CASE REPORT

Large, extra-abdominal leiomyoma of the round ligament with carneous degeneration

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SUMMARY

Round ligament tumours represent a rare entity that can present similarly to an incarcerated hernia. Basic understanding and appropriate preoperative management is imperative in order to differentiate between the two diagnoses. Leiomyoma is the most common type of round ligament tumour. It is associated with oestrogen exposure and is more common in the presence of uterine leiomyomas. Here we discuss a 68-year-old woman who presented with a palpable left inguinal mass that progressively grew in size, associated with pelvic pressure and discomfort. On surgical resection, the mass was found to be derived from the round ligament at the entrance of the external inguinal ring. Pathology confirmed a round ligament leiomyoma, measuring 25×9×8.5 cm. This case is the largest round ligament leiomyoma recorded to date and the first to exhibit carneous degeneration. A review of the current literature is also provided.

BACKGROUND

The round ligament of the uterus is an embryological remnant of the gubernaculum, originating from the uterine horns and exiting the pelvis via the deep inguinal ring. It passes through the inguinal canal and terminates in the labia majora where its fibres then spread and mix with the tissues of the mons pubis. Its composition is primarily made up

Figure 2 Sagittal section of an abdominal CT scan depicting communication between the abdominal wall mass and the uterus, noted by the red arrow.

of smooth muscle fibres and fibrous tissue, with blood supply from the branches of the inferior epigastric vessels. Tumours of the round ligament are exceedingly rare, with roughly 300 cases previously described in the literature; leiomyoma is the most commonly observed tumour type. These tumours have the ability to form at any point along the course of the round ligament and are further classified by intra-abdominal or extra-abdominal derivation.¹ Leiomyomas originating from the extra-abdominal round ligament are more commonly observed, likely due to the presence of a palpable mass within the inguinal canal or labium. They are typically unilateral, with a predilection for the right side and are associated with the presence of uterine leiomyomas.¹ Here we present a case of a large extra-abdominal leiomyoma of the round ligament that first presented as a left-sided inguinal mass, which progressively grew in size and eventually required complete surgical resection. A review of the current literature is included, which includes the pathogenesis, management and outcomes of this disease entity.

CASE PRESENTATION

The patient is a 68-year-old Dominican woman who initially presented to an outside hospital 30

Figure 1 Coronal section of the abdominal CT scan, depicting an abdominal wall mass measuring 27.75×9.41 cm at its largest dimensions.

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Figure 3 Inguinal mass, preincision, extending from the iliac crest to the mons pubis.

years ago with a chief complaint of sharp pain in the left lower quadrant of the abdomen. At that time, she was told she had a small inguinal hernia requiring no further treatment.

Thirty years later, the patient presented to another outside hospital after noticing the abdominal mass had rapidly increased in size over a 5-day period, associated with pain and overlying erythematous skin changes. A CT scan showed a large mass in the left external abdominal wall. The patient was subsequently taken to the operating room for a suspected strangulated inguinal hernia. On initial

Figure 4 Inguinal mass, circumferentially mobilised and denuded from surrounding structures, attached to the round ligament at the external inguinal ring.

Figure 5 Complete excision of inguinal mass, with a silk stitch denoting its attachment to the round ligament, measuring 25x9x8.5 cm.

exploration, the mass was without signs of strangulation, and an incisional biopsy was performed. The inner core was noted to be cystic and drained 1.3 L of serosanguinous fluid. Pathology revealed a leiomyoma with necrosis and degenerative changes. Postoperatively, the patient progressed well, however with persistent fevers that required intravenous antibiotics. The overlying skin changes resolved, and the patient was discharged after 6 days. The patient was then referred to surgical oncology for further management.

Her medical history includes chronic nephrolithiasis, status post right nephrectomy, and two caesarean sections due to a history of uterine leiomyomas. Menarche was at age 12, gravida 2, para 2 0 0 2, no history of oral contraceptive use and menopause occurred at age 50. Family history is pertinent for a daughter with clear cell ovarian cancer, diagnosed at age 31. On physical examination, a large mass,

Figure 6 Final result of the incisional wound after complete mass resection, closed with 3-0 Stratafix in a subcuticular fashion.

Table 1 Comprehensive review of the literature involving round ligament leiomyomas

Author, year	n, age	Presentation, (E/I)	Size (cm)	Laterality	Uterine fibroids	Oestrogen exposure	Diagnostic evaluation	Surgical management	Complications/recurrence
Ali <i>et al</i> , 2012 ²	1, 38	Irreducible inguinal mass (E)	5.0×5.0	Right	No	N/R	No imaging, no biopsy	CSR	None
Bakotic <i>et al</i> , 1999 ³	1, 69	Irreducible inguinal mass (E)	5.8	Left	No	N/R	CT	CSR	None
Bedir <i>et al</i> , 2016 ⁴	1, 32	Pelvic pain during pregnancy (I)	4.8×3.5×3	Right	No	N/R	U/S	CSR during C/S	None
Birge <i>et al</i> , 2015 ⁵	1, 28	Dyspareunia (E)	5.0	Left	Yes	G0	TVU, cystoscopy	CSR	None
Chang <i>et al</i> , 2013 ⁶	1, 52	Chronic pelvic pain (I)	4×3.5×5	Left	No	G6 P4	TVU	DL, CSR	None
Colak <i>et al</i> , 2013 ⁷	1, 40	Irreducible, painful inguinal mass (E)	6.0×6.0	Right	No	N/R	U/S	CSR	None—1 year
Deol and Arleo, 2017 ⁸	1, 68	Groin pain (E)	2.1×1.7×2.0	Left	Yes	G1 P0-0-1-0	TVU, MRI	N/R	N/R
Grossman and Cheung, 2007 ¹⁰	1, 49	Adnexal mass on imaging (I)	4.0	Left	No	N/R	TVU	DL	N/R
Guillen <i>et al</i> , 2009 ¹¹	1, 36	Acute pelvic pain (E)	8.0	Right	No	N/R	U/S	CSR	None
Harish <i>et al</i> , 2014 ²³	1, 40	Irreducible, painful inguinal mass (E)	3.0×4.0	Left	Yes	N/R	U/S, CT, FNAC	CSR	None
Hwang <i>et al</i> , 2010 ¹²	1, 51	Abdominal pain (I)	6.8	Left	No	G0	KUB, CT	DL, CSR	None
Kan <i>et al</i> , 2017 ¹³	1, 39	Irreducible inguinal mass (E)	8.0×7.0	Left	No	G0	TVU	DL, CSR	None
Kirkham <i>et al</i> , 2008 ¹⁴	1, 43	Irreducible inguinal mass (E)	9.0	Right	N/R	N/R	MRI, CT, biopsy	CSR	None
Klingbeil, 2017 (present article)	1, 68	Irreducible, painful inguinal mass (E)	25×8.5×9	Left	Yes	G2 P2	CT, biopsy	CSR, HR	None
Lazarevski and Pop-Ristova, 1990 ¹⁵	1, 53	Urinary incontinence (I)	N/R	Left	N/R	N/R	TVU	CSR	None
Losch <i>et al</i> , 1999 ¹⁶	1, 59	Palpable adnexal mass (I)	7.9×6.5×5.6	Right	Yes	G2 P2, HRT	TVU, CT	DL, CSR	None
Michel and Viola, 2003 ¹⁷	1, 50	Pelvic pain (I)	14×13×7	Right	Yes	G2 P2	U/S, CT, MRI	CSR	None
Najjar and Mandel, 2016 ¹⁸	1, 47	Irreducible inguinal mass (E)	4.0×3.0×2.0	Right	No	G1 P1	No imaging, no biopsy	CSR, SMP	None
Rhee <i>et al</i> , 1999 ¹⁹	1, 49	Irreducible inguinal mass, MRKHS (E)	9×8.5×5	Left	N/A	None	MRI	DL, CSR, HR	None
Salem Wehbe <i>et al</i> , 2016 ²⁰	1, 40	Infertility, MRKHS (I)	7.0×9.0	Right	N/A	None	TVU	DL, CSR	None
Vignali <i>et al</i> , 2006 ²¹	1, 40	Acute pelvic pain (I)	10×12×9	Right	No	N/R	TVU	DL, CSR	None
Warshauer and Mandel, 1999 ²²	1, 63	Groin pain (E)	3.0×2.0	Right	Yes	HRT	CT	CSR	None

CSR, complete surgical resection; C/S, caesarean section; DL, diagnostic laparoscopy; E, external; FNAC, fine needle aspiration cytology; G, gravida; HR, herniorrhaphy; HRT, hormone replacement therapy; I, internal; KUB, kidney ureter bladder radiograph; MRKHS, Mayer-Rokitansky-Kuster-Hauser syndrome; N/A, not applicable; N/R, not reported; P, para; SMP, synthetic mesh placement; TVU, transvaginal ultrasound; U/S, ultrasound.

approximately 30 cm in greatest diameter, was visualised in the superficial wall of the left lower quadrant of the abdomen. The mass was non-tender to palpation, slightly mobile and irreducible. A well-healed scar was visible from the previous incisional biopsy, with no overlying skin changes.

INVESTIGATIONS

A repeat CT scan of the abdomen was acquired for further evaluation of the abdominal mass. Imaging revealed a 27.75×9.41 cm, well-circumscribed heterogeneously enhancing mass in the subcutaneous soft tissue of the left abdominal wall, extending superiorly to the left iliac wing and inferiorly into the mons pubis (figure 1). It appeared to originate from either the external abdominal oblique or rectus abdominis muscles, however there was also evidence of continuation with the round ligament at the external inguinal ring (figure 2).

DIFFERENTIAL DIAGNOSIS

Inguinal masses in women have many possible aetiologies, of which the majority are relatively benign. The differential diagnosis includes inguinal or femoral hernia, inguinal lymphadenopathy, inguinal lipoma, round ligament varicosities, mesothelial or desmoid cyst, canal of Nuck hydrocele, round ligament tumour (eg, leiomyoma, endometrioma, adenomyoma), uterine leiomyoma, psoas abscess and femoral artery aneurysm.

TREATMENT

The patient was scheduled for surgery 1 week later for complete excision under general anaesthesia (figure 3). The mass was isolated from surrounding tissues using monopolar electrocautery in a circumferential manner through a clear fascial plane (figure 4). The root of the mass was in the region of the external inguinal ring, and on further dissection, it was found to

be embedded on the round ligament. The tumour was excised (figure 5), and the rest of the procedure was carried out in similar fashion to a Bassini inguinal hernia repair (figure 6).

OUTCOME AND FOLLOW-UP

The patient tolerated the surgery well with no complications postoperatively. She was discharged 2 days later. Final pathology identified a leiomyoma, 25 cm in largest dimension, with evidence of haemorrhage and necrosis, suggestive of carneous degeneration. There was no evidence of malignancy and the resected margins were negative.

At 2-week follow-up, the patient returned in good health. She was able to perform her regular daily activities with minimal pain. The incision site was healing as expected, without evidence of infection. She was advised to follow up in clinic only as needed.

DISCUSSION

The only previous review of the literature regarding round ligament leiomyomas was published in 1962 and reported 110 cases.¹ A new, comprehensive MEDLINE search of the literature regarding leiomyomas of the round ligament was completed and revealed 22 case reports published between 1962 and 2017 (table 1).^{2–23} The average age for presenting symptoms was 47.9 years. The majority of leiomyomas were located extra-abdominally, occurring in 59% of cases. The most common presentation was a painless, irreducible inguinal mass. Two cases occurred in the presence of Mayer-Rokitansky-Kuster-Hauser syndrome.^{19 20} There was no preference for laterality, and the average longest tumour dimension was 7.5 cm. A history of uterine leiomyomas occurred in 32% of cases. Two patients were taking hormone replacement therapy, and one patient was 28 weeks pregnant. With regard to preoperative diagnostic evaluation for intra-abdominal leiomyomas, transvaginal ultrasound (n=6) was used most often. Extra-abdominal leiomyomas were imaged using abdominal ultrasound, CT or MRI. Two cases were evaluated by clinical examination alone. Tumour biopsies were taken in three cases. In all patients, surgical management was the primary treatment option. A laparoscopic approach was most commonly used for intra-abdominal tumours, whereas extra-abdominal tumours were approached in a similar fashion to an inguinal hernia repair. No evidence of recurrence or complications were reported.

Leiomyomas, also known as fibroids, are a common benign tumour of smooth muscle tissue, previously estimated to occur in 20%–40% of women by 35 years of age. Recent population-based studies have shown that the prevalence is probably much higher, up to 70% in women by the age of 50.²⁴ Common forms of leiomyoma occur in the uterus, oesophagus and small bowel. Leiomyomas of the round ligament are an extremely rare finding. The pathogenesis of these tumours is relatively unknown; however, it is thought to involve complex interactions between somatic mutations, local growth factors and steroid hormones. Similar to other types of leiomyomas, those found in the round ligament have been shown to express both oestrogen and progesterone receptors.²⁵ Oestrogen is known to be a major promoter of growth, and previous cases have shown that patients who are pregnant⁴ or receiving hormone replacement therapy^{16 22} demonstrate an increase in tumour size. Furthermore, round ligament leiomyomas more frequently occur in premenopausal women.

As leiomyomas mature in size, they may eventually outgrow their blood supply, resulting in various types of degeneration. Hyaline degeneration is the most common type, occurring

in approximately 60% of cases.²⁶ Red or carneous degeneration is a rare type, caused by haemorrhagic infarction due to obstructed draining veins. This type of degeneration is typically associated with pregnancy and hormone replacement therapy.²⁶ Clinically, in the presence of a degenerating leiomyoma, patients may present with pelvic pain, low-grade fevers and leucocytosis lasting from a few days to weeks.

The diagnostic workup for a round ligament tumour is important due to the long list of possible differential diagnoses. Radiographic imaging is perhaps the most useful diagnostic tool, especially in the presence of an extra-abdominal mass concerning for an incarcerated hernia. Ultrasound imaging is often used for initial investigation. Leiomyomas appear as well-defined, hypoechoic masses, often with a whorled appearance.⁸ MRI is the preferred imaging modality, however, because of its greater sensitivity of identifying deep soft tissue structures.²⁶ Round ligament leiomyomas often appear with iso-intensity when compared with the myometrium on T1-weighted imaging.²⁶ Those undergoing degeneration may present as heterogeneous masses with variable presentations. Biopsy is typically not indicated during initial workup.

Patient's perspective

The last 2 months were very frightening for my family and I. I had what I thought was a hernia for a very long time, but then suddenly I started feeling sick and getting fevers, and the hernia seemed to double in size over 4–5 days. I went to two different hospitals before I came here. My family was frustrated because it seemed like I wasn't getting the help I needed. Then, when I finally found a doctor to do surgery, we had a tragedy. My husband fell and suddenly passed away. This happened just 2 days before I was supposed to have surgery. My children were devastated, and I know they were very scared about sending me into surgery without the support of my husband nearby. My daughter expressed a fear of losing me in surgery, too. I even stayed in the hospital an extra day because we were afraid that I would get dizzy and fall like my husband. Now, though, we are all very relieved. The recovery from surgery was easier than I expected, and I am doing very well. I am happy to be living without that tumour and the fear of getting sick again. I am very thankful to be healthy for my family.

Learning points

- Tumours of the round ligament are a rare finding that may present as an inguinal or labial mass, which can present similarly to an incarcerated hernia.
- Leiomyomas are the most common cause of a round ligament tumour and are known to grow rapidly in size, especially in the presence of high oestrogen levels, such as during pregnancy or with hormone replacement therapy.
- A thorough workup is necessary to ensure an accurate diagnosis. Ultrasound and MRI are the preferred imaging modalities. Biopsy of the inguinal mass is rarely indicated.
- Complete surgical resection is the optimal management for symptomatic leiomyomas, with concomitant repair of the inguinal canal defect to decrease the risk of future hernias.
- Postoperative outcomes of round ligament leiomyomas are satisfactory, with a low risk for complications and recurrence.

Surgical exploration is the primary treatment option to quickly differentiate between the various aetiologies of an abdominal wall mass, followed by complete excision of the mass at its base. If the mass involves the inguinal canal, a formal inguinal hernia repair is recommended, with or without mesh depending on the surgeon's preference.^{5 18} In the past 25 years, no evidence of round ligament leiomyoma recurrence has been reported, suggesting a good prognosis.

In conclusion, round ligament leiomyomas are a rare finding in female patients. Extra-abdominal tumours of the round ligament may present similarly to an incarcerated inguinal hernia; therefore, proper diagnostic workup is essential. Surgical exploration followed by complete excision is the recommended treatment option. In the currently reported case, the abdominal wall mass was found to involve the extra-abdominal round ligament and was excised successfully. On pathological evaluation, the mass was found to be benign, with evidence of necrosis and haemorrhage indicating carneous degeneration. The patient recovered without complications.

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