tender abdomen on physical exam. Serum laboratory values will reveal a drop in hemoglobin in the presence of hemoperitoneum. In hemodynamically unstable patients fluid resuscitation and a FAST exam is done. In the case presented the patient had free fluid on the FAST exam and was hemodynamically stabilized with fluid resuscitation. Since the patient was hemodynamically stabilized, an emergent CT was obtained to elucidate the cause of hemoperitoneum and for presurgical planning.

The superior modality for the evaluation of leiomyomata is US; however findings may be observed on CT in the evaluation of an acute abdomen. On US acute torsion of a subserosal leiomyoma can appear heterogeneous and hypoechoic with areas of cystic change. Free fluid will be seen in the presence of hemoperitoneum. Color Doppler shows peripheral vascularity and/or absence of color flow within the leiomyoma. On CT the appearance can vary with a heterogeneously enhancing mass [5]. On magnetic resonance imaging (MRI), the imaging appearance can vary depending on the degree of torsion and stage of blood products; often with heterogeneous T1 and T2 signal and variable enhancement. In our case an MRI was not obtained due to the adequacy of CT to confirm the diagnosis, time sensitivity of the patient's presenting condition and time intensive nature of MRI.

Differential Diagnosis:

The common causes of acute abdomen related to a myomatous uterus include torsion of a subserosal leiomyoma, torsion of a myomatous uterus, red degeneration of a leiomyoma and complications of uterine leiomyosarcoma[2].

Uterine torsion may present with gas in the uterine cavity on CT or radiographs[18]. Ultrasound can show a myomatous uterus with heterogeneous echogenicity and a lack of Color Doppler flow. On CT, twisting of the cervix can be observed as a whorled appearance with a large pelvic mass. After

contrast administration, areas of hypodensity and lack of enhancement of the uterus and/or mass can be observed [19]. On MRI uterine torsion can appear as loss of the normal H configuration of the upper vagina with resultant formation of an X configuration [20].

Red or carneous degeneration is a type of hemorrhagic infarction thought to occur secondary to thrombosed veins within the periphery of the tumor or rupture of intratumoral arteries [4]. On ultrasound degenerated leiomyomata can appear heterogeneous with areas of cystic change. On CT degenerated leiomyomata may appear complex with areas of fluid attenuation. Calcification confined to the periphery is thought to be secondary to thrombosed veins [17]. On T1 MRI degenerated leiomyomata can have a peripheral rim of high signal intensity from shortening effects of protein and/or blood products. The T2 appearance is variable with or without a low signal rim. Degenerating leiomyomata can have variable enhancement with minimal central enhancement or irregular patterns [17].

Uterine leiomyosarcoma is a rare soft tissue neoplasm occurring in 1% of uterine malignancies. Rarely, uterine leiomyosarcoma can present with spontaneous perforation and/or hemoperitoneum [21]. On imaging uterine leiomyosarcoma can appear identical to a leiomyoma with similar imaging features but will grow at a rapid rate on follow up imaging. On CT they can be heterogeneous with central areas of hypoattenuation representing necrosis [22]. On T1 MRI they are large heterogeneous masses that are isointense to muscle with areas of hemorrhagic necrosis that can appear as pockets of increased signal on T1 imaging. On T2, a background of intermediate signal intensity with pockets of high signal intensity can be seen. Areas of high T1 and high T2 signal correspond to regions of hemorrhagic necrosis and areas of low T1 signal and high T2 signal correspond to cystic necrosis [23].

In the case presented, contrast enhanced CT was chosen because CT is a rapid and sensitive exam for pelvic and abdominal etiologies of hemoperitoneum and an acute abdomen. MRI was deferred due to its time intensive nature. In the case presented the CT showed a large pedunculated heterogeneous mass extending from the uterine fundus with rupture of a vascular pedicle and contrast extravasation from the right uterine artery. The uterus appeared normal in configuration and did not contain intrauterine gas. There were no discrete cystic areas in the mass on CT to suggest degeneration or necrosis.

Treatment & Prognosis:

Hemoperitoneum with CT findings suggestive of acute arterial extravasation is an ominous sign requiring immediate intervention. If free fluid is seen in the abdomen on FAST exam, immediate surgical management is indicated. If left untreated hypovolemic shock can lead to death. Definitive management of a torsed leiomyoma in the presence of hemoperitoneum of a hemodynamically unstable patient requires emergent surgical intervention. Otherwise, approximately 80% of leiomyomata are asymptomatic and require no treatment [24]. For symptomatic patients treatment options include hysterectomy, myomectomy, myolysis, Gonadotropin releasing hormone analogs, and uterine artery embolization [6].

TEACHING POINT

Imaging findings of hemoperitoneum with active extravasation from a vascular pedicle of a leiomyoma is an uncommon cause of an acute abdomen but with fatal consequences. Early recognition on imaging could have life saving implications; this scenario should prompt immediate resuscitation and emergent surgical management.

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FIGURES

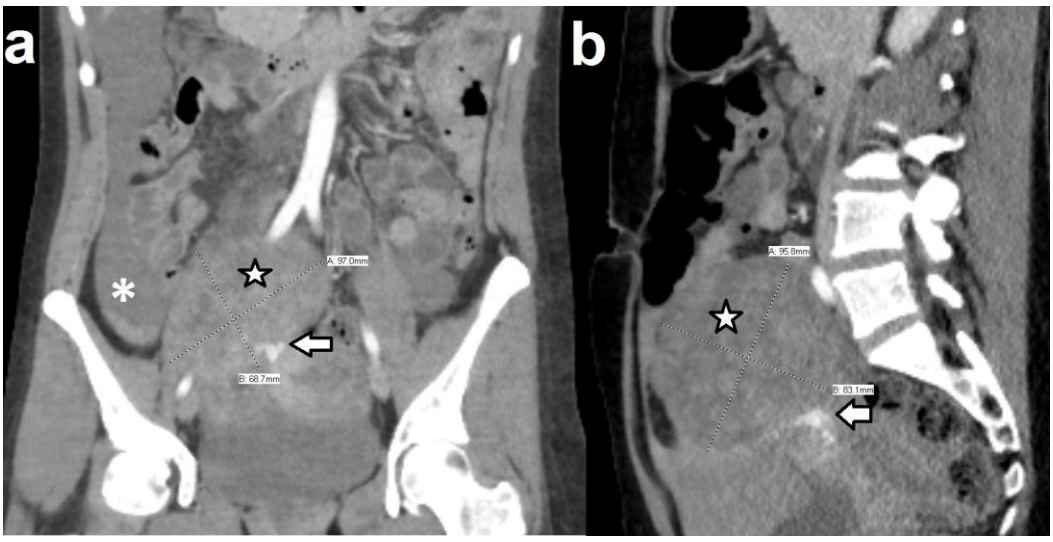

Figure 1: 39 year old female with acute arterial extravasation from spontaneous avulsion of a large pedunculated uterine leiomyoma. FINDINGS: a. Coronal contrast enhanced CT of the abdomen and pelvis in the late arterial phase shows a 9.7 x 6.9 cm heterogeneous mass (star) in the right aspect of the uterine fundus with an arterial blush at the base arising from a branch of the uterine artery (arrow). Layering high density fluid in the inferior right paracolic gutter (asterisk). b. Sagittal contrast enhanced CT of the abdomen and pelvis in the late arterial phase shows a heterogeneous mass (star) in the right aspect of the uterine fundus with an arterial blush at the base arising from a branch of the uterine artery (arrow). There are layering high density blood products superior to the bladder.
TECHNIQUE: Philips Brilliance 64 slice CT scanner, Coronal and sagittal reformatted CT, 120 kVp, 104 mAs, 5 mm slice thickness, 95 mL Isovue 370. CTDIvol=6.85 (mGy) and DLP = 467.4 (mGy*cm) with a 32 cm phantom.

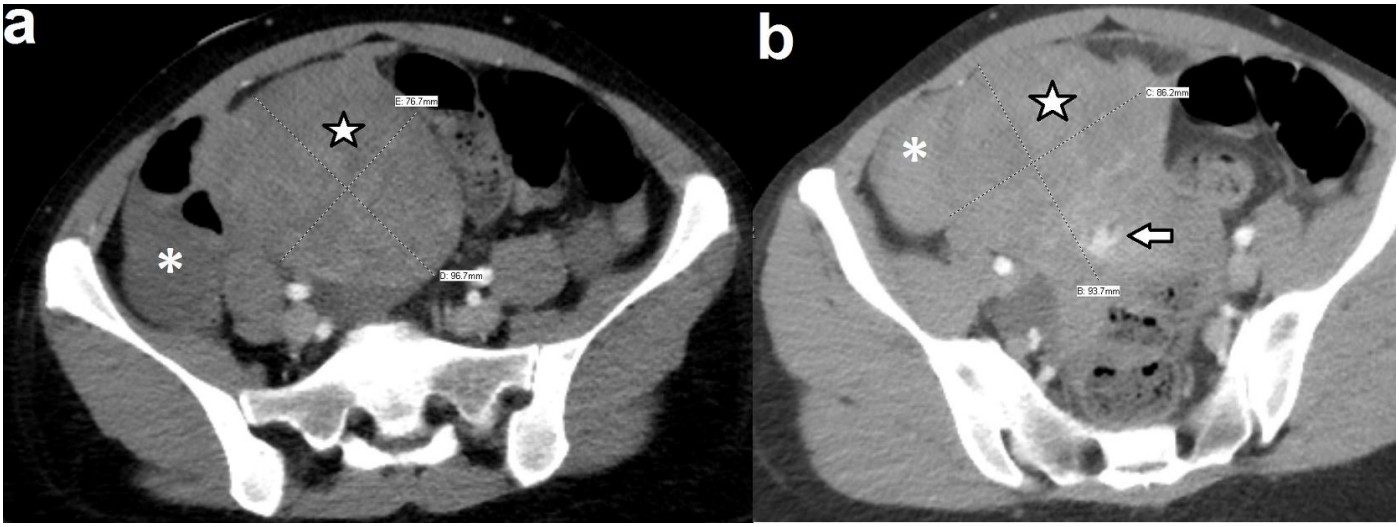

Figure 2: 39 year old female with acute arterial extravasation from spontaneous avulsion of a large pedunculated uterine leiomyoma.
FINDINGS: a. Axial contrast enhanced CT of the abdomen and pelvis in the late arterial phase shows a 9.7 x 7.8 cm heterogeneous mass (star) in the right pelvis and high density material in the inferior right paracolic gutter (asterisk). b. Axial contrast enhanced CT of the abdomen and pelvis in the late arterial phase shows a 9.4 x 8.6 cm heterogeneous mass in the right aspect of the uterine fundus with an arterial blush at the base arising from a branch of the uterine artery (arrow). High density layering blood products in the right paracolic gutter (asterisk).
TECHNIQUE: Philips Brilliance 64 slice CT scanner, Axial CT, 120 kVp, 104 mAs, 5 mm slice thickness, 95 mL Isovue 370. CTDIvol=6.85 (mGy) and DLP = 467.4 (mGy*cm) with a 32 cm phantom.

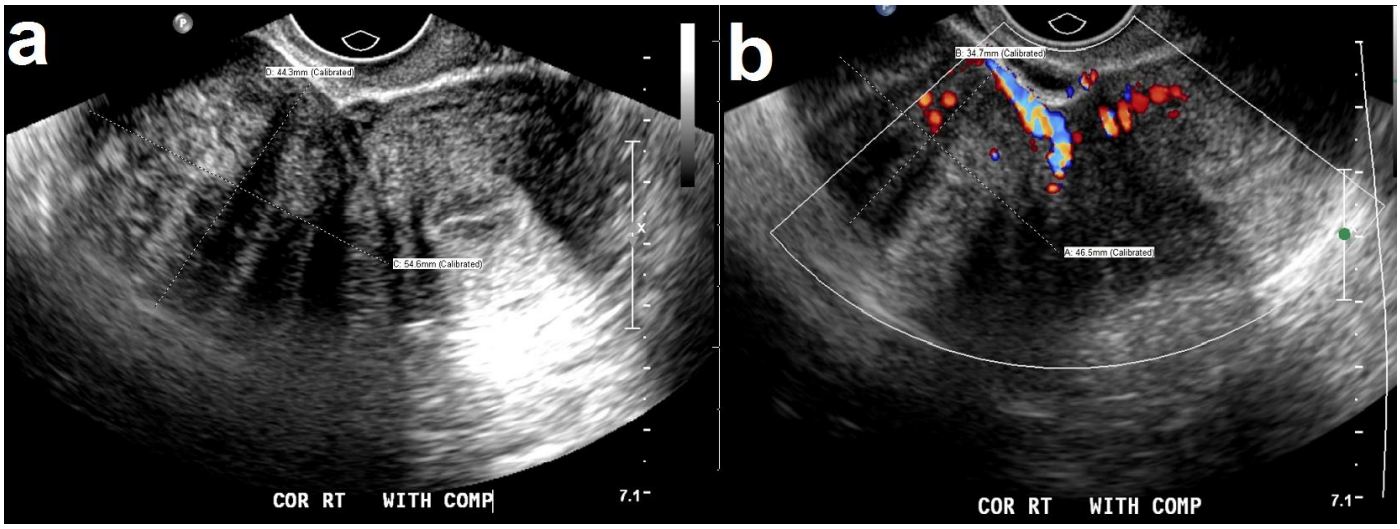

Figure 3: 37 year old female with a large pedunculated uterine leiomyoma extending from the right uterine fundus into the right adnexa. Sonographic imaging from two years prior in the same patient for pelvic pain.
FINDINGS: a. Coronal grayscale transvaginal ultrasound of the pelvis with an 8-5MHz 10 cm endocavitary probe with compression shows a subserosal heterogeneous mass that is predominantly isoechoic to myometrium in the right adnexal region. b. Coronal transvaginal Doppler ultrasound of the pelvis with an 8-5MHz 10 cm endocavitary probe with compression shows a subserosal heterogeneous mass predominantly isoechoic to myometrium in the right adnexal region with Color Doppler flow extending from the uterine fundus compatible with a vascular pedicle.
TECHNIQUE: Coronal grayscale and Color Doppler ultrasound, 8-5MHz 10 cm probe.

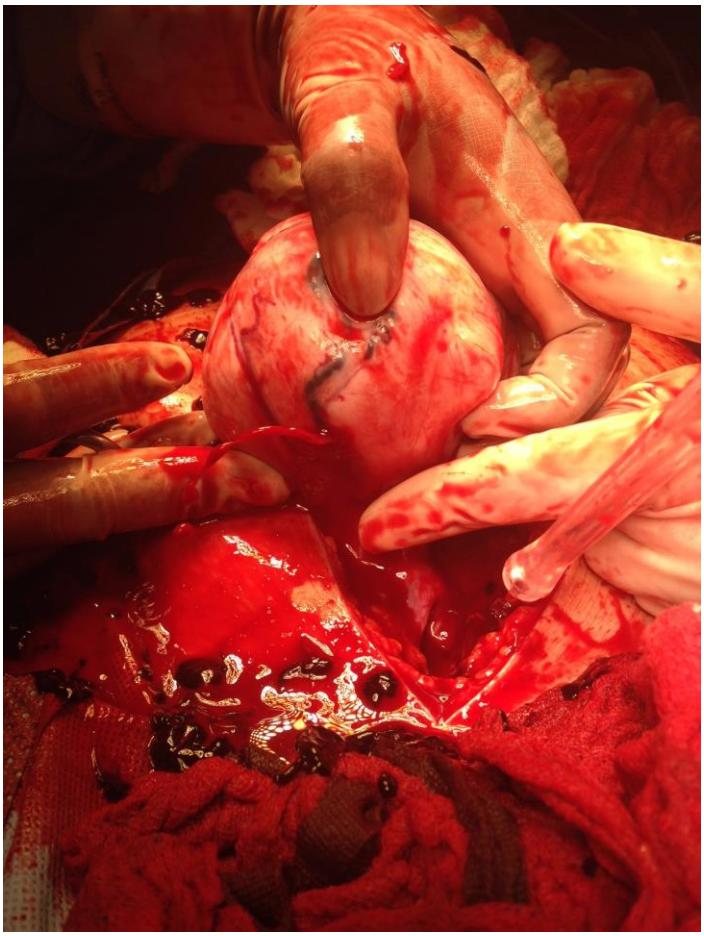

Figure 4: 39 year old female with acute arterial extravasation, hemoperitoneum and hypovolemic shock from spontaneous avulsion of a large pedunculated uterine leiomyoma. FINDINGS: Intraoperative photograph of the bleeding subserosal leiomyoma as seen on the CT scan.

Etiology	Benign tumors arising from smooth muscle cells of the myometrium[4].
Incidence	The most common pelvic tumor in women. Cumulative incidence by age 50 is greater than 80% in black women and approximately 70% for white women[5].
Gender Ratio	Exclusively females.
Age Predilection	Reproductive age[4]
Risk Factors	Black race, increased estrogen exposure, obesity, alcohol consumption, vitamin D deficiency, diet high in red meat, hypertension, genetic predisposition[5,9-15].
Treatment	Hysterectomy, myomectomy, myolysis, Gonadotropin releasing hormone analogs, and uterine artery embolization[4].
Prognosis	Excellent in the absence of spontaneous or traumatic rupture.
Findings on Imaging	*Radiographs:* Coarse or popcorn calcifications on radiographs. *CT:* Soft tissue density lesions with coarse peripheral or central calcifications. *US:* Predominantly hypoechoic but can appear isoechoic or hyperechoic. *MRI:* will show low to intermediate signal (compared to myometrium) on T1 weighted sequences and low signal on T2 weighted sequences with variable enhancement. *FDG PET:* mild or intense uptake

Table 1: Summary table for Uterine Leiomyoma.

	X-ray	US	CT	MRI	Pattern of Contrast Enhancement
Leiomyoma	Coarse or popcorn calcifications.	Predominantly hypoechoic but can appear isoechoic or hyperechoic.	Soft tissue density lesions with coarse peripheral or central calcifications.	T1: Low to intermediate signal (compared to myometrium) T2: Low signal	Variable.
Torsed Leiomyoma	None.	The leiomyoma will be heterogeneous with cystic change. Circumferential vascularity and/or absence of flow on Doppler.	Heterogeneous mass with reduced enhancement and hypodense areas.	Heterogeneous T1 and T2 signal depending on the stage of blood products and degree of torsion with variable enhancement.	Heterogeneous. If rupture of vascular pedicle, there may be acute arterial extravasation in the arterial phase on CT.
Uterine Torsion	Gas in the uterine cavity.	Heterogeneous echogenicity and absence of Color Doppler flow.	Gas in the uterine cavity. Whorled appearing cervix with a large pelvic mass.	X configuration of the upper vagina.	Variable. Lack of enhancement of the uterus and/or mass
Red Degeneration	Dense peripheral calcifications	Heterogeneous with hypoechoic foci representing cystic change.	Dense peripheral calcifications. Complex with areas of fluid attenuation.	T1: peripheral rim of high signal intensity. T2: appearance is variable with or without a low signal rim.	Variable enhancement with minimal central enhancement or irregular patterns of enhancement
Leiomyosarcoma	None	Predominantly hypoechoic but can appear isoechoic or hyperechoic. Grow rapidly in a short time interval.	Heterogeneous with central areas of hypoattenuation.	T1: heterogeneous masses isointense to muscle with areas of increased signal. T2: intermediate signal intensity with foci of high signal intensity.	Variable.

Table 2: Differential diagnosis table for torsed leiomyoma.

ABBREVIATIONS

CT = Computed Tomography
FAST = Focused assessment with sonography for trauma
FDG = Fluorodeoxyglucose
MRI = Magnetic Resonance Imaging
PET = Positron Emission Tomography
US = Ultrasound

KEYWORDS

Pedunculated leiomyoma; Hemoperitoneum; Spontaneous avulsion; Leiomyoma; Fibroid; Hypovolemic shock

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