

# True ectopic ovary in the right iliac fossa mimicking acute appendicitis and associated with ipsilateral renal agenesis

Paraskevi Kollia<sup>1</sup>, Christos Kounoudes<sup>2</sup>, George Veloudis<sup>1</sup>, Nikoletta Giannakou<sup>2</sup> and Stavros Gourgiotis<sup>1</sup>

<sup>1</sup>First Surgical Department, 417 NIMTS Military Veterans' Fund Hospital of Athens, and <sup>2</sup>Athens Medical Center, Athens, Greece

## Abstract

Ectopic ovarian tissue is a rare gynecologic condition. The presence of ectopic ovary may be accompanied by maldevelopments of the genital and urinary tract. We report an extremely rare case of a 39-year-old woman presenting with abdominal pain localized in the right lower quadrant. During the preoperative investigation and the exploratory laparotomy, an ectopic ovary in contact with the appendix accompanied by a single left kidney was found. The present report also includes a review of the related published work. To the best of our knowledge, this is one of the very few cases reported describing the co-occurrence of true ovarian ectopia and ipsilateral renal agenesis.

**Key words:** appendectomy, ectopic tissue, ipsilateral, ovary, renal agenesis.

## Introduction

Ectopic ovarian tissue is a rare embryological abnormality with less than 50 cases reported since 1959.<sup>1</sup> Ectopic ovaries have an estimated prevalence between 1 in 29 000 and 1 in 93 000 gynecologic admissions.<sup>2</sup> A more accurate diagnosis is difficult due to a confusing classification system, as well as their frequently asymptomatic nature. They can be distinguished in supernumerary and accessory ovaries although this terminology is sometimes confusing.<sup>3</sup> Furthermore, ectopia of the ovary may be rarely accompanied by maldevelopments of the genital tract and changes in the renal system.<sup>4</sup>

Appendicitis is the most common general surgical emergency in developed countries. In the majority of female patients, one or more gynecological disorders can mimic acute appendicitis, including pelvic inflammatory disease, ovarian torsion, tubal ectopic pregnancy, hemorrhagic cysts, recent ovulation, tubo-ovarian masses or infected cysts.<sup>5</sup>

We herein describe the clinical findings, diagnostic procedures and management of a 39-year-old female patient with symptoms of acute appendicitis, an ectopic ovary in contact with the appendix, and right (ipsilateral) renal agenesis. It is an extremely rare case, of the very few reported in the published work; to the best of our knowledge, it is the second one.

## Case Report

A 39-year-old female patient, gravida 0, para 0, presented with a complaint of abdominal pain localized in the right lower quadrant and a body temperature of 37.2°C. She stated that the localized pain had been intermittent during the last 2 months and had increased in intensity (8/10) 12 h prior her admission. The pain was not associated with nausea, vomiting or symptoms from her urinary tract. Her past medical and family history was unremarkable. She had a regular menstrual cycle of 26–30 days with average painless flow for 4–5 days. She had never undergone pelvic or

Received: February 27 2013.

Accepted: July 30 2013.

Reprint request to: Dr Stavros Gourgiotis, 161 Mesogeion Avenue, Athens 11526, Greece. Email: drsgourgiotis@tiscali.co.uk

Conflict of interest: There is no conflict of interest.

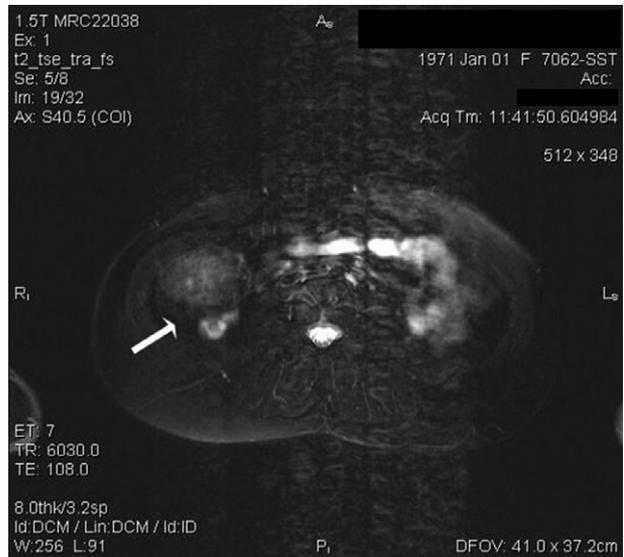
abdominal surgery. However, she reported similar pain attacks in the past at the middle of her cycle. The patient did not report infertility issues. She had been on oral contraceptives for 2 years and had stopped them 4 years prior. The pain attacks were during the middle of her cycle so we assumed them to be 'Mittelschmerz' syndrome. She did not report symptoms of dysmenorrhea or painful intercourse.

On admission, the pain was focused in the right iliac fossa with no radiation. McBurney's sign was positive while the digital rectal examination showed tenderness of the dorsal wall and was negative for blood. There was no abdominal distension, bowel sounds were normal, whereas liver and spleen were not palpable. Pregnancy and urine test were both negative. Her blood cell counts showed a marginal elevation of leukocytes 11 250/ $\mu$ L (4000–11 000). The clinical examination of the external genitalia and pelvis during admission was normal. There was no blood or bulging mass, no tenderness while the vagina and cervix were normal. The uterus was anteverted and was of normal shape and size.

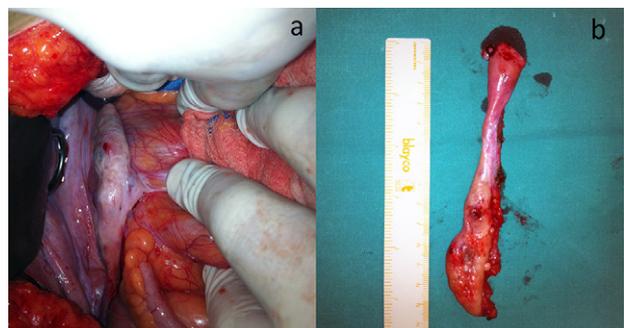
The abdominal ultrasound (US) showed a mass in the right iliac fossa rather retrocecal while the appendix could not be identified. The US of the pelvis showed a small amount (~20 mL) of fluid in Douglas pouch, a normal and full bladder, a normal uterus, a normal left ovary, while the right ovary could not be identified. A vaginal US was not performed.

The magnetic resonance imaging (MRI) identified a mass, approximately 12.5 cm  $\times$  3 cm  $\times$  2.5 cm in size, with regular margins and microcystic morphology, located retrocecal between the cecal pol and the psoas muscle (Fig. 1). The mentioned mass resembled a slightly hypoplastic, undescended ovary. As an incidental finding, the agenesis of the right kidney was revealed. Both the left kidney and the left ovary were of normal morphology and location. The MRI also revealed a normal uterus and left ovary and the ectopia of the right ovary inside the right iliac fossa.

The exploratory laparotomy revealed a retrocecal appendix with no macroscopic evidence of inflammation. Furthermore, the cystic mass was in contact with the antimesenteric border of the appendix (Fig. 2). Appendectomy and resection of the mass were performed. At the time of laparotomy, the uterus was of normal size and shape, both fallopian tubes were attached to it, and there was no sign of inflammation or mass in the pelvis. No abnormalities of the uterus and the fallopian tubes were observed. The right fallopian tube was attached to the uterus, but had no continuity with the cystic mass.



**Figure 1** Magnetic resonance imaging confirms the existence of a mass in the right iliac fossa (arrow).

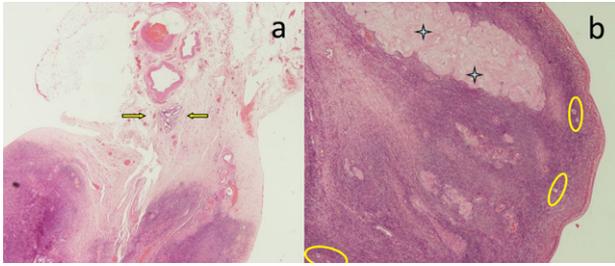


**Figure 2** (a) Intraoperative finding of the ectopic ovary in contact with the antimesenteric border of the appendix. (b) Resected ectopic ovarian tissue.

Histopathological examination of the appendix revealed no dilated lumen and minor inflammation of the mucosa with a small portion of purulent exudates relevant to the operative findings. Histopathological examination of the resected mass revealed ectopic ovarian tissue (Fig. 3). The patient was discharged the fourth postoperative day and has had no recurrence of symptoms 6 months later.

## Discussion

Ectopic or undescended ovaries are characterized by their attachment to an area above the level of the



**Figure 3** (a) Ectopic ovary, composed of normal-appearing cortex and medulla (bottom) and hilum, containing blood vessels and hilar mesonephric rests (arrows) (hematoxylin–eosin [HE], original magnification  $\times 20$ ). (b) Ovarian cortex, containing few primordial follicles (circles) and one corpus albicans (stars) (HE,  $\times 40$ ).

common iliac vessels.<sup>3</sup> There are two broadly acknowledged classifications. In 1959, Wharton classified ectopic ovaries as either accessory or supernumerary; being distinguished by their relationship to a normal ovary.<sup>6</sup> Accessory ovary was defined as excess ovarian tissue adjacent and connected to a normal ovary. On the other hand, supernumerary ovaries were described as those ovaries situated away from normal ovaries (e.g. in the retroperitoneum or omentum). This classification implies the presence of two normal ovaries and also does not account for abnormally located ovarian tissue secondary to previous pelvic surgery, such as found in ovarian implant/remnant syndrome.

On the other hand, the review by Lachman and Berman found that almost 50% of the reported cases since 1959 were in patients with previous pelvic surgery.<sup>3</sup> They also proposed a new classification of abnormally located ovarian tissue to further clarify the issue. They suggested that ectopic ovary should be the proper term and that this can be further subclassified as: (i) post-surgical implant; (ii) post-inflammatory implant; and (iii) true (ectopic) ovarian tissue.

Both the genital and the urinary system develop from the intermediate mesoderm, which extends along the dorsal wall of the embryo. This is the primordium of the urogenital ridge which can be divided into two parts: the nephrogenic ridge which gives rise to the urinary system and the gonadal ridge which becomes the genital system. The primitive genital system in the two sexes is similar. The gonads are derived from three sources: mesothelium, mesenchyme and primordial germ cells. By 18 weeks, many primordial ovarian follicles have already formed. The ovaries, located on the dorsal wall of the embryo, begin their descent by 20

weeks. Guided by the gubernaculum and processus vaginalis, the ovaries descend to a point inferior to the pelvic brim. The gubernaculum gives rise to the ovarian ligaments and round ligaments of the uterus.<sup>2</sup>

Ectopia of the ovary may be accompanied by additional maldevelopments of the genital tract which sometimes are accompanied by anomalous changes in the renal system. Renal agenesis is a relatively common congenital anomaly, although its etiology is unknown. This kind of agenesis may be unilateral or bilateral and is generally thought to result from: (i) absence of the metanephric blastema; (ii) ureteral bud maldevelopment; or (iii) lack of induction of the metanephric blastema by the ureteral bud. Unilateral renal agenesis is usually an incidental finding. Renal agenesis is also associated with ipsilateral urogenital anomalies.<sup>7</sup>

The ovary, which forms from the urogenital ridge, is present on the affected side but may be higher in the abdomen than its normal pelvic location.<sup>8</sup> The ovaries in such patients may be found in the upper abdomen, at the level of the pelvic brim, or in the inguinal canal. In our case, the right ovary was located in the right fossa and the right kidney was absent.

It is necessary to mention that renal agenesis and true ectopia of the ovary are both anomalies of the urogenital ridge, the common mesodermal ridge that forms the developing gonad and the mesonephros. Renal agenesis occurs when the nephrons fail to develop and the ureteric bud fails to branch. It may also arise if the interaction between the metanephric mesoderm and the ureteric bud fails to occur. The descent of the ovary into the pelvis depends on the formation of the cranial genital ligament, which forms the suspensory ligament of the ovary, and the caudal genital ligament, which forms the proper ligament of the ovary and the round ligament of the uterus. The uterus is formed by the fusion of both paramesonephric (Müllerian) ducts and abnormalities are due to partial or complete atresia of one of the ducts, which occurs after the formation of the ovary.

According to the facts mentioned above, we assume that in our case report the patient suffered a defect of the urogenital ridge which led to two correlated but independent abnormalities in two different steps of the development of the urogenital system. The uterus was normal because the paramesonephric ducts fused and the ovary failed to descend due to a malformation of its ligaments. We also characterize the ectopic ovarian tissue of our case as a true ectopic ovary, undescended due to a combined malformation of the urogenital tract which also appeared with the unilateral renal agenesis.

Furthermore, it is known that ectopic ovaries may cause menstrual irregularities, infertility or abdominal pain. In our case, we assumed that the ectopic ovarian tissue was the cause of the abdominal pain. The pain was associated with the patient's menses and there were no characterized symptoms of acute appendicitis like anorexia, vomiting or fever apart from the positive McBurney's sign. Finally, the level of the inflammation of the appendix, according to the histopathology report, was not high enough to justify the symptoms of the right lower quadrant as clear symptoms of acute appendicitis.

Magnetic resonance imaging and surgery is an option for the diagnosis and management of an ectopic ovary.<sup>9</sup> MRI is a non-invasive, highly sensitive and specific radiologic modality in the diagnosis of genital tract and concurrent renal anomalies and follow-up in patients with infertility or recurrent pelvic pain. Surgery remains the gold standard to reveal the cause and to manage ovarian problems. In the presented case, we preoperatively confirmed the absence of the right kidney and the presence of the mass in the right iliac fossa. The renal function was in normal range and we thought there was no need for further renal agenesis evaluation by i.v. pyelogram.

In conclusion, we present an extremely rare case of ipsilateral ovarian ectopia and renal agenesis concomi-

tantly. The true ectopic ovary was also mimicking some of the symptoms of acute appendicitis. We finally provide hints with which to guide the management of similar cases.

## References

1. Bayramov V, Sukur YE, Cetinkaya E, Berker B. Ectopic ovary autotransplanted over rectosigmoid colon: A case report. *Fertil Steril* 2009; **92**: 1496.e15-16.
2. Watkins BP, Kothari SN. True ectopic ovary: A case and review. *Arch Gynecol Obstet* 2004; **269**: 145-146.
3. Lachman MF, Berman MM. The ectopic ovary: A case report and review of the literature. *Arch Pathol Lab Med* 1991; **115**: 233-235.
4. Uyar I, Gulhan I, Sipahi M, Hanhan HM, Ozeren M. Ectopic ovary confirmed by ovarian stimulation in a case of unicornuate uterus. *Fertil Steril* 2011; **96**: e122-e124.
5. Shin LK, Jeffrey RB. Sonography and computed tomography of the mimics of appendicitis. *Ultrasound Q* 2010; **26**: 201-210.
6. Wharton L. Two cases of supernumerary ovary and one of accessory ovary, with an analysis of previously reported cases. *Am J Obstet Gynecol* 1959; **78**: 1101-1119.
7. Mishra A. Renal agenesis: Report of an interesting case. *Br J Radiol* 2007; **80**: e167-e169.
8. Reichman DE, Laufer MR. Congenital uterine anomalies affecting reproduction. *Best Pract Res Clin Obstet Gynaecol* 2010; **24**: 193-208.
9. Litos MG, Furara S, Chin K. Supernumerary ovary: A case report and literature review. *J Obstet Gynaecol* 2003; **23**: 325-327.