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CASE REPORT PEER REVIEWED | OPEN ACCESS

Acute appendicitis and hemorrhagic ovarian cyst in a patient with Mayer–Rokitansky–Küster–Hauser syndrome: A case report

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ABSTRACT

This case reports the strange coincidence of a patient with a previous diagnosis of Mayer–Rokitansky–Küster–Hauser syndrome (MRKHS), who was diagnosed with acute appendicitis in the emergency service; even more rare, a hemorrhagic ovarian cyst is described in laparoscopic findings. Therefore, MRKH type I syndrome is confirmed. There is no evidence in the bibliography of these three pathologies at the same time in a single patient. The case was resolved laparoscopically without complications and an acute suppurative appendicitis and an infarcted hemorrhagic corpus luteum cyst were reported in the histopathological study.

Keywords: Acute appendicitis, Hemorrhagic ovarian cyst, Laparoscopic, Mayer–Rokitansky–Küster–Hauser syndrome

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INTRODUCTION

Incidence of MRKHS is 1 in 4500–5000 born alive women [1], it has an hereditary pattern and it is classified in two types: type I or typical with hypoplasia or absence of uterus and the proximal two-thirds of the vagina, generally with a normal phenotype and functional ovaries, which corresponds to the patient presented in this case; and type II or atypical which includes congenital cardiac malformations [2, 3].

Acute appendicitis is one of the most common causes of lower abdominal pain, with a higher frequency in younger age groups (40–57%). The clinical diagnosis can be challenging. It is based on clinical, laboratory, and radiological findings; and it could be improved by clinical scoring systems. Laparoscopic appendectomy is the most effective surgical treatment [4].

No reported information of these two pathologies has been found in the same patient; however, in this case, another diagnosis was established: a hemorrhagic luteal cyst. Furthermore, one of the advantages of laparoscopic intervention in appendicular pathology becomes evident in this report, since it was possible to resolve the two pathologies in the same procedure, and the MRKH

syndrome was confirmed laparoscopically.

CASE REPORT

The case of a 22-year-old female patient with a history of MRKHS is presented, who attended the emergency service for abdominal pain lasting approximately 24 hours, located in the mesogastrium, which later migrated to the right iliac fossa with a 7/10 intensity on the visual analogue scale (VAS) for pain assessment, without apparent cause. The patient did not refer presenting this pain previously. It was accompanied by nausea that causes vomiting for one occasion, also anorexia and thermal rise were seen. The physical examination showed positive appendicular signs. In the laboratory tests, the following results were seen: leukocytes 11.3 × 10^3/uL, neutrophils 70%, hemoglobin 15.1 g/dL, hematocrit 47.3%, and C-reactive protein 15.17 mg/L. Additionally, abdominal ultrasound in the right iliac fossa reported anon-compressible tubular loop image, compatible with appendicitis.

The clinical history and the laboratory results were analyzed, and the patient had an adult appendicitis score (AAS) = 15, indicating an intermediate risk of appendicitis, additionally, ultrasound found an image compatible with appendicitis and no report of ovarian pathology, therefore acute appendicitis was diagnosed.

A laparoscopic appendectomy was performed, finding an appendix in suppurative phase (Figure 1). The presence of hematic fluid was evident, with an approximated volume of 50 mL. Furthermore, a right ovarian cyst with laminar bleeding covering it was found (Figure 2). A cystectomy with hemostasis control was also performed. No complications were reported. The post-surgical evolution was adequate, so discharge was decided 24 hours after the procedure. The histopathological study reported a diagnosis of acute suppurative appendicitis and an infarcted hemorrhagic corpus luteum cyst. The patient evolved clinically and surgically in an appropriate way.

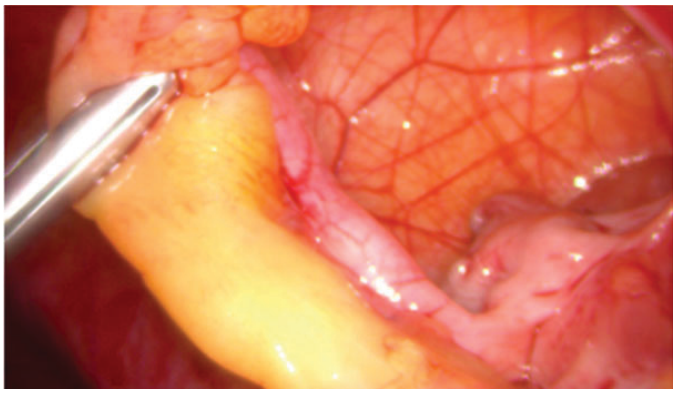

Figure 1: Laparoscopic image of phase I appendicitis.

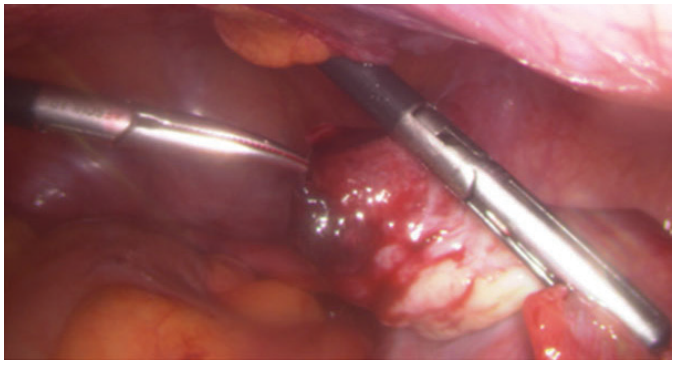

Figure 2: Laparoscopic image of a hemorrhagic ovarian cyst.

Figure 3: Laparoscopic image of the pelvis, with absence of uterus.

DISCUSSION

Mayer–Rokitansky–Küster–Hauser syndrome, also known as Müllerian agenesis, is presented by alterations of structures derived from the Müllerian ducts, also called Müllerian structures, during development [1]. It consists of two paramesonephric ducts located laterally in the urogenital crest and ending in the Müllerian eminence of the primitive urogenital sinus [5]. It manifests with hypoplasia or absence of the uterus, cervix, and upper two-thirds of the vagina in women with a 46XX chromosomal pattern, with normal ovarian function and phenotypic development [2]. Its main manifestation is primary amenorrhea.

The incidence of MRKHS is approximately 1 in 4500–5000 newborn baby girls [1]. It is considered the most severe malformation of the female reproductive tract, and can be classified in type I or typical, having the previously manifested characteristics or associated structural abnormalities, mainly in the renal or skeletal muscle system; and in type II or atypical, different authors include the Müllerian aplasia, renal aplasia, and cervicothoracic somite dysplasia (MURCS) variant in this group, which includes cardiac malformations [3, 6–8]. A different genetic subtype is also described, associated with mutations in the WNT4 gene in which a state of hyperandrogenism occurs.

Etiology of MRKHS is not fully established, and is characterized as heterogeneous, with a hereditary component. Since both sporadic and familial cases are

reported, in which the expression and penetrance is incomplete, it seems to have an autosomal dominant type [1]. Some genes have been related, such as HOXA7 [9], HOXA13, PBX1 [10], AMH, RBM8A, and TBX6 [11]. However, sufficient evidence on abnormalities of these genes and the development of the syndrome has not been established yet [8].

Acute appendicitis is the most common cause of low abdominal pain in the emergency setting and the most frequent cause of acute abdomen in young patients [12]. The lifetime risk of suffering from MRKHS is 9% in the United States of America [13]; in other guidelines it is mentioned in a lower percentage, 8.6% in men and 6.7% in women [4].

Mortality risk in cases of acute non-gangrenous appendicitis is less than 0.1%, it increases to 0.6% when it is gangrenous and to 5% when it is perforated [12]. It is recommended that clinical assessment scales should be used to diagnose the need of complementary imaging tests. The appendicitis inflammatory response (AIR) score and the AAS are recommended [12]. Evidence recommends the use of a combination of clinical parameters and ultrasound to improve diagnostic sensitivity and specificity [12]. It is also essential to rule out adnexal gynecological pathology, especially in young women.

Treatment should be performed using a laparoscopic appendectomy as it has a lower incidence of surgical site infection and is associated with less post-intervention morbidity and shorter hospital stay compared to an open approach [12].

Hemorrhagic ovarian cyst is formed by bleeding within a follicular or luteal cyst, its main manifestation is abdominal pain and it is also one of the main differential diagnoses. Most are functional and benign, very few are neoplastic; their management is surgical if they are greater than 5 cm in diameter, in the presence of persistent severe abdominal pain, spontaneous lack of resolution, or absence of benign characteristics by ultrasound [14].

A case of a patient with MRKHS is presented, due to the low frequency with which this syndrome occurs, in addition to the possibility of documenting the images during the laparoscopic appendectomy performed In a patient who presents MRKHS (Figure 3). Cases of patients with MRKH syndrome presenting acute appendicitis and hemorrhagic ovarian cyst have not been found in the literature review.

CONCLUSION

Mayer–Rokitansky–Küster–Hauser syndrome is rare and even more if it is associated with acute appendicitis and hemorrhagic ovarian cyst. The importance of laparoscopic treatment in the setting of acute abdomen for acute appendicitis is now the gold standard treatment since it is technically less invasive and allows resolution of other intra-abdominal pathologies if warranted, in addition to other already known advantages.

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Author Contributions

Michelle V Peñaherrera – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically

for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jhony A Delgado – Conception of the work, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ruth A Rojas – Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Oscar P Villacres – Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Diego M López – Conception of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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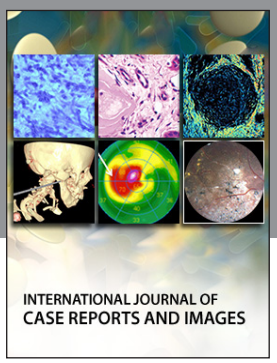

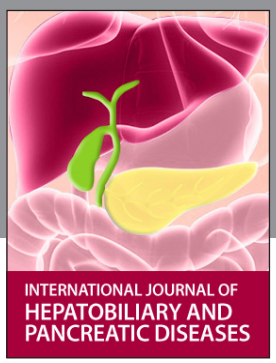

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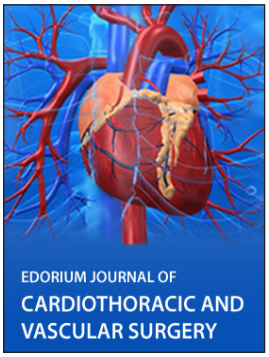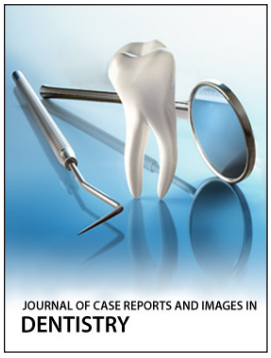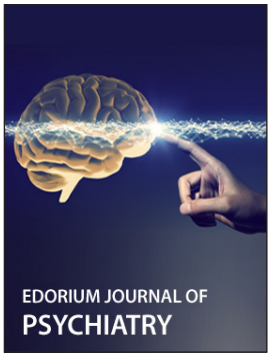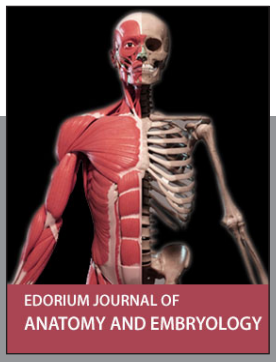
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