Ipsilateral Fallopian Tube and Ovary Agenesis or Absence? Case Report and Review of the Literature

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Submission: November 12, 2017; Published: December 18, 2017

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Abstract

The ipsilateral absence of fallopian tube and ovary is rare. The etiology is not clear and the esteemed incidence is 1:11.240. The unilateral absence is rarely associated to genital and extra-genital malformations. We refer the case of 21 years of women who declared primary amenorrhea and dyspareunia. She was found affected by mammary Tanner stage 3; low 17-β-estradiol and high FSH and LH levels; BMI 32.2; delayed bones development; moderate hyperinsulinemia; normal karyotype. No urinary apparatus anomalies. Her homozygote twin-sister is normal. The diagnostic laparoscopy shows uterine hypoplasia, normal right fallopian tube and ovary and complete absence of left tube and ovary. A review of literature is reported. The patient is now under sequential estro-progestinic treatment and on a diet to improve hypogonadism, sexual life and metabolic system.

Keyword: Adnexa agenesis

Introduction

The unilateral agenesis and/or absence of ipsilateral fallopian tube and ovary (adnexa) is rare and rarely described in the literature. The esteemed incidence is 1:11.240 [1,3]. The etiology is not clear: ipsilateral tubal and ovarian absence could be caused by the torsion of ovarian peduncle before birth or before puberty [2,3]. Adnexa agenesis is always associated to uterine and to urinary apparatus malformations; unilateral agenesis or absence are rarely associated to genital and extra-genital malformations. The majority of affected patients has no special symptoms and signs and the diagnosis follows a laparoscopy or a laparotomy performed for different causes. We report the case of young women affected by primary amenorrhea and review the literature regarding adnexal agenesis and/or absence.

Case Report

S.V. 21 y. o., 93Kg, 170cm high (BMI: 32.2) comes to our Maternity Unit because of primary amenorrhea and dyspareunia. Her homozygote twin sister is regularly menstruated since age 13 and has no gynecological problems.

Modest hyperinsulinemia (13.4UI/ml), low 17-β-estradiol (<10pg/ml) and high FSH (54.2mU/ml) and LH (15mU/ml) levels deposite for an hyper-gonadotropic hypogonadism. At inspection extern genitais appear normal; moderate presence of pubic hair; mammary development at Tanner stage 3 [4]. Striae rubrae are present on both flanks and subcutaneous fat and body hair distribution are of normal feminine habitus. At exploration and inspection the vagina seems normal; cervical hypoplasia with a punctiform external orifice. At transvaginal echography uterus appears hypo-plastic with no endometrial thickness. The right ovary has reduced volume: a little follicle is visible. Left ovary not detectable. RX for bone age (Figure 1) shows transparent rhymes at proximal phalanges and at distal ulnar and radial extremities which prove a delay in bone development for age.

Figure 1: Left hand: delayed bones development.

Karyotype is 46, XX with absence of sequences at locus SRY. Diagnostic laparoscopy shows an hypo-plastic uterus with normal round ligaments; normal right fallopian tube and right ovary seat of little cyst which is aspirated; total absence of left tube and ovary.
tube and ovary; no pelvic adhesions (Figure 2). A successive NMR excludes urinary tract anomalies [5]. After multidisciplinary counselling the patient has been treated with tran-dermic lowering natural estrogens with the attempt to stimulate the secondary sexual characters and to reduce FSH and LH levels and induce menarche. Afterward, natural progesterone per as has been added to sham ovarian cycle.

**Figure 2:** Laparoscopy Hypoplasia of uterus and right fallopian tube and ovary. Absence of left adnexa.

**Discussion**

Unilateral agenesis of adnexa is rarely reported in literature: it seems to be even more rare in absence of uterine anomalies. A review of literature (PubMed) reports 28 cases (Table 1):

> five cases are associated to urinary tract malformations [6-9];
> five cases are associated to uterine anomalies [6,9-12];
> three cases signal contemporary endometriosis [3,7,9];
> twelve cases are associated to primary sterility [1-3,12-16].

In three cases of tube and ovary agenesis is present a contra lateral stenotic tube [1,3,15] and in two cases is present a contralateral ovarian cyst [17,18]. Our patient never had spontaneous menarche: one only similar case is reported in literature [12] typified by hypoplastic uterus, normal karyotype, primary amenorrhea and sterility but this case had bilateral ovarian agenesis and absence of distal tract of both fallopian tubes.

With regard to etio-pathogenesis, is known that the malformation of our clinic case includes organs originating from Muller ducts and from yolk sac mesoderma. During the sixth week of gestation Muller ducts migrate on the median line and their fusion creates the uterus and the proximal tract of the vagina. Rostrally the Muller ducts create the fallopian tubes. The gonads instead derivate from migration of mesodermal primordial germinal cells of the yolk sac to the genital crests giving origin to the sexual cords. In case of male embryo the SRY gene of the short Y arm [19] keeps the sexual cords proliferating deep in the medullar and creates the seminiferous tubules. When the embryo is female, the sexual cords split in series of cellular heaps of granulosa cells surrounding the primordial germinative cells.

**Table 1:** Unilateral Fallopian tube and ovarian agenesis: literature review.

<table>
<thead>
<tr>
<th>Author</th>
<th>Uterine Associated Malformation</th>
<th>Infertility/ Sterility</th>
<th>Urinary Apparatus Associated Malformations</th>
<th>Associated Endometriosis</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pabuccu [13]</td>
<td>Unknown</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Right ovary absence; right fallopian tube anomaly</td>
</tr>
<tr>
<td>Vaiarelli [24]</td>
<td>No</td>
<td>Yes</td>
<td>Unknown</td>
<td>No</td>
<td>Contralateral tubal stenosis</td>
</tr>
<tr>
<td>Eustace [2]</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Left tubal distal tract absence and left ovarian agenesis</td>
</tr>
<tr>
<td>Sivanesaratnam [15]</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Casual check up during surgical detorsion of contra lateral fallopian tube torsion.</td>
</tr>
<tr>
<td>Uckuyu [16]</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Contra lateral tubal stenosis</td>
</tr>
<tr>
<td>Rapisarda [2]</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Diagnosis during tubal sterilization</td>
</tr>
<tr>
<td>Suh [10]</td>
<td>Septed Uterus</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td></td>
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<tr>
<td>Eustace [2]</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>Sivanesaratnam [15]</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Haydardedeoglu [6]</td>
<td>Unicorne Uterus</td>
<td>Unknown</td>
<td>Ipsilateral kidney Agenesis</td>
<td>No</td>
<td></td>
</tr>
</tbody>
</table>
The agenesis of both ipsilateral ovary and fallopian tube with normal uterus is an embryologic condition difficult to explain. Some studies speculate a defect of the genital crest and of the caudal area of the Muller ducts caused by an insufficient vascularization during migration in the pelvis of the caudal section of the para-mesonephrum [2,3,18]. Other studies support 1) an asymptomatic adnexal torsion with secondary ischemia and reabsorption [2,3,18], 2) a congenital absence due to a defect in development of the Muller ducts and of the ipsilateral genital crest [2,7,15,18], 3) a vascular anomaly cause of ischemia in prenatal life [11]. Literature reports the case of a women who ten years before diagnosis of right fallopian tube and ovary absence had an important and persistent pain in the right ovarian region. The authors [19,20] speculate an ovarian torsion with further ischemia and adnexal re-absorption [20-27].

**Conclusion**

In our clinic case, the normal karyotype with absence of SRY gene exclude a condition correlated to a genetic cause. Also the peculiar homo-zygotic twins trails to a noxa happened after the embryologic development of the sexual organs. Moreover, the contra lateral hypogonadism and the uterine hypoplasia suggest the presence of contemporary factors however happened after the embryologic period. For these reasons we suppose a distinctin between agenesis and absence. The treatment with estrogens and estro-progestins as well as an adequate diet has the intent to induce the uterine and the secondary sexual characters development as well as to improve the sexual life and the metabolic system.

**References**


