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Right inguinal hernia with the presence of a uterus, fallopian tube, and ovary associated with Mayer-Rokitansky-Küster-Hauser syndrome

Hernia inguinal derecha con presencia de útero, trompa de Falopio y ovario, asociada a síndrome de Mayer-Rokitansky-Küster-Hauser

Carlos Humberto Ramírez-Mendoza,* Luis Enrique Sánchez-Sierra,‡ Ana Romero-Lanza,§ Araceli Chicas Reyes¶

Keywords: Inguinal hernia, Mayer-Rokitansky-Küster-Hauser syndrome, fallopian tube, ovary.

Palabras clave: *Hernia inguinal, síndrome Mayer-Rokitansky-Küster-Hauser, trompa de Falopio, ovario.*

ABSTRACT

Introduction: abdominal wall hernia is a frequent pathology, with an estimated prevalence of nearly 5% in the general population; the most frequent presentation is inguinal type, accounting for about 70% of the total. Mayer-Rokitansky-Küster-Hauser syndrome is a rare congenital anomaly of the genital tract; its etiology is unknown. It usually presents primary amenorrhea in adolescent females with normal external genitalia and growth. It may be associated with other alterations, especially at the genitourinary level. **Clinical case:** 25-year-old female patient, with pathological history of primary amenorrhea and surgical history of left inguinal hernioplasty, with a history of protrusion in the right inguinal region of three years of evolution, which increases with physical exertion. Physical examination revealed a bump in the right inguinal region of approximately 5 cm in diameter with Valsalva, without color or inflammatory changes, auscultation showed normal bowel sounds, and palpation revealed a mass in the right iliac fossa of soft, depressible, painful, and reducible consistency. On gynecological examination, the patient with female phenotype and typical secondary sexual characteristics presented symmetrical labia majora and minor, permeable vaginal duct. The clinical diagnosis was an uncomplicated right inguinal hernia, surgical treatment was decided, and she was scheduled for elective surgery, finding the right inguinal hernia, the hernial sac containing the uterus, fallopian tube, and ipsilateral ovary.

RESUMEN

Introducción: *La hernia de pared abdominal es una patología frecuente, se calcula que la prevalencia es cercana a 5% en la población general; la presentación más frecuente es de tipo inguinal, cerca de 70% del total. El síndrome de Mayer-Rokitansky-Küster-Hauser es una rara anomalía congénita del tracto genital, se desconoce su etiología. Por lo general se presenta como amenorrea primaria en mujeres adolescentes, con genitales externos y crecimiento normales. Puede asociar otras alteraciones, especialmente a nivel genitourinario.* ***Caso clínico:*** *Paciente femenino de 25 años, con antecedente patológico de amenorrea primaria y antecedente quirúrgico de hernioplastía inguinal izquierda, con historia de protrusión en región inguinal derecha de tres años de evolución, que aumenta al realizar esfuerzo físico. Al examen físico se observó protrusión en región inguinal derecha aproximadamente de 5 cm de diámetro con Valsalva, sin cambios de color o inflamatorios, a la auscultación presentó ruidos intestinales normales, a la palpación se detectó masa en fosa iliaca derecha de consistencia blanda, depresible, dolorosa, y reducible. En el examen ginecológico, paciente con fenotipo femenino y características sexuales secundarias normales presentó labios mayores y menores simétricos, conducto vaginal permeable. El diagnóstico clínico fue hernia inguinal derecha no complicada, se decidió tratamiento quirúrgico, y se programó para cirugía electiva que encontró hernia inguinal derecha, saco herniario que contiene el útero, trompa de Falopio y ovario ipsilateral.*

* General Practitioner.
‡ Medical trainee with a master's degree in Public Health.
§ Medical Specialist in General Surgery.
¶ Medical Specialist in Gynecology and Obstetrics.

Honduran Institute of Social Security, Honduras.

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INTRODUCTION

A hernia is a defect in the continuity of the structures of the fasciae, muscles, or aponeurosis of the abdominal wall, which allows the protrusion of structures that do not usually pass through them.[1]

Abdominal wall hernia is a frequent pathology; the prevalence is estimated to be close to 5% in the general population; the most frequent presentation is of the inguinal type, close to 70% of the total, which in turn is 25 times more frequent in men.[2] The presence of the uterus and adnexa in the sac of an inguinal hernia is an infrequent event, with less than 1% of cases.[3]

Mayer-Rokitansky-Küster-Hauser syndrome (MRKH syndrome) was first described by Mayer in 1829, followed by Rokitansky in 1838, Küster in 1910, and Hauser in 1961.[4] It is a rare congenital anomaly of the genital tract of unknown etiology. It usually presents primary amenorrhea in adolescent females with normal external genitalia and growth. It may be associated with other alterations, especially at the genitourinary level.[5]

It is a rare disease that affects one in 5,000 women, whose main symptom is the absence of menarche of unknown origin, but characterized by the convergence of multiple factors, among which a probable genetic cause, the lack of sex hormone receptors in the Müllerian ducts, as well as a deficit of the enzyme galactose-1-phosphate uridylyltransferase are not ruled out.[6]

Due to the infrequency of this pathology, we present the case of a 25-year-old woman who underwent elective surgery, finding the uterus, fallopian tube, and right ovary as contents of the hernial sac, together with bilateral renal ectopia, with the kidneys being in the pelvic cavity.

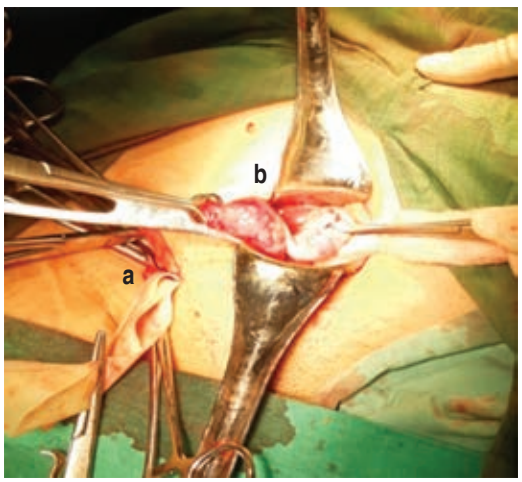

Figure 1: a. *Incision in the right inguinal region (general surgery) for utero inguinal hernioplasty.* ***b.*** *Pfannenstiel incision (obstetrics and gynecology) for revision of pelvic organs.*

CLINICAL CASE PRESENTATION

A 25-year-old female patient from rural Honduras with a pathologic history of primary amenorrhea without previous evaluation and left inguinal plasty 13 years ago. She reports protrusion of a mass in the right inguinal region after three years of evolution, which increases with physical exertion and decreases with rest.

Physical examination revealed a hypochromic hypertrophic scar in the left inguinal region, an increase in volume in the right inguinal region of approximately 5 cm in diameter, with no change in color when performing the Valsalva maneuver, soft, depressible, painful, and reducible consistency. On gynecological examination, she had a female phenotype, symmetrical labia majora and minora, and a permeable vaginal canal. Laboratory studies were within normal parameters.

Surgical management was proposed, which she accepted. A longitudinal incision was made in the right inguinal region identifying an indirect inguinal hernia. A hernial sac was dissected, and its content was a hypoplastic right uterus, hypoplastic right fallopian tube, and ipsilateral dystrophic ovary, as shown in *Figures 1 and 2*. Due to the findings, joint management by gynecology was requested, who performed an abdominal exploration using a Pfannenstiel incision. The presence of the left ovary in the pelvic cavity was confirmed; digital release and reduction of the hernial content were performed, with reincorporation of the hernial content to the abdominopelvic cavity and subsequent inguinal repair with placement of polypropylene prosthetic mesh with Lichtenstein technique.

Figure 2: a. *Hypoplastic uterus.* ***b.*** *Right fallopian tube.* ***c.*** *Dystrophic ovary.*

The postoperative diagnosis was a right inguinal hernia with the uterus, fallopian tube, and ipsilateral ovary, integrating a Mayer-Rokitansky-Küster-Hauser syndrome.

An abdominal ultrasound was performed to identify bilateral renal ectopia in the pelvic cavity to complete the diagnostic approach and rule out other associated anatomical alterations. She was discharged 48 hours after the procedure with a 30-day follow-up in the outpatient clinic, where she was found with an adequate evolution. She is currently under follow-up by gynecology for the management of Mayer-Rokitansky-Küster-Hauser syndrome.

DISCUSSION

Anatomically, the female inguinal canal has two main contents: *gubernaculum ovarii* and *processes vaginalis*. The former is a ligamentous structure that attaches to the uterine *cornua*. The latter is a small evagination of the parietal peritoneum that is typically effaced by the eighth month of embryological development.[7] Müllerian ducts give rise to the uterus, fallopian tubes, and upper two-thirds of the vagina; the renal system forms from Wolf's ducts. The Müllerian ducts stop their development around the fifth week of gestation and are vulnerable to alterations[8] such as aplasia of the Müllerian ducts.[9]

The presence of the uterus within the hernia sac and the uterine adnexa is a rare pathology in an inguinal hernia; it appears as a palpable and asymptomatic inguinal mass that occurs early in life.[4,10,11] Uterine adnexa are found in up to 31% of inguinal hernia sacs in girls, but as age advances, the frequency decreases; therefore, it is a rare finding in an adult female.[7,12]

Mayer-Rokitansky-Küster-Hauser syndrome can be of two types: type I is associated with the isolated absence of the proximal two-thirds of the vagina. Type II is characterized by malformations such as aplasia or hypoplasia of Müllerian ducts, renal ectopia or agenesis, and dysplasia of cervicothoracic somites.[9] This case was associated with type II because it coincides with its anatomical features, hypoplastic uterus, and vagina associated with bilateral pelvic ectopic kidneys.

Currently, this syndrome is the second most common cause of primary amenorrhea,[9] and the first cause is gonadal dysgenesis.[13]

In 2010 Chacón-Barboza published a Mayer-Rokitansky-Küster-Hauser syndrome in a 27-year-old woman with female phenotype, breasts, and pubic hair with Tanner stage 4. In the gynecological examination, the vulva was macroscopically normal, and the vagina consisted of a closed pouch with a depth of 2 cm, without evidence of a uterine cervix.[14]

Riggall and Cantor reported the first case of inguinal hernia containing the uterus in 1980 in a woman with a female karyotype and phenotype; however, the hernial sac had only the uterus. An anatomic anomaly with primary weakness of the uterine and ovarian suspensory ligaments is suspected. Thomson offered a hypothesis: if there is a failure of fusion of the Müllerian ducts leading to excessive mobility of the ovaries plus non-fusion of the uterine *cornua*, the possibility of herniation of the entire uterus, ovary, and fallopian tube into the inguinal canal increases.[3]

On the other hand, Fowler theorized that elongated ovarian suspensory ligaments were a hernia's primary cause or secondary effect. Okada et al. suggested that weakness of the broad or ovarian suspensory ligaments may contribute to the inguinal ring herniation, which is exaggerated by increased intra-abdominal pressure.[3,15]

An inguinal hernia with ovarian contents is not at risk for compression of its blood supply but torsion and infarction. Therefore, management aims at preserving ovarian function by repositioning the gonad to ensure an adequate source of oocyte and estrogen production. Repositioning and herniorrhaphy are advisable as soon as the condition is recognized, regardless of the Mullerian status. An open or laparoscopic approach can perform this repositioning.[4]

The management of Mayer-Rokitansky-Küster-Hauser is multidisciplinary, involving gynecological and psychological aspects. Psychological counseling is necessary to mitigate the emotional effects. Non-surgical creation of the vagina is the most common method, with daily manual self-dilation of the vaginal dimple. Surgery is considered in patients with failure of manual self-dilation or in patients who prefer the surgical creation of a vaginal canal to allow sexual intercourse. Surgical intervention in cases of inguinal hernia should be timely to prevent and relieve torsion, restore normal perfusion to the adnexa, and prevent subsequent infertility.

CONCLUSION

The management of an inguinal hernia should be adjusted to what is described in the different clinical guidelines; in the case of infrequent findings of the contents of the hernia sac, such as a hypotrophic uterus and adnexa, it is necessary to suspect rare pathologies and involve the gynecology department in the management. The diagnostic and therapeutic approach to Mayer-Rokitansky-Küster-Hauser syndrome is multidisciplinary.

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Correspondence:
Luis Enrique Sánchez-Sierra
E-mail: luensasi90@hotmail.com