Four Cases with Congenital Unilateral Absence of Ovary and Fallopian Tube: Review of the Literature

Konjenital Tek Taraflı Over ve Tuba Yokluğu olan 4 Olgu ve Literatürün Gözden Geçirilmesi

Congenital Tubal And Ovarian Absence

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Özet

Yazımızda Etlik Zübeyde Hanım Kadın Hastalıkları Eğitim Araştırma Hastanesinde tanı alan konjenital tek taraflı tuba ve ovaryan yokluk ile karakterize dört vakayı sunduk: 23 yaşında adnexal kitle varlığı ile başvuran hasta, 21 yaşında adnexal mass ve kronik pelvik ağrı ile başvuran hasta, 25 yaşında adnexal kitle nedeni ile başvuran hasta ve 36 yaşında, 18 aylık primer infertilite nedeni ile başvuran hasta. Hastaların hepsine laparoskopi yapıldı: Aynı taraflı komplet tubal ve ovarian yokluk, karşı taraf overde kitle saptandı: Bu durum nekroz ve rezorpsiyonla sonuçlanabilen asemptomatik adnexal torsion, vaskuler problemler veya konjenital malformasyonlar nedenlerinden kaynaklanabilir.

Anahtar Kelimeler

Konjenital Unilateral Over Yokluğu; Konjenital Unilateral Tubal Yokluk

Abstract

We present four rare cases of congenital unilateral tubal and ovarian absence that were diagnosed in and admitted to the Etlik Zubeyde Hanım Women's Health Teaching and Research Hospital. The patients were a 23-year-old female with an adnexal mass, a 21-year-old female with chronic pelvic pain with an adnexal mass, a 25-year-old female with an adnexal mass, and a 36-year-old female with primary infertility for 18 months. All patients were evaluated by laparoscopy. The cases showed complete tubal and ovarian absence with a contralateral adnexal mass. The absence of one ovary and tube may be explained by three possible etiopathogenic causes: asymptomatic adnexial torsion that leads to necrosis and resorption, vascular problems, or congenital malformation.

Keywords

Unilateral Fallopian Tube Agenesis; Unilateral Ovarian Agenesis

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Introduction

Asymptomatic unilateral absence of a fallopian tube with or without ovarian involvement is a very rare condition. The true incidence is unknown; Sivanesaratnam estimates it as 1 in 11,241 women [1]. This rare condition can be related to three etiologic causes. The segmental torsion of the uterine tube and/or ovarian pedicle may occur for undetermined reasons during intrauterine fetal life, childhood, or adult life. Consequently, torsion may give rise to necrosis and auto-amputation. Tubal and ovarian maldevelopment secondary to ischemia due to a vascular accident may be another etiological factor. Alternatively, the absence of these organs may be congenital, associated with developmental alterations of the mesonephric and paramesonephric ducts.

In this study four cases diagnosed as the absence of ovary and fallopian tube will be presented and compared to the cases published in the literature.

Case Report

Case 1

A 23-year-old nulliparous single woman with acute pain in the left lower quadrant and dysmenorrhea was admitted to our clinic. She had menarche at the age of 11 and continued to have regular menstrual periods at 30-day intervals. The patient did not complain of nausea, vomiting, or fever. On pelvic examination, a left adnexal cystic mass of approximately 10 cm diameter was palpated. Transabdominal sonography revealed a 10x8x6 cm hypoechogenic uniloculated cystic mass in the left ovarian location. No vascularity was detected by doppler ultrasonography. The left and right ovary could not be visualized separately. Serum Ca 125 level was 35 IU/ ml. She had no history of abdominopelvic surgery. Laparoscopic surgery was planned for diagnosis and treatment of the adnexal mass. On laparoscopic evaluation, the uterus and Pouch of Douglas had a normal appearance, but there was a typical endometriotic lesion with a diameter of 1 cm on the serosal surface of the bladder. The right ovary and fallopian tube were absent. There was a thick and fibrotic band on the right side of the round ligament close to the uterus. On the left side there was a pseudocytic mass of 10 cm within a conglomerated tubo-ovarian complex. The content of the pseudocyst was drained and an excisional biopsy was carried out from the cystic component of the mass and was sent for histopathological evaluation. The endometriotic implant was biopsied and cauterized. The histopathological analysis of the biopsy specimen from the cystic mass reported normal ovarian tissue.

Case 2

A 21-year-old nulliparous single woman who applied to our clinic with chronic pelvic pain with a simple ovarian cyst of 32x30 mm that had been diagnosed 4 months earlier. Her menarche had occured spontoneously at the age of 12 and she had regular menstrual periods at 28-day intervals since then. She had no history of any gynaecological complaint and no previous operation. On pelvic examination, bilateral adnexal fullness was palpated. Transabdominal sonography revealed a 29x26mm hypoechogenic cyst with mural solid component in the left adnexal location. The right ovary was not detected by ultrasonography. Serum Ca 125 level was 71.9 IU/ml. She was diagnosed as having a persistent left adnexal mass and a laparoscopy was planned. During laparoscopy, the right round ligament was not observed, while a free avascular fibrous band extended between the uterus and the small intestine. This fibrous band ended on the serosal surface of the intestine with tissue mimicking the fimbrial end of the tube. Under this band a remnant white opaque ovarian tissue of 0.5cmx0.5cm was detected. On the left adnexal region there was a 2 cm ovarian haemorrhagic cyst. The cyst was aspirated and a punch biopsy was performed from the cyst wall. The histopathology was benign.

Case 3

A 25-year-old nulliparous single woman with irregular menses was admitted to our clinic with an adnexal mass. The patient had no other complaint and no history of previous pelvic or abdominal surgery. Her menarche had occurred at the age of 12 and she had regular menstrual periods at 30-day intervals until 7 months prior to her admission. She had used oral contraceptives for treatment of irregular menses for 7 months. A right adnexal cystic mass approximately 6-7 cm diameter was palpated. Transvaginal sonography revealed a 65x35mm hypoechogenic biloculated cystic mass in the right ovarian site. No vascularity was noted on doppler ultrasonography. The left ovary could not be visualized separately. Serum Ca125 level was 15 IU/ ml. She was diagnosed as having a benign right adnexal mass and a laparoscopic approach was planned. On laparoscopic evaluation there was a saggital sulcus on the fundal part of uterus. The left ovary and fallopian tube were absent. On the right ovary there was a cytic mass of 7 cm. Laparoscopic cystectomy was performed and the specimen was sent for histopathologic evaluation. The histopathological report of the specimen was benign cyst adenoma.

Case 4

A 36-year-old nulliparous woman with regular menses applied to our clinic for infertility. She had been trying to conceive for the last 18 months and had no other complaint. Her menarche had occured at the age of 13 and she continued to have regular menstrual periods at 25-day intervals. She had no history of abdominal and pelvic surgery or pelvic inflammatory disease. During pelvic examination a cystic mass was palpated on the right side. Basal infertility investigations of the couple were normal, except for hysterosalpingography (HSG). The left fallopian tube was not visualized and the right tube showed hydrosalpenx, while the uterine cavity had a unicorn shape by HSG. The right ovary was not visualized by ultrasonography whereas there was a 69x29 mm cyctic mass on the right adnexal side. A laparoscopy with concomitant hysteroscopy surgery was planned. On laparoscopic evaluation a right unicorn uterus was detected. The right tube was hydropic and was about 5x6 cm; it was twisted twice. The right ovary was absent. On the left side a rudimentary non-communicant nonfunctional horn was detected. The left ovary and tube were normal. There was no communication between the rudimentary uterine horn and the left tube. The right tube was detorsioned and laparoscopic fimbrioplasty was performed. When the patency of the right tube was evaluated by laparoscopic dye pertubation, methylene blue

failed to pass through the right tube. Laparoscopic salpengectomy was performed and the right tube was sent for histopathologic evaluation. On hysteroscopic evaluation only the right ostium was seen.

Only laparoscopy was performed in the first three cases as all three patients were single and refused to have a hysteroscopy as they were virgins. All patients had complete evaluation of the urinary system via ultrasonography and intravenous pylography after laparoscopy. No urinary tract anomaly was detected.

Discussion

Complete absence of bilateral or unilateral ovarian and tubal structures with a normal uterus is rare, as shown in Table [1-16]. There are three hypotheses for the etiology of these rare cases. The first is the mechanical hypothesis that involves asymptomatic torsion of one or both adnexa or tube/ovary during adult life or childhood, or even in the fetal stage, that results in auto-amputation and necrosis secondary to related ischmia [1,14]. Adnexal torsion can involve either the tube, the ovary, or the adnexa.

The second cause is thought to be congenital. The uterus and fallopian tubes develop from the Mullerian duct whereas ovaries develop mainly from the mesenchymal elements of the urogenital ridge, the coelomic epithelium, and the germ cells arising from the yolk sac. The unilateral congenital absence may originate from a defect in the development of the Mullerian system on one side [14] or a defect of the caudal end of the Mullerian duct and the genital ridge. Paternoster et al.

Table 1. Absence of fallopian tube and/or ovary with uterus, urinary tract, other system anomalies: Literature Review

Authors (reference)	Absence of fallopian tube and/or ovary	Uterine anomaly	Urinary tract anomaly	Other	Indications for the surgery
Sivanesaranam V(1) Case 1 Case 2	Absent left fallopian tube and left ovary Absent right fallopian tube and right ovary	Nm* Nm*	Nm* Nm*	Nm* Nm*	Tubal ligation Infertility
Georgy FM et al. (2)	Absence of left fallopian tube and ovary	Nm*	Nm*	Nm*	Menorrhagia
Lashgari M. (3)	Absence of left ovary	Nm*	Nm*	Nm*	Sterilisation
Sirisena LA. (4)	Absence of left fallopian tube and ovary	No	No	No	Right ovarian cyst
Zaitoon MM. (5)	Absence of left fallopian tube and ovary	No	Renal ectopia	No	Tubal ligation
Sinha MR. (6)	Absence of right fallopian tube and ovary	Nm*	Nm*	Nm*	Torsion of one left ovarian cyst
Silva PD et al .(7)	Absent right fallopian tube and ovary	Nm*	Nm*	Nm*	Cystic mass
Mylonas et al. (8) Case 1 Case 2 Case 3	Absent right tube, right ovary Absent right tube, right ovary Right adnexial aplasia, left ovarian agenesis, left adherent tube	No No No	Nm* Nm* Nm*	No Endometriosis No	Diffuse lower abdominal pain Diffuse lower abdominal pain Diffuse lower abdominal pain
Mulayım et al(9).	Complete absence of left tube and ovary	Unicornuate uterus	Pelvic kidney	No	Infertility
Haydardedeoglu et al.(10)	Absent right ovary	Unicornuate uterus with noncommunicated horn	Ipsilateral renal agenesis	Nm*	Menometrorrhagia, pedunculated submucous leimyoma
Demir et al. (11)	Absent Left tube and left ovary	Unicornuate uterus	No	No	Incidental at cesarean delivery
Muppala et al. (12)	Absent right ovary, tube and round ligament	No	Right renal agenesis	Pyloric stenosis, Endometriosis	Infertility
Suh et al. (13)	Absent left distal tubal segment and left ovary	Uterine septum	Nm*	No	Infertility
Uckuyu et al.(14) Case 1 Case 2 Case 3 Case 4	Absent left distal tubal segment, streak left ovary Absent right distal tubal segment, normal right ovary Twisted left tube, absent right ovary Left ovarian agenesis, normal left tube	No No No No	No No No No	Separeted tissue in the fossa ovarica Separeted tissue in the lateral pelvic wall Separeted tissue in the omentum No	Infertility Infertility Infertility Infertility
Rapisarda et al. (15)	Absent Left tube and left ovary	No	Nm	No	Infertility
Pabuccu et al. (16)	Complete absence of left tube and ovary	No	No	No	Infertility
Presented Cases, Etlik Z.H Case 1 Case 2 Case3 Case 4	Complete absence of the right tube and ovary Complete absence of the right tube and ovary Complete absence of the left tube and ovary Complete absence of the right ovary	No No No Unicorn uterus	No No No No	Endometriosis Endometriosis No No	Ovarian cyst Adnexal mass Adnexal mass Infertility

* Nm:not mentioned

presented 2 cases with absence of fallopian tubes and pointed out the possibility of incomplete tubal development due to inadequate blood supply during the pelvic descent of the caudal part of the Mullerian duct [18]. Absence of the fallopian tube may be related to the development of a unicornuate uterus. In the presence of any described anatomic malformation of the uterus or urinary tract, the hypothesis of the congenital defect is rather strong. Unilateral absence of the right ovary with a unicornuate uterus was observed in the presented case-4, suggesting an incomplete development of the uterovaginal canal. In our series none of the patients had an accompanying urinary tract abnormality.

The third hypothesis is that the tubal and ovarian maldevelopment is secondary to ischemia due to a vascular accident [1, 8, 10, 14]. Duelholm and Praest [20] have pointed out that adnexal torsion might be related to the presence of adnexal masses, hemodynamic abnormalities, sudden body position changes, trauma, surgery, or disease. Torsion of the ovarian pedicle or mesosalpinx can lead to avascular necrosis, separation of those tissues, and resorption [17]. Adnexal torsion can be related to either the tube or the ovary or to both. Although it is thought to be rare before menarche and after menapause, Jamieson and Soboleski [21] and Goktolga et al. [22] reported adnexal torsion in prepuberty. Torsion most likely manifests itself with acute abdominal pain, nausea, and vomiting, but it can also be asymptomatic and therefore diagnosed only in operations performed for other reasons. In the cases reported by Sivanesaratnam [1], Dueck et al. [23] and Guvenc et al. [24], they were also asymptomatic, suggesting that the event had occurred during the fetal stage.

Laparoscopy provides a definitive diagnosis of unilateral ovarian and/or tubal absence. The patients may be operated on for a variety of indications (Table 1). In the presented cases none of the patients had a previous surgery or an acute pelvic pain episode. However, the previous-torsion hypothesis is not easy to prove. The differential diagnosis between congenital agenesis of the tubes and/or ovaries and auto-amputation secondary to silent torsion can only be made by previous radiologic or occult observation of both tubes and ovaries followed by a later failure to detect them. The other sign that implies auto-amputation secondary to torsion is detection of histologically-proven remnants of the ovary and/or the tube in the abdominal cavity [17]. In the second case in our series, remnants of the ovary were observed on the right round ligament and pelvic wall.

The true etiology of unilateral ovarian absence with or without the fallopian tube is unclear in most of the reported cases but this possibility should be considered even in asymptomatic patients with no significant medical history. The absence of any other anatomical structures (such as uterine or renal abnormalities) together with histologically-proven remnant tissues on the ipsilateral side would support the hypothesis of vascular ischemia secondary to torsion rather than a developmental anomaly [18].

Disclosure Statement

We disclose that we don't have any financial relationship with a biotechnology manufacturer, a pharmaceutical company, or other commercial entity that has an interest in the subject matter or materials discussed in the manuscript.

Competing interests

The authors declare that they have no competing interests.

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