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CASE REPORT

TORSION OF OVARY IN MRKH SYNDROME PRESENTING AS IRREDUCIBLE SLIDING INGUINAL HERNIA

Sneha Palepu[1], Uday Kumar[2], Md. Jawed Akhter[3], Yaramati Avinash[4]

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ABSTRACT: Inguinal hernias may have very unusual sac content. The Fallopian tube and ovary are found in 15% to 20% of the sliding hernias in females. Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, is one of the associated genital anomalies with an incidence of 1:4500 female births. We report a rare case of sliding inguinal herniation of ovary with torsion & fallopian tube in a 20 year old woman with MRKH syndrome type I. Preservation of ovarian function by repositioning the gonad followed by herniorrhaphy was done.

KEYWORDS: MRKH Syndrome, Sliding inguinal hernia, Torsion of ovary.

INTRODUCTION: Inguinal hernias may have very unusual sac content.[1,2] Sliding inguinal hernias of the uterus, fallopian tube, and ovary are rare in adult women.[3] Most cases have associated congenital anomalies of the genital tract.[4] Ovarian torsion and infarction have been encountered in 2% to 33% of the patients presenting as non-reducible groin swellings.[5] We report a case of sliding inguinal herniation of ovary & fallopian tube with torsion of the right ovarian cyst in a 20 year old woman with MRKH syndrome type I.

CASE REPORT: A 20 years old female with bilateral reducible groin swellings since childhood and primary amenorrhea presented with right groin pain for 2 days. On examination 7x5cm oval, moderately tender irreducible swelling in the right inguinal region and 3x3cm oval, non-tender reducible swelling in the left inguinal region were present. Patient was thin built with a female phenotype and normal stature. Her breast development & pubic hair pattern corresponded to tanners stage III, with sparse axillary hair.

Her external genitalia were normal. Per vaginal examination revealed blind vagina. Ultrasonography of abdomen and pelvis revealed 6x4cm mixed echogenic lesion in the right inguinal canal without vascularity and 3x2cm oval mixed echogenic lesion in the left inguinal canal without uterus and both ovaries in the pelvis. Magnetic Resonance Imaging (MRI) revealed well defined heterogeneously hyperintense T1 & contrast enhancing lesions in the bilateral inguinal canals with multiseptated T2 hyperintense lesion in the right inguinal canal [Figure 1]. Her karyotype was 46XX and her hormonal profile was normal.

Intra-operatively, a sliding hernia of right side revealed an atretic ovary with a 5x3cm congested cyst and a 12cm long rudimentary fallopian tube [Figure 2]. On the left side, a reducible sliding hernia with normal ovary and a paraovarian mass of 3x2cm was identified. The right ovary and the tube were excised followed by bilateral herniorrhaphy. Biopsy of the specimen revealed normal ovarian tissue with fallopian tube showing congested vessels and oedematous stroma with a thin walled cyst [Figure 3].

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DISCUSSION: Embryologically, female inguinal canal encompasses two main anatomic structures- gubernaculum ovarii and processus vaginalis. Gubernaculum ovarii is a ligamentous structure which attaches to uterine cornua midway along its course. Superior to this attachment, it is called ovarian ligament which prevents ovarian descent into the inguinal canal and inferior to the attachment, it is the round ligament of uterus. Processus vaginalis is a small anterior evagination of parietal peritoneum which is usually obliterated by 8th month of development.[6] Incomplete obliteration may result in indirect inguinal hernia.

An ovary in a hernia sac might be assumed to be a descended gonad, mimicking descent of the testis rather than simply prolapsed which presents as sliding hernia.[7] The ligament that lies in the inguinal hernia sac in females believed to be the round ligament of uterus is actually the suspensory ligament of the ovary.[8] It is supposed to be the female gubernaculum with altered anatomy and localization because of the absence of androgen responsiveness.[7]

MRKH is characterized by congenital aplasia of the uterus, upper 2/3 of the vagina & normal ovaries in women with normal female phenotype and normal 46, XX karyotype. Type I with isolated genital anomalies is less frequent than Type II.[9] This was a case of a 20 year old adult married woman with MRKH syndrome type I with an irreducible sliding inguinal hernia containing ovary and fallopian tube with torsion of the ovary which is extremely rare.

Strubbe et al. identified 6 among 91 MRKH patients with inguinal ovarian hernia.[10] Bazi et al. identified 7 well documented cases of MRKH with inguinal ovarian hernia, their patient became eighth.[11] An inguinal ovary is not at risk of compression of its blood supply but of torsion and infarction.[5] Management therefore is aimed at preserving the ovarian function by repositioning the gonad followed by herniorrhaphy as soon as the condition is recognized.[11]

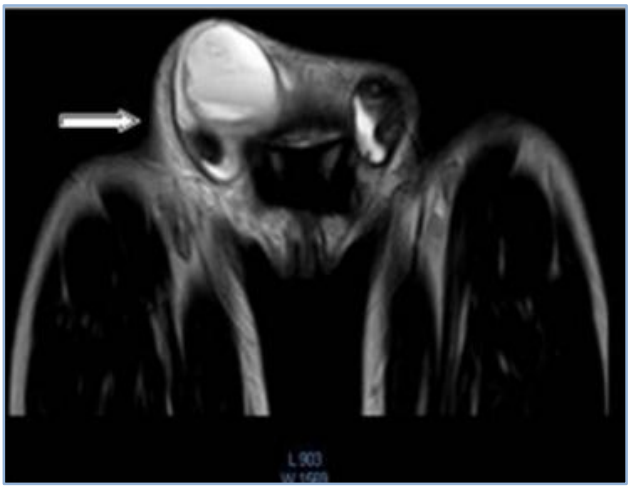

Fig. 1: (MRI of B/L Inguinal Region)

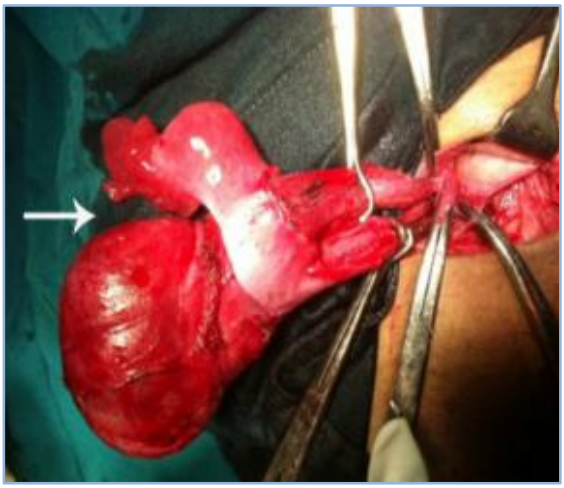

Fig. 2: (Ovary, Congested, Cyst & Fallopian Tube)

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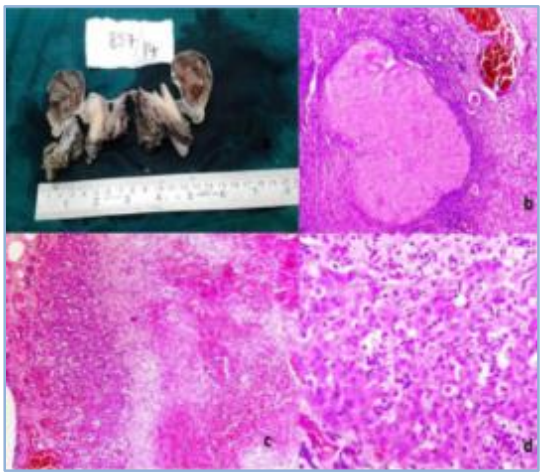

Fig. 3: (HP of Ovary, Congested Cyst & Fallopian Tube)

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AUTHORS:

1. Sneha Palepu
2. Uday Kumar
3. Md. Jawed Akhter
4. Yaramati Avinash

PARTICULARS OF CONTRIBUTORS:

1. Post Graduate, Department of General Surgery, Mamata Medical College, Khammam, Telengana.
2. Post Graduate, Department of General Surgery, Mamata Medical College, Khammam, Telengana.
3. Professor, Department of General Surgery, Mamata Medical College, Khammam, Telengana.
4. Post Graduate, Department of General Surgery, Mamata Medical College, Khammam, Telengana.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Md. Jawed Akhter,
Department of General Surgery,
Mamata Medical College,
Khammam-507002, Telengana.
E-mail: dr_mdjawedakther@yahoo.com

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