CASE REPORT

Mayer-Rokitansky-Kuster-Hauser Syndrome Presenting as Irreducible Inguinal Hernia

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Abstract Inguinal hernia is the most common type of hernia experienced in the surgical OPD due to the muscular anatomy of the region being weak along with the natural weaknesses like deep ring with indirect hernia being more common than direct. Inguinal hernias may have varying unusual sac contents, a case presentation of rudimentary uterus and ovaries as contents of inguinal hernia in an adult female with Mayer-Rokitansky-Kuster-Hauser (MRKH) type II syndrome. This syndrome may be attributed to abnormal development of blastema of cervicothoracic somites and pronephriducts.

Keywords MRKH · Inguinal hernia · Rudimentary uterus and ovaries · Mullerian agenesis

Introduction

Mayer-Rokitansky-Kuster-Hauser (MRKH) consists of vaginal aplasia with other mullerian duct abnormalities. Type I is associated with isolated absence of proximal two thirds of the vagina, whereas type II is characterised by other malformations including mullerian duct aplasia/hypoplasia, renal agenesis/ectopy and cervicothoracic somite dysplasia (MURCS) [7].

The syndrome is transmitted as an autosomal dominant trait with an incomplete degree of penetrance and variable expressivity. It has shown to be associated with HOX gene but still under research. This was a case was of 45-year-old married female with obstructed irreducible inguinal hernia containing uterus and ovaries along with MRKH type II syndrome, whose incidence is 1:4500 female births.

Case Report

A 45-year-old female presented with a swelling in left groin gradually increasing in size with dragging pain on and off since 2 years and primary amenorrhea. Swelling was better appreciated on coughing and straining with pain aggravation. On examination, a 5 × 7 cm oval, irreducible, inguinal swelling along with tenderness was present. Per rectal examination was normal. Also, the left kidney was congenitally absent in her. The patient was moderately built, and her breast development and pubic hair pattern corresponded to Tanner stage IV.

Per vaginal examination revealed a blind vagina. Ultrasonography of abdomen and pelvis revealed a mixed echogenic lesion with no fluid in the sac. The assessment of FSH, LH and estradiol was within normal limits.

The patient was operated with an open approach, external oblique aponeurosis was opened and the sac was dissected. It showed mullerian duct or an agnetic tissue as its contents of indirect obstructed inguinal hernia with minimal tissue discoloration and haematoma. It was drained followed by Lichenstein's repair.

Histopathology reports shows microscopically of bundles and fascicles of smooth muscle cells with absence of endometrial tissue suggestive of rudimentary uterus and ovary as contents of the sac (Figs. 1 and 2).

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Fig. 1 Myometrium—histopathology slide

Discussion

Anatomically, female inguinal canal has two main contents—gubernaculum ovarii and processus vaginalis. The former is a ligamentous structure attaching to uterine cornua and latter is a small evagination of parietal peritoneum which is typically obliterated by 8th month of development [4].

The presence of uterus and ovaries in the inguinal hernia sac is rare. Although it is an unusual content, presence of appendix was first reported by de garengot in femoral hernia in 1731 [1]. Later, an enormous literature got published with presence of contents such as hydrocele of canal of Nuck, sliding hernia with both tubes and ovary, Amyand's hernia containing vermiform appendix, undescended testes, haematomas, benign and malignant tumours [2, 3].

Uterine adnexa are found in as many as 31% of inguinal hernia sacs in infant girls, but as age advances the incidence decreases; hence, it is a rare finding in an adult woman [5].

Approximately 4 to 37% cases present with irreducible ovary in inguinal hernia, while about 2 to 33% of these are complicated with torsion or infarction [6].

Ideally, MRI and karyotyping should be performed, but since the patient belonged to a poor socio economic background they could not afford one.

Hence, ultrasonography should be done routinely to diagnose contents of sac [8] and various other malformations to intervene early and give prompt treatments to such patients.

Management is aimed to preserve the contents of hernia and reposition them back into the body; since in our case contents were atrophied already, we sacrificed it and repaired the hernia.

Due to the low incidence and rarity of presentation, such syndromes go unnoticed until the patient presents with complications. With advancement in vitro fertilizations technique and surrogacy, there may be an increase of availing such options in near future. Hence, characterisations of genetic mutations associated with the syndrome play a vital role in management of the patient.

Fig. 2 Hernial sac opened with rudimentary uterus exposed

Compliance with Ethical Standards

Consent Written informed consent was obtained from the patient for publications of this case report and accompanying images. A copy of the written informed consent is available for review by the Editor-in-Chief of this journal on request.

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Consent of the Patient The verbal consent of the patient was taken regarding publishing the details of her case, keeping her identity anonymous. If the journal requires a written consent for the same, kindly let us know.

Conflict of Interest The authors declare that they have no conflict of interest.

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