Lesions of the Round Ligament and Canal of Nuck - It’s Not Always an Inguinal Hernia: A Review

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Article begins on next page
Lesions of the Round Ligament and Canal of Nuck-It’s Not Always an Inguinal Hernia-a review

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Precis:
Lesions of the round ligament and canal of Nuck are reviewed.

Key words: round ligament, canal of Nuck, uterine neoplasms, female
Abstract:

Background:

Lesions arising in the round ligament and canal of Nuck are uncommon, and may prove challenging to clinicians.

Objectives:

To review the scope of lesions affecting the round ligament and canal of Nuck.

Methods:

A literature review was conducted. A Medline search was used, using the round ligament, canal of Nuck, uterine neoplasms, female

Results:

Reported lesions of the round ligament and canal of Nuck were reviewed. Many of these lesions can be mistaken for each other clinically.

Conclusions:

A review of the scope of rectovaginal lesions is presented to assist in developing a differential diagnosis if a patient with such a lesion is encountered.
Introduction:

The uterine round ligament and canal of Nuck may give rise to a variety of lesions. Except for round ligament pain in pregnancy, and perhaps round ligament leiomyomata, clinicians may be unfamiliar with the range of pathology in this area, and many of these lesions present clinically as inguinal hernias. This review aims to fill in the other areas of specific pathology that can occur in the round ligament (table 1).

Methods:

A literature review was conducted. A Medline search was used, using the round ligament, canal of Nuck, uterine neoplasms, female.

Results:

Reported lesions of the round ligament and canal of Nuck were reviewed. Many of these lesions can be mistaken for each other clinically.

Discussion:

Embryology, Anatomy & Histology

The round ligament is derived from the gubernaculum, which travels through the abdominal wall along with the processus vaginalis peritonei which is the peritoneum outside the round ligament, traversing the inguinal canal. While the traditional thinking is that the round ligament terminates in the labium majus, in an anatomic study conducted by Attah and Hutson (1), the round ligament was found to end just beyond the external inguinal ring, implanting either into...
the abdominal wall muscle or the adipose of the mons pubis. The processus vaginalis peritonei usually obliterates in utero, but may persist as the canal of Nuck(2). The round ligament hence has intrapelvic and extrapelvic components, and is composed primarily of smooth muscle(figure 1), with blood vessels and connective tissue, and a mesothelial investment(3), although skeletal muscle has been described in close to 3% of one series, thought to represent persistence and maturation of fetal rhabdomyoblasts(4). Blood supply to the round ligament is primarily from the inguinal duct and abdominal wall(2)

**Congenital Anomalies**

Persistent patency of the processus vaginalis(Canal of Nuck) allows for pelvic contents to enter into the inguinal canal, resulting in inguinal hernias. Some syndromes of sexual differentiation may have a component involving the canal of Nuck, with cases presenting as inguinal hernias. This may include an inguinal testis, ovary or uterine remnant. This is most common in Androgen Insensitivity Syndrome, but has also been described in Mayer-Rokitansky-Küster-Hauser syndrome(5). In addition, a variety of uterine anomalies may be associated with incarceration of a uterine horn in the inguinal canal(6). Inguinal gonads or uteri may incarcerate or torse(7). In addition to herniation of a normal ovary, an accessory ovary(connected to a normal ovary ligamentously), already rare, has been described in a round ligament with endometriosis(8). Percutaneous oocyte retrieval of an inguinal ovary has been reported as well(9).
Round ligament syndrome

The growing uterus can exert traction on the round ligaments, which can cause severe pelvic and back pain. Specific exercises and acupuncture are some of the modalities that have been used to treat this condition. Examination is indicated to rule out more medically severe mimics(10).

Trauma

Traumatic laceration of the round ligament has occurred with coitus, without accompanying vaginal laceration(11). In addition, a case of pelvic pain in an adolescent was diagnosed at laparoscopy as a partial detachment of the uterine ligament at its uterine insertion. Attempted reapproximation created significant traction on the uterus, and it was thought that this would increase the pain, so no repair was performed, and the girl’s pain decreased over time(12).

Benign Tumors and Tumor-like Lesions

Hydrocele of the Canal of Nuck and Cyst of the Round ligament

A hydrocele may develop, if there is persistent patency of the processus vaginalis with collection of fluid in the Canal of Nuck, presenting as a cystic mass in the inguinal canal or inguinal hernia(13). There are three types of hydrocele. The most common is without
communication with the peritoneal cavity forming a cyst around the round ligament; a hydrocele that does communicate with the peritoneal cavity; and the “hourglass” type, where the inguinal ring constricts one portion, and one portion communicates with the peritoneal cavity(14). In one highly unusual case, a patent Canal of Nuck was the underlying etiology for marked unilateral labial swelling in a young woman with ascites associated with ovarian hyperstimulation after treatment with human menopausal gonadotropins. Unlike usual labial edema, this mass was cystic, and treated with aspiration(15). It appears the mass was actually a fluid-filled patent canal of Nuck. In a very unusual case, an ectopic pregnancy was found to arise in a cyst of the canal of Nuck(16). Unlike a prior case, where there was herniation of a tubal ectopic pregnancy(17), the authors could not explain how this case arose, as there was no connection between the cyst and the peritoneal cavity detected. Ultrasonography may be helpful in the workup of canal of Nuck cysts(18). Repair has been performed laparoscopically, extraperitoneally(14). Cysts of the Canal of Nuck may become infected. The case presented by Ameh(19) was of a neonate with a rapidly growing cystic inguinal mass, which grew out Klebsiella.

Mesothelial cysts of the round ligament have been described(20). It is debatable whether this is a distinct entity from a hydrocele of the canal of Nuck, given the similar ultrasound and histologic findings. Mesothelial cysts are lined by a flattened mesothelium. Since the processus vaginalis is composed of peritoneum(mesothelium), these lesions may be identical, however another theory is that there is embryonic mesothelial tissue incorporated during formation of the round ligament, which would make a mesothelial cyst distinct from a cyst of the canal of Nuck(20). Ryley et al(21) presented a case of a woman who presented with what was thought to be an incarcerated inguinal hernia presenting after gonadotropin stimulation of the ovary, and
attributed this presentation of a previously unrecognized mesothelial cyst to the therapy, which may have led to enlargement of the round ligament. Mesothelial cysts of the round ligament are often associated with inguinal hernias(22), which may be small.

**Other Cysts of the Round Ligament**

Epidermal inclusion cysts have rarely been reported in the round ligament. Kim et al report such a case arising in the intraperitoneal portion of the round ligament in a 21 year old female, treated by laparoscopic excision. In cases where there is no possibility of iatrogenic causation such as in a prior episiotomy, the underlying etiology may be epithelial rests that have been situated during fetal life, or seeding during germ cell migration(23).

**Hematoma and hemorrhage**

A hematoma of the Canal of Nuck arose spontaneously in a woman on anticoagulant therapy as a painful inguinal mass. In addition to hematoma, incarcerated hernia was in the differential (24). A case of hemorrhage into the round ligament itself, presenting as an incarcerated inguinal hernia was reported(25).
Varices of the Round Ligament

Pregnancy is a risk factor for the development of inguinal hernias, however not all masses in the groin presenting in pregnancy and the postpartum period are inguinal hernias. Although expectant management during pregnancy is often practiced, if the mass becomes painful, concern about incarceration is raised, and surgery may be contemplated. It has been urged that additional studies beyond physical examination, such as ultrasonography, be employed, lest other entities that are usually nonsurgical be misdiagnosed, such as varices(26). Round ligament varices, like an inguinal hernia, will transmit cough impulses(27). Varices of the round ligament may develop due to compression of the pelvic veins by the gravid uterus, increased blood volume of pregnancy, or increased action of higher progesterone levels on receptors within round ligament veins during pregnancy(28). Aside from pain, other risks of these varices include thrombosis, and potentially rupture(29,20). Magnetic resonance imaging and computerized tomography have also been utilized in treatment planning(31). Management is usually expectant for round ligament varices without complications.

Round ligament varicosities may be suspected if groin swelling occurs in pregnancy along with vulvar and leg varicosities. Less common is accompanying uterine varicosities(32), which could potentially rupture, with risk of hemoperitoneum(33) or possibly interfering with an operative delivery.

Endometriosis

Inguinal endometriosis affects less than one percent of women with endometriosis(34), most often involving the round ligament, and in particular the right round ligament(34,35), for unclear
reasons. Not surprisingly, endometriosis is much more common in the intrapelvic portion of the round ligament(36). Extrapelvic round ligament endometriosis is highly associated with pelvic endometriosis(34), causing some investigators to recommend diagnostic laparoscopy to rule out pelvic endometriosis when inguinal endometriosis is detected(37). Inguinal endometriosis is also associated with an inguinal hernia, in about a third of cases(35). Inguinal endometriosis may present with a groin mass, and often with cyclic groin pain with menses. Although the underlying etiology may be extension of pelvic endometriosis, in one case(38), the authors attributed the inguinal endometriosis to the functioning incarcerated uterine horn in the hernia, supporting the retrograde menses with subsequent implantation theory. Inguinal endometriosis may not be suspected preoperatively. In addition to imaging, fine needle aspiration biopsy may be helpful in the preoperative workup.

Rare cases of malignant transformation of inguinal endometriosis have been reported, as can happen in intrapelvic endometriosis(39,40). While these cases, like intrapelvic ones, are usually endometrioid or clear cell adenocarcinoma, a case of papillary serous carcinoma arising from inguinal endometriosis was reported in a young women who had a successful pregnancy after therapy(40). Endometrial stromal sarcoma arising in endometriosis of the round ligament has also been reported(41).

**Adenomyosis**

Tabrizi et al(42) described two cases of nodular adenomyosis of the uterus adjacent to the uterine portion of the round ligament, detected during diagnostic laparoscopy, and leading to severe groin pain in both patients. Both patients were relieved by excision. One of the two cases
specifically reported excruciating groin pain during menses. Both cases had hemorrhage into the center of the nodules.

**Leiomyoma**

Leiomyomata are the most common neoplasm of the round ligament, of no surprise considering the round ligament is mostly smooth muscle. They are often associated with uterine leiomyomata(43). They may cause pain by traction on the round ligament. They can occur in the intrapelvic portion of the ligament, or extraperitoneally in 2/3 of the cases, in the inguinal canal or out on the vulva(44).

**Lipoma of the Round Ligament**

Lipomas, benign neoplasms composed of adipose tissue, are often incidental findings at the time of inguinal hernia repair, which is more common in men. The term lipoma, however, may be inaccurate, as most of these lesions more likely represent protrusion of extraperitoneal fat rather than a true neoplasm(45). However, lipomas may occur in the region in the absence of hernia, and may occur in the round ligament in women as well. Seven of the nine cases of round ligament lipoma presented in Lilly’s series(45) presented with pain. Six did not have an associated hernia. Five of these six had significant pain relief after laparoscopic surgical excision and repair. A case of round ligament lipoma occurring in a 24 weeks pregnant woman, mimicking appendicitis has been reported(46).
Other Rare Benign Masses and Neoplasms

Other rare groin masses have included cystic lymphangiomas of the inguinal canal, angiomyolipoma of the canal of Nuck, and round ligament masses such as nodular hyperplasia in pregnancy, solitary fibrous tumors (which rarely metastasize), and mature teratomas (47-52). In one unusual case (53), teratomas were present in both round ligaments and one ovary in a woman undergoing cesarean section. Ultrasound is useful in assisting with the differential diagnosis (47).

Malignant Neoplasms:

Primary malignancies of the round ligament or canal of Nuck are rare. Most adenocarcinoma has been described as arising in a background of endometriosis, but occasional cases of adenocarcinoma have been reported to arise directly from the canal of Nuck (54, 55). Sun’s case of endometrioid adenocarcinoma was attributed to Müllerian metaplasia or remnants, while the papillary serous histology in Peison’s case (55), in a woman with prior TAH-BSO for benign disease, was considered a primary peritoneal papillary serous adenocarcinoma of the peritoneum of the canal of Nuck.

Other rare primary malignant neoplasms have included sarcomas, including leiomyosarcoma (56), and perivascular epithelioid cell tumor (PEComa) (57).
Secondary malignancy of the round ligament has been reported, including direct extension from an endometrial carcinoma (51), as has metastatic spread of ovarian carcinoma (58). We have seen metastatic fallopian tube carcinoma to the round ligament (figure 2).

**Conclusions:**

The round ligament and canal of Nuck can show a wide range of pathology, most of which is rare enough not to be encountered in a routine gynecologic practice. The most common presentations are of mass or pain, and the most common clinical diagnosis is inguinal hernia. Preoperative imaging has been shown to be very helpful. Many of these lesions are amenable to minimally invasive surgical procedures. A knowledge of the range of lesions of the region will be helpful if such a patient is encountered.
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Legends:

Figure 1- Round ligament, composed of smooth muscle bundles

Figure 2-Metastatic poorly differentiated adenocarcinoma of fallopian tube(right) metastatic to round ligament(left).