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CASE REPORT

Unilateral ovarian absence

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Introduction

Unilateral ovarian absence (UOA) is a rare event. It has a prevalence of one in 11,240 women [1]. Even rarer is the absence of both the ovary and distal portion of the ipsilateral fallopian tube. It has been suggested that this phenomenon may be a result of a congenital malformation or an ischemic event due to mechanical alterations (e.g., torsion and subsequent resorption of the ovary) that occur during fetal life or childhood [2]. In this case report, we describe a patient with UOA as an unexpected finding during diagnostic laparoscopy for acute pelvic pain.

Case presentation

A 19-year-old female patient G0P0 presented with acute pelvic pain of increasing intensity. Clinical examination revealed right-sided abdominal tenderness with guarding. Her uterus, vagina, and vulva were normal. Her body temperature was normal, there were no urinary or bowel symptoms and she was on day 22 of her menstrual cycle. Her history was unremarkable, and a β-hCG test was negative. A transvaginal ultrasound scan revealed a normal anteverted uterus, a cystic mass on the right ovary, and free fluid in the lower pelvis with a floating uterus. The left ovary could not be identified during the scan.

After consultation, she consented to and underwent an emergency diagnostic laparoscopy. The laparoscopy confirmed the diagnosis of right ovarian cyst rupture, but also revealed absence of the left ovary, its associated ligaments, and the distal part of the ipsilateral fallopian tube (Fig. 1). The patient was followed up with a whole-abdomen CT scan, which did not reveal urinary tract variations or malformations or ectopic ovarian tissue. Karyotyping was also advised.

Discussion and conclusion

UOA is a very rare condition and can coexist with total or partial absence of the ipsilateral fallopian tube. When the entire fallopian tube is missing, it is probably the result of a congenital malformation (agenesis or aplasia). This is explained embryologically by developmental alterations of the paramesonephric (Müllerian) ducts and unknown paracrine and autocrine signals, which result in agenesis of the associated gonad [2]. This condition potentially coexists with malformations of the uterus and/or urinary tract, such as unicornuate uterus, unilateral renal agenesis, and other variations [3].

The first published case of UOA was reported by Dannreuther in 1923, as mentioned by Alexander [4] in 1947. In his case presentation, UOA was associated with absence of the left broad ligament, round ligament, salpinx, kidney, and ureter and the presence of a unicornuate uterus.

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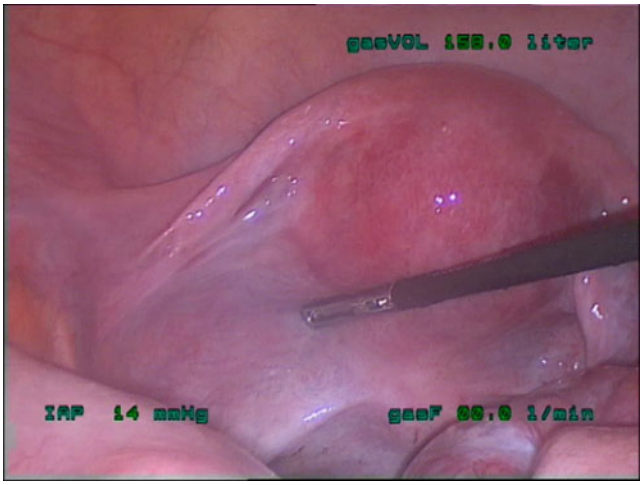

Fig. 1 Illustrating the abrupt discontinuation of the left fallopian tube and the absence of the ipsilateral ovary 361 × 203 mm (72 × 72 DPI)

In 2003, Mylonas et al. [5] reported three such cases and reviewed the literature, which contained a total of 13 cases of UOA and/or fallopian tube absence. In 2007, Demir et al. [6] reported incidental UOA and unicornuate uterus during a cesarean section. In 2008, Suh et al. [7] presented a case of UOA, tubal hypoplasia, and septate uterus and in 2009, Rapisarda et al. [8] reported a case diagnosed during urgent laparoscopy for suspected corpus luteum hemorrhage. In 2009, four new cases of UOA were reported [9]. The most recent case was reported in 2011 [10].

When only the distal part of the ipsilateral fallopian tube is missing along with UOA, the proposed mechanism is ovarian torsion with necrosis and resorption, which can occur either antenatally or postnatally. Adnexal torsion and infarction is usually associated with severe symptoms and is a well-documented surgical emergency in both adults and children. Consequently, some authors [11] support the view that "asymptomatic" torsion cannot occur postnatally. Other authors [2] have suggested that symptoms may be minimal or even absent, especially when torsion occurs during pregnancy.

In our case, there was an abrupt interruption of the fallopian tube and absence of any associated uterine, renal, or other abnormalities. The combination of these findings, along with no indication in the patient's history, suggests that the most likely cause was an asymptomatic ischemic event and resorption during fetal life or after birth.

It has been stated [5] that the incidence of UOA could be higher than that reported in 1986 and the ever-increasing number of cases in the literature supports this view. This could be because of either an unknown environmental factor or the more widespread use of laparoscopy, which has revealed cases that otherwise would have remained undiagnosed. As has been demonstrated, women with UOA and/or fallopian tube absence are fertile [6], so quite possibly some of these women were fertile and might have raised offspring while unaware of their condition.

In 2008, Garret [12] proposed an interesting theory based on his observations of discontinuous fallopian tubes in non-sexually active adolescent girls. This theory stated that the fimbria could have an embryologic origin distinct from that of the rest of the fallopian tube and potentially in common with the ovary. This suggestion is in line with studies involving the fimbria as the likely site of origin of ovarian epithelial neoplasms [13]. The variety of congenital and acquired malformations could offer new insight into the embryologic and developmental processes involved and may prove beneficial for the management of ovarian epithelial neoplasms.

Diagnostic laparoscopy plays a vital role in the diagnosis of these abnormalities. We recommend the principle of "three-cycle inspection" as a standard step in all laparoscopic procedures. The first cycle is a clockwise inspection of the upper abdomen before placing the patient in the Trendelenburg position. The second cycle is carried out after insertion of the accessory trocars and preparation of the surgical field and refers mainly to the anatomical structures that enter the pelvis at the level of the pelvic brim. The third cycle involves visualization of the internal genitalia and lower pelvis with the assistance of a uterine manipulator when required and is performed after placing the patient in the maximum Trendelenburg position.

It is also important to note that in the clinical setting of evaluating common, acute gynecological emergencies, the suspicion of such a rare abnormality may be raised by preoperative ultrasound imaging findings, drawing the surgeon's attention to actively search for the "missing" ovary. If the ovary cannot be clearly visualized during the ultrasound study, the possibility of UOA should be considered as a differential diagnosis. We believe that a thorough inspection of the whole abdomen is mandatory whenever laparoscopy is performed despite the adverse conditions that can be encountered during an emergency procedure.

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