A Case Series Depicting Agenesis Of Uterine Adenexa At Different Age Groups

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Abstract: Unilateral absence of fallopian tube and ovary, unilateral ovarian agenesis are extremely rare findings to be discovered on laparoscopy. The causes for absence of ovary on one side could be as a result of defect in embryological development or asymptomatic torsion. The absence of fallopian tube on one side could be due to defect in development of the mullerian duct on the corresponding side. The absence could also be due to asymptomatic torsion with subsequent atresia. We are reporting a case series depicting uterine adenexal agenesis at different age groups.

Keywords: Fallopian tube agenesis, ovarian agenesis, Mullerian agenesis, primary infertility, appendicitis.

I. INTRODUCTION

Developmental abnormalities of the female genitalia, includes the external genitalia, vagina, cervix, uterus, fallopian tubes and ovaries. Mullerian duct abnormalities are the most commonly observed amongst the anomalies. Anomalies involving the fallopian tubes, ovaries and vagina are less frequently seen. Unilateral absence of ovary with the absence of the fallopian tube on the same side, unilateral ovarian agenesis being extremely rare finding. The exact pathophysiology is not known but could be as a result of defect in the embryological development or asymptomatic torsion of the fallopian tube and ovary.

Figure 1: depicting normal right fallopian tube and right ovary

Figure 2: Depicting absence of left fallopian tube and left ovary, a tag of tissue visible at its place suggesting adenexal torsion as the likely pathology
A TEENAGER WITH UNILATERAL AGENESIS OF IPSILATERAL OVARY AND FALLOPIAN TUBE WITH ACUTE APPENDICITIS

CASE 1: 19 years old unmarried girl presented with pain in the right iliac fossa with fever for five days. Her vitals were stable. She had come with complaints of pain abdomen in left iliac fossa with urinary tract infection and constipation, three months prior to getting hospitalized and treated. She had no menstrual problems. There is no significant history of any health problem in the past. Abdominal examination revealed severe tenderness in the right iliac fossa. There was no mass or any organomegaly. Ultrasonography showed a left ovarian cyst measuring 4.2 x 4.0 cm. Right ovary could not be visualized. CT scan of the previous hospitalization also had shown a left ovarian cyst measuring 4.6 x 4.3 x 3.8 cm with areas of calcification with an impression of dermoid cyst. Chest X ray being normal. Hemoglobin-13.0 gm%, total WBC count – 13,200 differential count – neutrophils:64%, lymphocytes:26%, eosinophils:04%, monocytes:06%, random blood sugar-120, serum creatinine-0.96, sodium- 137, potassium-3.83. Tumour markers CA-125-5.4, LDH-504, beta HCG<-2.0, alpha fetoprotein-1.3, CEA-0. A working diagnosis of acute appendicitis with a dermoid cyst of left ovary was made and the patient was subjected for laparoscopy. Laparoscopy showed flimsy adhesions present in the pelvis near adnexal areas on both sides and the appendix area. uterus and left fallopian tube normal with a follicular cyst of the ovary on the left side. Right fallopian tube and ovary were absent and appendix showed inflammation. Peritoneal washings was sent for cytology which showed chronic inflammatory exudate with mesothelial reaction. Ovarian aspirate revealed lymphocyte in haemorrhagic background. Histopathological report revealed acute appendicitis with peri-appendicitis. Laparoscopy was proceeded with appendicectomy and puncturing of the ovarian cyst wall. Intraoperative and postoperative period was uneventful.

CASE 2: INFERTILE PATIENT PRESENTING WITH UNILATERAL AGENESIS OF OVARY AND FALLOPIAN TUBE

A 26-year-old nulligravida was admitted to the Department of Gynecology with a diagnosis of primary infertility. The patient had not conceived despite regular unprotected intercourse for two years. Menarche occurred at 12 years of age, and the menstrual cycle was regular with 27–28 day intervals and a 5–6-day menstrual period without dysmenorrhea. The overall health of the patient was good and there was no history of abdominopelvic surgery. On physical examination, no surgical scars were observed. The external genitalia, vagina, cervix and uterus appeared normal on gynecological examination, and the results of sex hormone analysis were also normal. The patient’s husband submitted a semen analysis, which was within the normal limits. Transvaginal ultrasonography revealed that the uterus and ovaries (only the right ovary was visualized) were normal. Hysterosalpingography was performed which revealed a normal uterine cavity; however, the fallopian tubes did not fill bilaterally. Furthermore, genetic analysis revealed a normal karyotype (46,XX).

Subsequently, a diagnostic laparoscopy and hysteroscopy were performed. During the hysteroscopy, the endometrial cavity was observed to be normal, as well as the left and right tubal ostia. The laparoscopy revealed a single, normal-sized uterus with a smooth surface. No adhesion between the uterus and the intestinal serosa, the cecum and the pelvic wall was observed. The rectouterine pouch was inspected and no ectopic tissues were identified. The left adnexa was not completely visualized; however, a 2-cm tubal remnant with an...
intact left round ligament was observed. The right fallopian tube, right ovary (with a corpus luteum) and right round ligament were found to be normal (Fig. 1). The broad ligaments were also normal without any adhesions. In addition, the peritoneal and omental surfaces were analyzed and no ectopic tissues or remnant structures were observed. Methylen blue chromopertubation did not result in spill from the right fallopian tube and the postoperative course was uneventful.

Since adnexal agenesis often coexists with malformations of the urinary tract, abdominopelvic magnetic resonance imaging (MRI) was performed to investigate the urinary system. The MRI scan revealed that the kidneys and ureters were normal bilaterally, while the left ovary was unable to be imaged.

CASE 3: PRIMARY INFERTILITY WITH UNILATERAL OVARIAN HYPOPLASIA

A 34 years old nulligravida presented with primary infertility of 5 years duration. Her menstrual cycles being regular with unremarkable gynaecological and medical history. Her husband’s semen analysis being normal within limits by WHO criteria.HSG revealed normal uterine cavity, patent right fallopian tube and minimal patency of left fallopian tube. Patient had undergone 6 cycles of failed IUI, despite documented ovulation in all cycles of treatment.

On general physical examination- normal external genitalia, normal sized anteverted & anteflexed uterus and a normal cervix. On saline infusion sonography a normal uterine cavity with a 7mm x 5mm endometrial polyp. On transvaginal sonography left ovary was unvisualized. Lab. Investigations revealed a serum anti-mullerian hormone level of 5.2 ng/ml, FSH and s.estradiol of 5.4 mlu/ml and 40 pg/ml respectively. Laparoscopic assessment revealed normal uterus, right ovary and right fallopian tube. Left fallopian tube being normal, left ovary being atrophic i.e. maximum diameter < 10 mm. Left ovary was found to be adherent to left pelvic sidewall. Flimsy adhesions between the two were lysed. On chromopertubation bilateral tubal patency was established. Hysteroscopic resection of the endometrial polyp was done via resectoscope. 6 months following laparoscopy patient conceived spontaneously. Currently the patient is 32 weeks pregnant. Her ANC period being uneventful uptil now.

II. DISCUSSION

The unilateral absence of ovary and fallopian tube is an extremely rare finding. The causes of this still remains unclear. There are three possible hypothesis:

✔ Women with mullerian duct anomalies are more prone to ovarian torsion due to abnormal anatomic connection between ovary and pelvic side wall.

The etiologies for development of ovarian torsion include:
- Anatomic abnormalities
- Physiological abnormalities
- Hemodynamic abnormalities
- Trauma, surgery

Ovarian torsion may lead to organ autoamputation. Calcified ovarian tissue may be found free floating in pelvis during laparoscopy. In some cases parasitic ovaries have been found.

✔ The absence of any other anatomical structure and chromosomal abnormalities along with histological examination of remanant tissue is suggestive of vascular accident.

✔ A defect in development of mullerian and mesonephric system, either entirely on one side or localized to region of genital ridge and caudal part. Hox gene plays a important role in regional characterization of structures found along crano-caudal axis of female reproductive tract.

WNT7a is required for proper Hox expression and radial axis patterning of mullerian ducts.

The incidence of ovarian agenesis is estimated to be <1 %. The suggested etiologies include torsion of ovarian pedicle in fetus or early neonate resulting in ischaemia & atresia. Failure of embryological development ma also lead to ovarian hypoplasia. Ovarian torsion in adults is usually associated with abdominal pain, nausea, vomiting. Adenexal torsion might result from ovarian enlargement by tumours or cysts. Women with mullerian duct abnormalities are more prone to torsion due to abnormal anatomic connection between ovary and pelvic side wall. Ovarian torsion might lead to organ autoamputation.

III. CONCLUSION

The unilateral absence of ovary and fallopian tube is an extremely rare finding. The causes of this still remain unclear, even though congenital malformation or torsion of the ovarian pedicle are thought to be likely causes. Absence of ovary and fallopian tube on same side in our case have been due to torsion leading to subsequent atresia as she had recurrent episodes of pain abdomen in the past in first case report. Absence of other congenital reproductive organ or renal anomalies makes the hypothesis of embryological anomaly less likely in our reported cases.

REFERENCES