Al-Kindy College Medical Journal (KCMJ)

Case Report

Unicornuate Uterus with a Non-Communicating Rudimentary Horn in a Palestinian Patient with Familial Mediterranean Fever: Case Report

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Article history:
Received 1 January 2023
Accepted 20 February 2023
Available online 30 April 2023

https://doi.org/10.47723/kcmj.v19i1.936

Keywords: Mullerian Duct Anomalies, familial Mediterranean fever

ABSTRACT

Rudimentary non-communicating functional uterine horn with unicornuate uterus, originating from anomalous embryological development of one Mullerian duct, is prone to different complications either at the gynecological or obstetrical level such as chronic pelvic pain, hematometra, subfertility and decreased quality of life. This unique case report presents a 14-year-old female with a history of severe chronic pelvic pain. She was diagnosed with Familial Mediterranean Fever (FMF) and had an appendectomy for suspected appendicitis within the symptoms' interval. Ultrasound showed a right 5*6 cm right complex cystic mass assuming ovarian in place. She underwent a suspected endometrioma cystectomy operation and was diagnosed with left unicornuate uterus with right functional non-communicating rudimentary horn. The patient was followed up and mentioned marked improvement of her previous pain attacks. Her family members observed marked improvement in her usual daily activities and quality of life. Occult non-communicating uterine horn are frequently misdiagnosed due to its rarity and unspecific symptoms. The diagnosis of Mullerian anomalies should be added to the differential diagnoses for women with infertility, chronic abdominal and pelvic pain, and dysmenorrhea to avoid the patients' agony and to alleviate their quality of life.

Introduction

Unicornuate uterus with rudimentary horn is a rare Mullerian anomaly results from abnormal development of one or two paramesonephric ducts which exists in 0.2% of fertile woman and 0.6% of infertile woman. (1) Type IIb (II: for unicornuate. b: for non-communicating) represents our case when there is a non-communicating horn (American Fertility Society,1988), causing the patient to suffer from several gynecological and obstetric complications such as pre-term delivery that is partially avoided by the resection of the rudimentary horn. It is also the most common unicornuate subtype and, due to the functioning endometrial cavity inside the horn, the subtype that causes the most significant clinical sequelae. (2)

Case Presentation

A 14-year-old, nulligravid female previously diagnosed with Familial Mediterranean Fever (FMF) carrying heterozygous mutations in C2177T>C (M694I) ,C2082G>A (V726A), sought medical help due to debilitating dysmenorrhea and abdominal pain

for 6 months, dull aching in nature, gradual in onset and progressive to which the pain sometimes was not relieved even with opiates analgesia. The pain is not associated with nausea, vomiting or fever. Her menarche was at age of 13 with regular menstrual period every 28 days, for five days with average amount of menstrual bleeding. She was on combined estrogen (ethinyl estradiol) and progestin (drospirenone) pills for 2 months to alleviate the pain. The patient's past surgical history consists of a previous open appendectomy in 2020. Blood test was performed on the 30th of January 2021 and the following results were obtained; Alpha Feto-protein was normal (1.19 ng/mL), CA-125 was elevated (516 U/mL) with normal Lactate Dehydrogenase (271 IU/mL). The patient was admitted to Al-Makassed Hospital on 9th February 2021 for the laparoscopic resection of the complex ovarian cyst.

On admission, during examination the patient was well, not in pain, abdominal examination was normal. BMI was 18.26 kg/m2 (underweight). Complete Blood Count showed mildly elevated platelets count (465k/mL), but otherwise normal. Beta HCG was negative. Urine analysis showed few bacteria with WBCS of (8/HPF). She underwent an abdominal ultrasound that showed right complex ovarian cyst 5*6 cm with thick wall and small projections (Figure1).

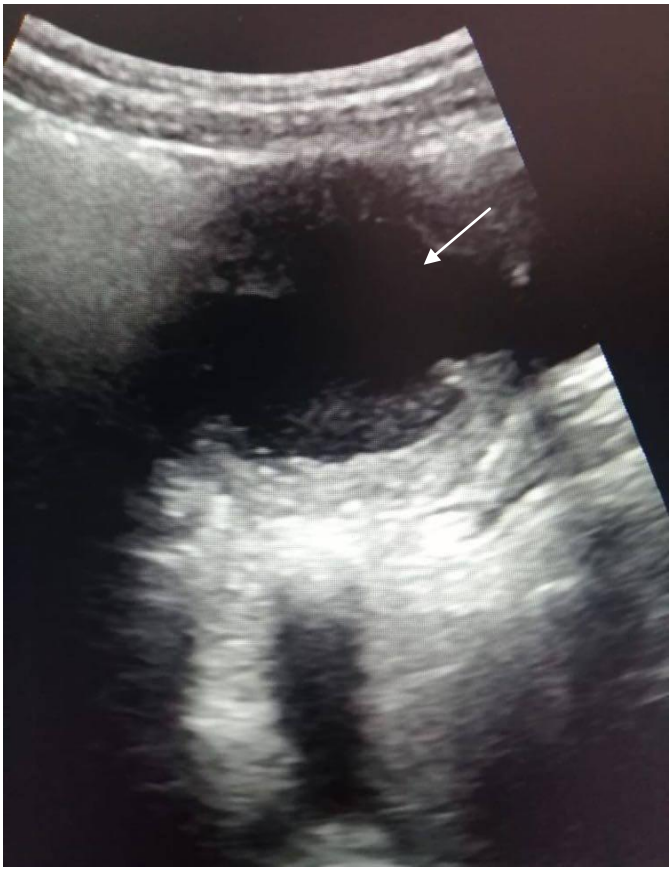

Figure 1: The functional rudimentary non-communicating uterus under ultrasound.

Normal left ovary and normal size uterus were observed. Another abdominal ultrasound was performed earlier at a private clinic on 24th December 2020 showed both kidneys were normal in size, shape and echo pattern with normal corticomedullary differentiation. There was no kidney stone, no ureteric stone and no hydronephrosis. Urinary bladder was well defined, no filling defects, no wall thickening. It also showed no free fluid, no thickening of the bowel loops and no abnormality in liver, gallbladder, pancreas and spleen.

On 10th February 2021, laparoscopy was performed at our hospital for ovarian cystectomy. During the laparoscopy, there was a right non-communicating rudimentary horn of uterus with left unicornuate uterus to normal ovaries, tubes, cervix and vagina (Figure 2).

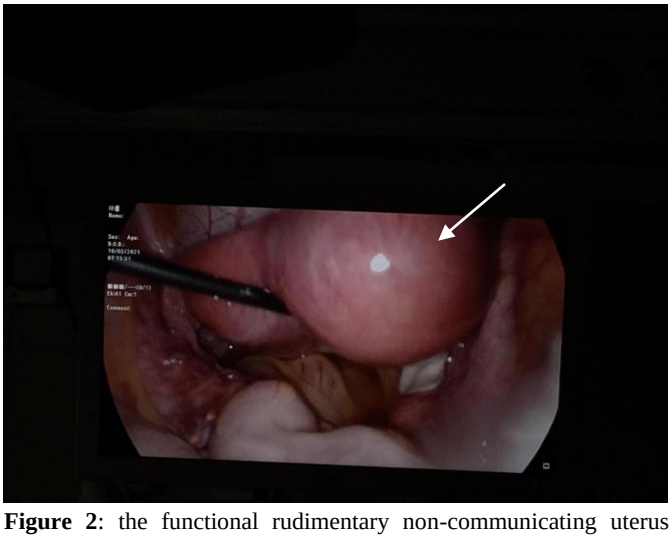

Figure 2: the functional rudimentary non-communicating uterus under laparoscopy.

With the new findings, the plan was to do an open lower transverse laparotomy for excision of the rudimentary horn with right salpingectomy after the family of the patient was addressed and consent was taken (Figure 3,4).

Postoperatively, the patient was clinically stable. Intravenous fluids, antibiotics and opioids were administered.

The patient was then seen one week later. She was in good general condition. The stitches were removed then with no complications. She was followed up for a month duration after the operation, no pain attacks occurred at all, despite her family members mentioning non-compliance of the patient to colchicine medications for a previously diagnosed FMF. The final pathology report of the excised fallopian tubes and myometrium confirmed normal unremarkable histology. Horn's endometrial biopsy showed decidualized endometrium. After laparotomy, regarding her FMF, she did not have any previous history of recurrent fever, chest pain, arthritis/arthralgia, myalgia, or erythema, though she claims that she is not compliant with her Colchicine therapy. Her next period was regular with normal amount and the menstrual pain was only in the first day.

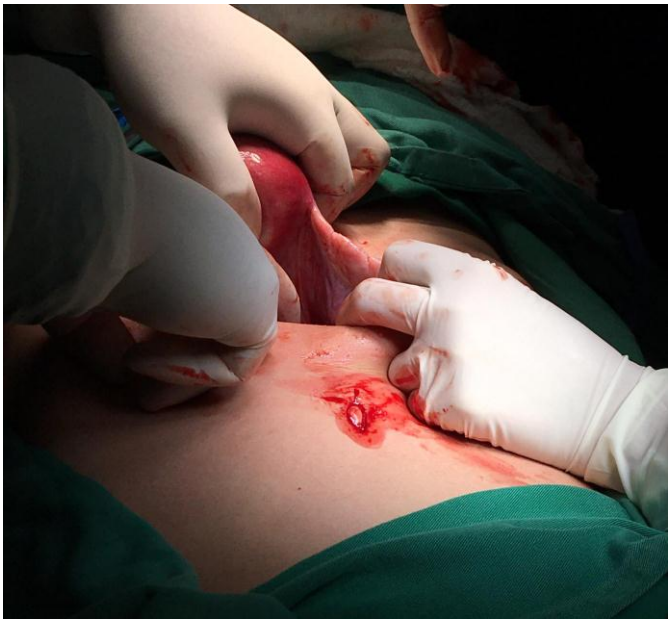

Figure 3: the functional non-communicating rudimentary horn in the laparotomy.

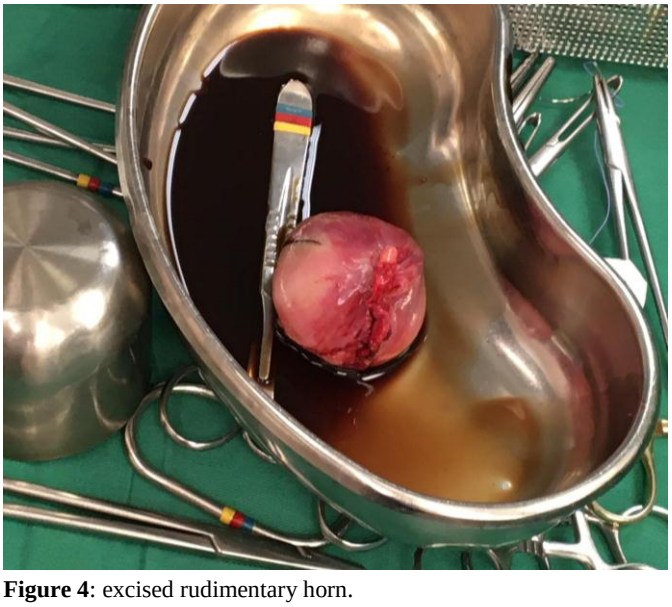

Figure 4: excised rudimentary horn.

Discussion

During embryological development, uterus is formed by the fusion of the two paramesonephric (Mullerian) ducts, and the complete process of forming the uterus is completed by the 22nd week of gestation. However, in the case of unicornuate uterus, one Mullerian duct partially or totally fails to elongate since the 9th week of gestation, and some studies showed the connection in same case that it can exist as a result of agenesis in the urogenital ridge causing a coexisting ipsilateral ovarian agenesis. (3,4,5)

The American fertility society (AFS) classified the case of non-communicating rudimentary horn with unicornuate uterus as IIb. A study analyzing the pregnancy outcomes of unicornuate class showed that 54.2% live births and 34.4% spontaneous abortions. (6)

Due to the interconnectedness of urogenital sinus and Mullerian ducts, there is a probability that an associated urologic anomaly could be present with the unicornuate uterus in (44%). Among them, 67% of the cases are associated with ipsilateral renal agenesis. (7) It could also be associated with VACTERL malformations. (8)

Non-communicating horns are the most common subtype of unicornuate uterus, and it shows the most clinical complaints due to the obstruction which causes hematometra, chronic pelvic pain and increases the risk of endometriosis. (2) Some complications related to pregnancy include uterine rupture, missed abortion and transperitoneal sperm migration into the fallopian tube of the non-communicating horn is theorized to be the reason of ectopic pregnancy in this case. (9,10,11)

Surgical resection is indicated in this case for symptomatic relief and facilitating a better obstetric prognosis. Sometimes, the rudimentary horn is connected to the unicornuate uterus with myometrium which needs resection before the horn is removed.

We observed cases of appendectomy in similar cases of right non-communicating horn before its diagnosis, so the need to consider a Mullerian anomaly in the differential diagnosis of young female patients with right iliac fossa pain before proceeding with doing appendectomy is encouraged. (12,13).

Conclusion

It is recommended to include the diagnosis of Mullerian anomalies in young female patients complaining of dysmenorrhea, chronic attacks of abdominal pain to the differential diagnoses to avoid delayed or improper diagnoses. Familial Mediterranean fever could be one of the misdiagnoses in our patient age group, so it is recommended for the patient to be evaluated clinically to confirm that the symptoms are not caused by any concomitant disorder. All patients with Mullerian anomalies should also be screened for associated abnormalities in urinary tract due to embryological interconnectedness. The patient should be counselled thoroughly about possible obstetric complications in the future such as subfertility, preterm deliveries, increased miscarriage rate, uterine rupture, malpresentation and increased need for cesarean section.

Consent for Publication

The patient and her guardians were informed and consent taken for the pictures and publication. Written consent was sent to the editor of the journal.

Funding

This research did not receive any specific fund.

Conflict of Interest

No conflict of interest

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To cite this article: Darras O, Abu Osba A, Massarweh K. Unicornuate Uterus with a Non-Communicating Rudimentary Horn in a Palestinian Patient with Familial Mediterranean Fever: Case Report. Al-Kindy College Medical Journal. 2023;19(1):127–30.