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Case Report

Robotic Assisted Laparoscopic Pelvic Lymphadenectomy for Primary Gynaecologic Chylous Reflux

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ABSTRACT

Background: Gynaecologic chylous reflux is a rare condition with significant impact on patient quality of life and nutritional status. While no long-term treatment options have been successful this article looks at the possibility of using robotic-assisted surgical management as a less invasive option for treatment.

Case Report: This patient had milky vaginal discharge which contained high levels of triglycerides. A robot-assisted pelvic lymphadenectomy was performed which resulted in resolution of discharge for eight months before returning to pre-surgical volume. Symptoms significantly improved after a second operation and dietary modification.

Conclusion: Robot-assisted lymphadenectomy may be a minimally invasive approach for the treatment of primary gynaecological chylous reflux.

Introduction

Primary chylous reflux is a rare condition caused by incompetent lymphatics. Several mechanisms have been proposed including lymphatic hypoplasia and incompetent valves [1]. Primary chylous reflux is typically congenital, however, it often presents later in life. Secondary chylous reflux may be caused by neoplasm, radiation, trauma, surgery, or filariasis [2]. Gynaecologic chylous reflux can present as dermal vesicles, lymphedema, vulvar edema, chyluria, hypomenorrhea, chylovulvorrhea, or chylocolporrhea [1]. The diagnosis can be made based upon analysis of the discharge and requires at least two of the following criteria: appearance of milky fluid, fluid triglyceride level greater than 101 mg/dL, or lipoprotein findings in chylomicrons [3].

The early diagnosis of chylous effusions is essential to prevent significant loss of fluid volume, disruption of daily activities, as well as the risk of malnutrition. Daily discharge can be significant, with up to 2000 ml of discharge daily [2]. Thus, in addition to disrupting daily activities there are additional risks including lipid and protein loss, malnutrition, anemia, hypoproteinemia, hypocalcemia, hypolipidemia, and increased risk of infection [3].

This case presents a patient whose primary complaint was leukorrhea. Herein, we describe the technique for the first documented robot assisted laparoscopic pelvic lymphadenectomy. After diagnosis, a robotic assisted, minimally invasive technique was used to ligate the diseased lymphatics with full resolution of symptoms at six weeks postoperative. At one year postoperative, symptoms had recurred, and a reoperation was completed with resolution of symptoms in the immediate postoperative period. Within a few months, symptoms recurred but were improved with dietary modification.

Case Report

A 19-year-old female presented to the clinic complaining of copious liquid vaginal discharge. She had been seen by multiple providers at various institutions over several years and undergone multiple tests and pelvic exams without being offered any effective treatment options. Of note, she had been treated with antifungals and antibiotics which had not alleviated the symptoms. The odorless vaginal discharge began spontaneously at age twelve and ranged from a milky yellow to a light green colour. Hydration, stress, and a fatty diet increased the volume of discharge. The patient used five to eight pads daily to manage the copious amounts of fluid discharge. She underwent menarche at age

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twelve and had regular menses lasting an average of three to four days. Her menses occurred at regular intervals and was mixed with the fluid discharge. She had no previous history of sexual activity. Her medical and surgical history were unremarkable. Her only allergy was to tree nuts. Family history was positive for fibrosarcoma in her mother. Her review of systems was positive only for fatigue. She was a college student and denied a history of alcohol or tobacco use.

The physical exam showed a Caucasian female with a BMI of 19.8. She had appropriate Tanner stages of development. On the gynaecologic exam, her hymen was intact, with pooling of milky liquid in her vagina. The cervix was difficult to visualize secondary to patient anxiety. She had well supported hyper-rugated vaginal walls which appeared almost multicystic.

The differential diagnosis included:

i. Urinary leakage (urge incontinence, fistula)
ii. Vaginal discharge (bacterial/ fungal infection)
iii. Sexually transmitted infection
iv. Urinary tract infection
v. Medication or drug side effect
vi. Metabolic condition
vii. Neurologic condition (Multiple sclerosis)
viii. Ectopic ureter (ureter at proximal urethra)
ix. Chylous reflux

A complete blood count, complete metabolic panel, prolactin, and thyroid stimulating hormone all came back within normal limits. Methylene blue and oral pyridium tests were both negative. Analysis of the vaginal fluid showed a high fat content with triglycerides of 1101 mg/dL. Imaging included an MRI and a CT scan from five years prior which were both within normal limits. Diagnostic laparoscopy was also unremarkable. A diagnostic hysteroscopy showed whitish discharge from the external cervical os. No lesions or focal abnormalities were found on the cervical or endometrial canals and both ostia were within normal limits. Hysteroscopy was otherwise unremarkable. Radiopaque contrast was injected into the groin and pericervical lymphatics after which fluoroscopy was performed. Contrast was seen in the pelvis; however, no areas of abnormality were appreciated. Despite the negative imaging, a presumed diagnosis of chylous reflux was made based on the presence of milky white discharge with high triglyceride levels.

i. Preserve fertility
ii. Preserve anatomy, in particular, the blood supply to the uterus
iii. Remove only diseased lymphatics (if possible)
iv. Avoid excessive dissection so as to prevent lymphedema
v. Perform the surgery in a minimally invasive manner for benefits to patient
vi. Use pericervical indocyanine green to aid in dissection
vii. Use technology where appropriate.

Prior to dissection of the left broad ligament, lysis of adhesions was required. Upon dissection of the broad ligament, it was noted that there was a wet appearance inside of the ligament. An abnormal saccular vessel was identified and dissected. Despite being ninety minutes after injection of indocyanine green, the abnormally dilated left pelvic lymphatic vessels did not fluoresce. During dissection, there was an inadvertent injury to a branch of the right uterine artery which was repaired via careful suture ligation instead of cautery in order to preserve adjacent branches. The left abnormal lymphatics were removed and sent to pathology for further study (Figure 1). Arista (Plant-based absorbable hemostatic particles) was applied to coagulate any remaining open vessels.

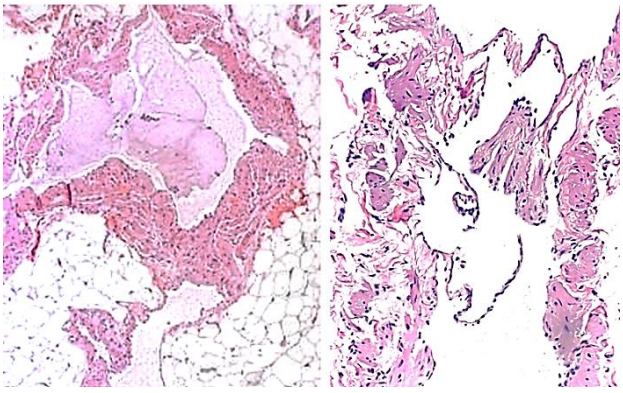

Figure 1: Left: Dilated varicose mega lymphatic vessels with smooth muscle hyperplasia in abnormal thickened walls (H&E stain, 100x). Right: Thick incompetent lymphatic valve (H&E stain, 200x).
A surgery was planned with the following goals to:

Six weeks postoperatively the patient presented for follow-up. She was doing well with no complaints. Incisions were well healed, and she had no residual pain. Patient no longer had bothersome vaginal discharge. She had normal menses for a duration of two days. Speculum examination revealed physiologic vaginal discharge in the posterior fornix and cervical mucus at the os but no liquid vaginal fluid from the external os.

The patient followed up one year after surgery with recurrent symptoms which had started approximately seven to eight months postoperatively and had gradually worsened to the original severity. The decision was made to reoperate with similar methods as discussed above. Of note, during the second surgery peri-cervical indocyanine green still did not result in fluorescence of any abnormal lymphatic vessels from time of injection to one hour after injection. In the immediate postoperative period, no chylous discharge was noted. At six-month follow-up the patient reported partial recurrence of symptoms using up to six pads daily. However, she has found that excluding gluten and dairy from her diet decreases discharge to two non-saturated pads daily. Overall, she is very pleased with the results of the operations and dietary modification.

Discussion

Primary gynaecologic chylous reflux is a rare lymphatic condition. Presentations include dermal vesicles, lymphedema, vulvar edema, chyluria, hypomenorrhea, chylovulvorrhea, and chylocolporrhea [2]. While the spectrum of presentations are broad and not always obvious, several diagnostic criteria have been proposed and include at least two of the following: a milky appearance of discharge, fluid triglyceride levels greater than 101 mg/dL, or lipoproteins in the chylomicrons [3]. In this case, the patient presented with only chylocolporrhea which had a milky appearance with elevated fluid triglyceride levels.

Several imaging studies have been used to find anomalous lymphatic vessels including magnetic resonance imaging (MRI), lymphangiogram,

and computerized tomography (CT). Due to the challenge of distinguishing between uterine, cervical, or vaginal wall discharge, imaging modalities are crucial for accurate anatomic diagnosis. MRI has been used to define the soft tissues of the pelvis including the tortuous, serpiginous lymphatic vessels typical of chylous reflux [4]. However, lymphangiogram is the diagnostic test of choice as dye can be run through different lymph vessels until the diseased vessel is clearly found [5]. CT has also been used particularly after a lymphangiogram to show the full scope of diseased vessels and leakage points [5]. This case presented exceptional challenges in diagnosis of anatomical defects as none of the imaging modalities showed any abnormalities.

Disease complications can vary from patient to patient. In patients with whom vaginal discharge is the primary complaint, as in this case, concerns often include patient discomfort and loss of fluid. In some severe instances, discharge has been recorded up to 2000 ml a day. Large volumes of discharge may necessitate patients to wear multiple adult diapers daily. The result of such volume loss can cause nutrient deficiencies with hypoproteinemia, hypocalcemia, and hypolipidemia among the most commonly documented deficiencies [3]. General malnutrition and anemia have also been reported. Increased risk of infection has been noted in these patients with several possible causes including malnutrition and increased exposure to bacterial entry into the lymphatic system [6]. There is one documented case of a patient who succumbed to severe malnutrition and recurrent bacterial infections despite dietary modification and parenteral nutrition [3]. While the patient in this case had not yet manifested any nutritional deficiencies, it was important to address her large volume of discharge to prevent future complications.

Treatment options for gynaecologic chylous reflux have varied from conservative dietary treatments to surgical ligation of lymphatics. Conservative measures have been shown to be somewhat beneficial in patients with mild disease. Dietary modification is often first line and included encouraging a low fat, high medium chain triglyceride diet [1]. In some patients this is sufficient to resolve discharge and improve malnutrition [1]. In this case, the patient did note that symptoms worsened with a fatty diet, however, a low-fat diet was not sufficient for resolution of symptoms. Medications such as diuretics have also been beneficial for some patients with complete resolution of symptoms [1]. Recently several studies have been published which showed that interventional radiology embolization of affected nodes and vessels may provide resolution of symptoms for up to 12 months [7, 8].

Few cases of surgical management of gynaecologic chylous reflux have been documented with open being the most common approach [5]. Even in cases of laparotomy, symptoms may resolve for a period of time, however, cases with longer follow up documented show recurrence weeks to months after surgery [6, 9, 10]. However, due to the benefits of robot-assisted surgery including improved precision, magnified visualization of enlarged vessels, access to deep pelvic spaces without large incisions, quick recovery and little blood loss, this technique was considered to be a possible surgical approach for resolving more severe chylous reflux [11].

This case presented a patient with primary gynaecologic chylous reflux who failed conservative dietary management. She was treated with robot assisted laparoscopic pelvic lymphadenectomy with resolution of symptoms at six weeks follow up but recurrence at seven to eight months postoperatively. A repeat surgery was performed which also had a short-term resolution of symptoms followed by recurrence. After the second recurrence, the patient noted that eliminating gluten and dairy from her diet significantly decreased the vaginal discharge. While there was a recurrence of symptoms, similar to the results of other treatment modalities, this case indicates that minimally invasive lymphadenectomy may be a less invasive surgical option for pelvic chylous reflux.

Précis

Robot-assisted lymphadenectomy may be a minimally invasive approach for the treatment of primary gynaecological chylous reflux.

Highlights

i. Robot assisted laparoscopic pelvic lymphadenectomy is a preferred approach for chylocolporrhea.
ii. Differential for continuous liquid vaginal discharge should include gynaecologic chylous reflux.
iii. If concern for chylous reflux is present, test fluid for triglyceride levels.

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Conflicts of Interest

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Consent

The patient has provided written informed consent for publication.

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