European Journal of Radiology Open 7 (2020) 100215

Contents lists available at ScienceDirect

European Journal of Radiology Open

journal homepage: www.elsevier.com/locate/ejro

Inguinal herniation of left ovary, fallopian tube and rudimentary left horn of bicornuate uterus associated with type 2 Mayer-Rakitansky-Kuster-Hauser (MRKH) syndrome in a teenage girl: A case report and literature review

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ARTICLE INFO

Keywords:
Uterus
Ovary
Inguinal
Imaging
Hernia

ABSTRACT

The incidence rate of ovarian herniation in the inguinal canal is 3 %. Nevertheless, the herniation of ovary with uterus is extremely scarce. Most of these cases are accompanied with congenital genitourinary malformations such as Mayer-Rakitansky-Kuster-Hauser (MRKH) syndrome and Mullerian duct malformations. Herein, a 13-year-old girl presented with type2 MRKH syndrome together with left groin swelling and pain. Sonography reports ovarian herniation into left inguinal canal and magnetic resonance imaging (MRI) confirmed the diagnosis and demonstrated the left ovary, fallopian tube and rudimentary left horn of bicornuate uterus in the left inguinal canal accompanied with ectopic renal of left lower quadrant. Finally, the patient was designed for surgical inguinal exploration and left normal ovary, fallopian tube and rudimentary left horn of bicornuate uterus were replaced in the pelvis.

1. Introduction

Inguinal hernia (IH) is the most common hernia in adult population but it is not common in female [1]. IH sac usually contains omentum, appendix and urinary bladder may herniate into inguinal canal [1]. However, ovarian herniation to IH is scarce in adult female, it is more common in infants and pediatrics [1], but protrusion of uterus to IH sac is very uncommon [2]. In herniation of ovary, fallopian tube and uterus congenital genitourinary tract anomalies such as Mayer-Rakitansky-Kuster-Hauser (MRKH) syndrome and Mullerian duct anomalies should be considered [1,2]. In this study, we report a teenage girl of MRKH syndrome associated with inguinal herniation of left ovary, fallopian tube and rudimentary left horn of bicornuate uterus

2. Case presentation

A 13-year-old female with history of swelling and pain in left groin referred to our radiology department for further assessment. On physical examination revealed a mass over the left groin which was not tender or warm. On sonography, there was herniation sac containing ovarian-like structure and magnetic resonance imaging (MRI) suggested for further evaluation. MRI shows left ovary with normal size and shape, left fallopian tube and rudimentary left horn of bicornuate uterus lie in left inguinal canal. Normal right ovary and right horn of bicornuate uterus were also noticed in pelvis (Fig. 1 and 2).

Surgical examination of the left inguinal canal indicated an oblique hernia and after was open the hernia sac and it was included the left ovary and the left fallopian tube, along with left uterine horn and then, the left uterine horn in the left inguinal canal was well developed and finally, left inguinal canal was explored and normal left ovary, left fallopian tube and rudimentary left horn of bicornuate uterus were replaced in pelvis, and the indirect defect was shut by 0 nylon (Fig. 3)

Due to the association of this type of herniation with congenital genitourinary tract anomalies such as MRKH syndrome and Mullerian duct anomalies, she underwent further accurate evaluations. In secondary evaluation she had history of primary ameorrhoea and in physical examination the patient had female phenotype, normal stature and normal development of secondary sexual characteristic.

On the other hand, abdominal and pelvic imaging was performed for detection of other probable anomalies and the only abnormal finding was ectopic renal of left lower quadrant

Overall, this patient had primary amenorrhoea, normal phenotype and normal development of secondary sexual characteristic, who presented with left inguinal ovary, fallopian tube and rudimentary left horn of bicornuate uterus which are compatible with MRKH syndrome. Furthermore, because of its association with ectopic renal of left lower

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https://doi.org/10.1016/j.ejro.2020.01.004
Received 7 January 2020; Accepted 17 January 2020
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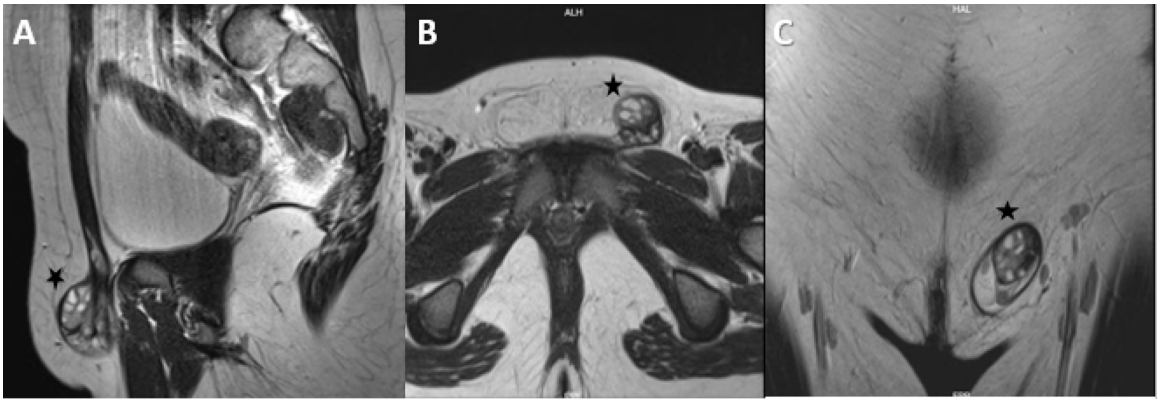

Fig. 1. sagittal (A) axial (B) and coronal (C) T2W MRI shows left ovary with normal size and shape in left inguinal canal (black star).

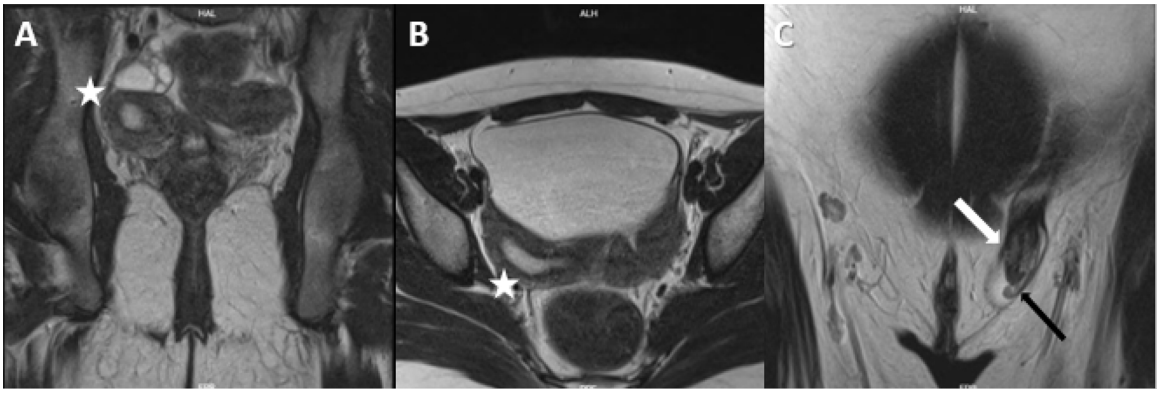

Fig. 2. coronal (A) and axial (B) T2W MRI shows normal Right ovary and Right side cornuate of uterus (white star), and coronal T2W MRI shows left fallopian tube (black arrow) and rudimentary left cornuate of uterus (white arrow) in left inguinal canal.

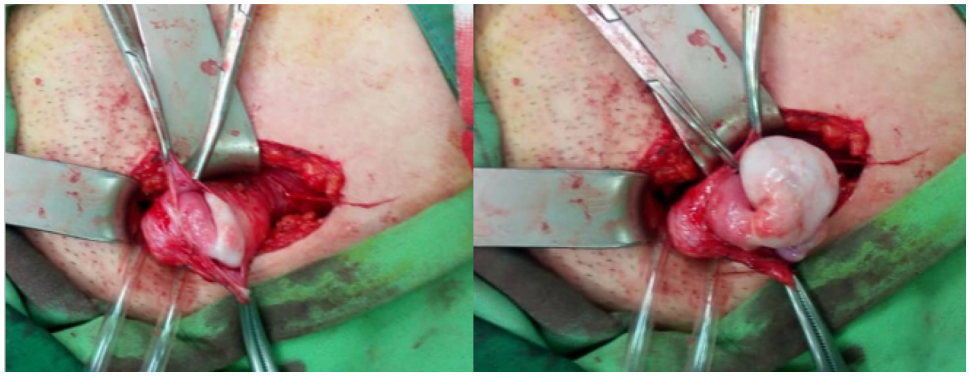

Fig. 3. pictures from operation room shows normal left ovary, left fallopian tube and rudimentary left cornuate of uterus in left inguinal canal.

quadrant, it is considered as type 2 of MRKH.

In patient follow up, on the fourth postoperative day, she was discharged and was visited in the clinic 3 weeks after postoperative and the patient was no any complication and all parts e.g. inguinal and vaginal wounds were healthy as well as the role of clinical management in maintaining ovarian function with gonad reuse is considered to provide an sufficient source of oocytes and estrogen production.

3. Discussion

The inguinal canal is a 4 cm narrow, diagonal passage in the lower anterior abdominal wall and is lined by the aponeurosis of the external oblique, internal oblique and transverse abdominis muscle. Deep inguinal ring is in transverse fascia and the superficial one is in the aponeurosis of external oblique muscle [3]. In female embryos, gubernaculum adheres to the ovary from its cranial terminal and to the inner surface of the labioscrotal fold from the caudal terminal. Its midpoint attachment to the uterus barricades the ovary from descending into the inguinal canal [4]. IH containing ovary is not scarce in in female infants and pediatrics, but herniation of uterus is extremely rare [1].On the other hand, MRKH syndrome is distinguished by congenital absent or rudimentary bipartite uterus, aplasia of the upper two-thirds of vagina with normal ovaries, fallopian tube and secondary sexual specifications [1,5].

In this report, we present an utero-ovarian herniation into left inguinal canal in a teenage girl with type 2 MRKH syndrome. These cases should be considered because of two important aspects. First, because of probable associations to other anomalies general physical examination and evaluation should be done. Al Omary et al., reported a young woman with right utero-ovarian inguinal herniation with MRKH

syndrome who also was missing the left kidney [7]. Other point, for preventing ovarian complications such as torsion and also preserving fertility, replacing IH sac should be done as soon as possible.

Very few cases of ovarian herniation with and without fallopian tube and uterus in MRKH syndrome have been reported. For example Khan et al., 2019 [6] and Mohanty et al., 2017 [1] reported cases of MRKH syndrome with inguinal herniation of ovary. Bazi et al. et al.,2006 presented the bilateral inguinal ovarian herniation in a young girl with Mullerian agenesia and this literature also reviewed some other similar literature of MRKH syndrome and Mullerian agenesia with ovarian herniation accompanied with uterus either normal types or rudimentary one [5].In addition, Graul et al., 2014 mentioned another conditions that leads to adnexal entrapment especially in infants having a short inguinal canal, oblique direction of canal in abdominal wall and diverticulum of Nuck [8]. Furthermore, some literatures reported inguinal herniation of ovary with or without fallopian tube and uterus into the inguinal canal regardless of congenital genitourinary syndromes. Such as Daib et al., 2017 reported a 1-month year old infant with utero-ovarian herniation into left inguinal canal [2] and another study by Ueda et al.,2016 indicated the inguinal herniation of uterus, ovary and fallopian tube in an old woman [9].

4. Conclusion

In summary, our case suggested that clinical management of such a clinical problem must be holistic including extensive counseling because of its presumptive relationships, reposition of uterus, fallopian tube and ovary for preserving fertility and preventing complications such as torsion and repairing of the hernia.

Contributors

RJ collected the data and conceived the idea of the case report and collected the data and revised the manuscript. MJ reviewed the manuscript and wrote the manuscript. FD contributed to the data and wrote the initial manuscript. All authors read and approved the final manuscript

Funding

No funding from an external source supported the publication of this case report

Patient consent

Patient and parental consent for publication were obtained

Provenance and peer review

This case report was peer reviewed.

Declaration of Competing Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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