

A rare case of intrinsic ureteral endometriosis causing hydronephrosis in a 40-year-old woman.

A case report and literature review

**E. Papakonstantinou¹, F. Orfanos¹, Th. Mariolis-Sapsakos¹, D. Vlahodimitropoulos²,
A. Kondi-Pafiti³**

¹Department of Surgery, Evgenideion Hospital University of Athens; ²Pathology, Evgenideion Hospital University of Athens and

³Department of Pathology, Areteion Hospital, Athens (Greece)

Summary

Endometriosis is a multifactorial disease with unclear pathogenesis. Urinary tract endometriosis occurs in about 1% of all endometriotic lesions while isolated ureteral endometriosis is extremely rare. We present a case of intrinsic ureteral endometriosis causing ureteral stenosis in a 40-year's old woman, in combination with intestinal, extensive peritoneal and ovarian endometriosis.

The clinicopathological features and investigation methods used, as well as the treatment approach are discussed. An individual therapy plan depending mainly on the patient's age, desire for children and the extent of the endometriotic foci should always be attempted. Collaboration between gynecologists and urologists was essential in our cases.

Key words: Ureter; Endometriosis; Ureteral endometriosis; Intestinal endometriosis; Ovaries.

Introduction

Endometriosis is a multifactorial disease with unclear pathogenesis affecting around 5-15% of women of reproductive age. It is characterized by the presence of uterine endometrial tissue outside its normal location, mainly in the ovaries, the pelvic peritoneum, and the recto-vaginal septum and rarely in the pericardium, pleura and even brain [1-3].

Although endometriosis was first described in 1960, it is a very controversial and enigmatic disease, and many theories regarding its pathogenesis have been advanced. Most studies of pelvic endometriosis support the implantation theory of Sampson, which proposes that during menstruation, endometrial tissue flows back via the Fallopian tubes into the abdominal cavity where it can implant [4, 5]. The theory of Mullerian metaplasia of coelomic epithelium was initially introduced by Mayer [6], which was extended later to the induction theory. The development of endometriosis to distant sites may be explained by dissemination through lymphatic and venous vessels and implantation [7-11].

Current investigation focuses on the factors favoring the growth and development of endometrial tissue in the peritoneal cavity in some women and its regression in others. Genetic predisposition, the presence of a familial tendency and the influence of environmental factors suggest a polygenic/multifactorial manner of development [9-12].

There are two histological types of the rare form of ureteral endometriosis: intrinsic and extrinsic. Intrinsic is defined when the lesions infiltrate the muscularis of the

ureteral wall, while it is considered as extrinsic when the infiltrating lesions are responsible for significant ureteral obstruction but without involvement of the ureteral muscularis. Severe ureteral endometriosis causes significant obstruction to urinary flow with ureteral stenosis resulting in hydronephrosis [13-16].

In this case report, the clinicopathological findings of a rare case of intrinsic ureteral endometriosis in combination with extensive peritoneal, intestinal and ovarian endometriosis are presented, and the therapeutic approach is discussed.

Case Report

A 40-year-old woman visited the Gynecological Clinic of Evgenideion Hospital complaining of severe lower abdominal pain that usually presented during the third and fourth day of her menstrual cycle.

Clinical examination revealed mild abdominal tenderness, especially in the hypogastrium. No masses were palpated. Rectal examination showed a painful mass at the Douglas space. Menstrual irregularities were not referred. The patient had two children by cesarean section and the history of a surgical removal of a uterine mass that after histological examination was reported to be leiomyoma.

Urination was normal with greater frequency during menstrual periods. Constipation is also reported with occasional episodes of bloody and mucus diarrhea. Mild blood hypertension was noticed.

The patient reported that the symptoms appeared during the previous five months, thus she underwent CT scan and MRI of the lower abdomen which revealed: a cyst of the left ovary, 13 mm in diameter with hemorrhagic content; a similar cyst 18 mm in diameter on the left side of the Douglas space; a non-homogeneous intense nodule close to the left ovary and at the rectosigmoid junction; and a thickening of the left side wall of the uterus (consistent with the history of the removed leiomyoma).

Revised manuscript accepted for publication May 23, 2011

Fig. 1

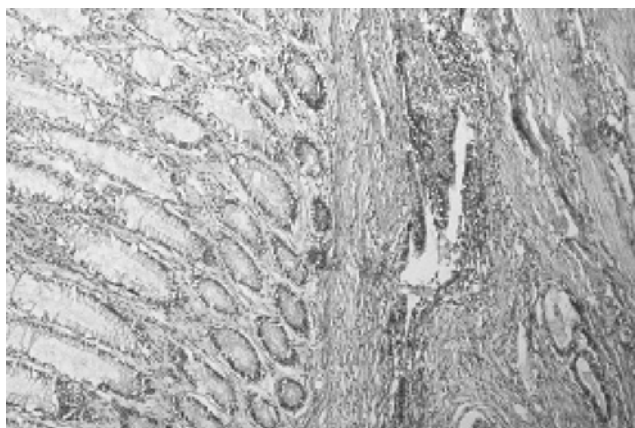


Figure 1. — Histological section of colonic wall showing a focus of endometriosis in the muscular wall (H-E x 120).

Figure 2. — Histological section of ureteral muscular wall showing an ectopic endometrioid gland (H-E x 200).

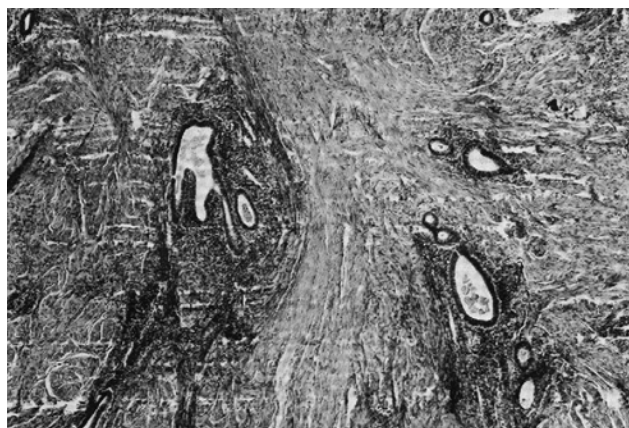


Fig. 2

A colonoscopy was performed and abnormal mucosa of the rectosigmoid colon with intense erythematic lesions was revealed. Biopsy specimens obtained from the lesion showed benign glandular formations inside the thickening wall of mucosa similar to endometriotic glands, suggesting endometriosis of the rectum (Figure 1). The rest of the colon was normal.

When the patient was admitted to our clinic, a transabdominal US was performed to obtain a more detailed description of the anatomical structure and it revealed a left unilateral stricture of the ureteral pelvic tract with secondary, severe dilatation of the upstream tract of the left ureter and hydronephrosis of the left kidney, with no conclusive radiological evidence about the nature of the stenosis. After ureteral involvement was revealed, renal function was checked by kidney scintigraphy which revealed partial loss of left renal function.

During open laparotomy, extensive endometriosis of the peritoneum was observed, as well as cystic structures of the left adnexa, infiltration of the left adnexa and the left ureter from endometriotic lesions, as well as endometriotic infiltrates of the right side of the Douglas pouch, firmly attached to pelvic peritoneum and the rectal wall. A left salpingo-oophorectomy was performed along with excision of a segment of the left ureter en bloc with the periureteral mass that caused dilatation of the left ureter and insertion of a pigtail stent (7 fr). Finally, diathermo-coagulation ablation was performed for all the endometriotic foci of the peritoneum. Histological examination revealed a tumor-like form of ureteral endometriosis engulfing the left ureter, while the left adnexa including the ovary contained abundant endometrial glands and supporting stroma (Figure 2).

Immediately, after the surgery, all the patient's abdominal symptoms abated and hydronephrosis was alleviated. Follow-up investigation revealed complete recovery and no relapses of the ureteral disease were highlighted. Medical hormone suppression was used as adjuvant therapy to surgery and as a preventive therapy for relapses.

Discussion

Endometriosis can be classified according to the revised American Fertility Society classification (rAFS) into minimal, mild, moderate or severe endometriosis, based on a number of points given for the presence of ovarian or peritoneal endometriosis (subdivided into

superficial or deep), the presence of adhesions and posterior cul de sac obliteration [2, 3].

The prevalence of extra pelvic endometriosis is unknown because of a lack of epidemiologically well designed studies. The variety of symptoms, signs and locations and the difficulty in establishing the diagnosis of the disease are the main difficulties in estimating the prevalence of the disease [1-3].

Endometriosis, which is biologically much similar to benign neoplasia, is a pathological entity marked by high local invasiveness and high recurrence. It mainly affects women of fertile age, and low or lack of parity, hormone therapies, previous gynecological surgery and cesarean section are regarded as risk factors [1-4].

Urinary tract endometriosis is a rare event, observed in about 1% of all endometriotic lesions, and about 30% of patients suffer from reduced kidney function at the time of diagnosis [13-16]. The most frequently affected side is the left one and, according to Vercellini *et al.* this fact may be ascribed to the sigma creating locally favorable conditions for cell seeding in a retrograde manner from the uterine cavity [14].

The response of the ectopic endometrial tissue to hormone stimulation results in cyclical bleeding of the lesion and its subsequent desquamation, necrosis and fibrosis, all contributing significantly to the development of ureteral stenosis. Severe ureteral endometriosis causes significant obstruction to urinary flow with ureteral stenosis resulting in hydronephrosis during radiologic examination [15-18].

Isolated ureteral endometriosis is rare. It is usually considered as a multifocal pathology [16]. Patients usually present with associated histologically proven endometriotic lesions mainly in the intestine [19-23] and our case seems to confirm this finding.

It is essential to consider this tendency for multifocal development of endometriosis when considering the surgical modalities for these patients. An interdisciplinary approach is necessary to detect and excise all the endometriotic lesions in a single operation. The optimum

method of surgical management requires the diagnosis be made preoperatively and not fortuitously during surgery.

The difficulty is that the diagnosis of ureteral endometriosis is challenging because the clinical presentation is nonspecific [17]. Indeed, clinical symptoms are often silent and abdominal pain is probably the cardinal symptom of endometriosis. Infertility is another commonly associated complaint. The diagnosis of endometriosis still presents several problems resulting from similarities in clinical symptoms to other benign or malignant diseases. Diagnosing is usually aided by the symptoms chronology linked to the menstrual cycle but, most of all by the patient's positive medical history for endometriosis [18].

The gastrointestinal tract is the most common site of extra pelvic endometriosis, affecting 5-15% of women with pelvic endometriosis. Among women with intestinal endometriosis, the rectum and sigmoid colon are the most commonly involved areas (75-90%). Other parts of the bowel commonly affected are the distal ileum (2-16%) and appendix (3-18%). Superficial intestinal endometriosis is often characterized by visible serosal implants [19, 20]. It seems that intestinal endometriosis is more specific in the symptoms presented, including abdominal pain, abdominal mass, obstruction, rectal bleeding, infertility, diarrhea and increasing urinary frequency [21]. The classic triad of dysmenorrhea, dyspareunia and infertility, as a result of concomitant pelvic disease, may also exist [22, 23].

Various methods of investigation (intravenous pyelogram, retrograde pyelography, US, kidney scintigraphy) have been proposed in cases of clinical suspicion of ureteral endometriosis [24]. MRI techniques have been increasingly improved during the past ten years and numerous recent studies have underlined the advantages of MRI for the diagnosis of endometriotic lesions [25]. Transabdominal US of the pelvis is the initial method of choice to identify and characterize adnexal structures. Identification of specific locations of deep endometriosis is usually achieved by MRI, which remains the key examination for ureteral and bowel endometriosis [26].

Endometriosis is particularly difficult to treat and the treatment of choice is surgical, medical or a combination of both approaches [23, 27].

Factors to consider in management include the age and reproductive desires of the patients, stage of the disease and most importantly the symptoms. Oral contraceptives, androgenic agents, progestins and gonadotrophin releasing hormone analogs have all been used successfully.

Ureteral obstruction develops slowly from periureteral fibrosis and often results in asymptomatic hydronephrosis, loss of renal function and hypertension. Conservative procedures such as ureteral stenting, associated with medical treatment usually lead to favorable outcomes. Surgical management depends on the clinical form of the ureteral endometriosis (minimal or severe). For patients presenting minimal ureteral endometriosis, conservative treatment with excision of endometriotic lesions associ-

ated with ureterolysis is an effective approach. However, not all cases of ureteral endometriosis can be treated with simple ureterolysis.

Ureterolysis with possible omentopexy should be considered in case of localized pelvic extrinsic endometriosis with limited ureteral involvement but this method is not adequate in cases of extensive involvement of the ureter or intrinsic endometriosis, which is hard to determine without histological examination.

In segmentary ureterectomy the active disease, the diseased part of the ureter and all surrounding fibrosis are removed. Ureteral termino-terminal anastomosis may be performed in less severe cases when the distal ureteral tract shows no signs of endometriosis. However, in many cases the spread of disease in the pelvic area prevents the surgeon from using the distal pelvic ureter and therefore uretero-cystoneostomy is performed [18].

When the endometriosis penetrates through the entire depth of the organ wall, complete resection and re-anastomosis of the ureter or bowel can be safely performed, either laparoscopically or with open surgery [27].

Whereas the intrinsic form of ureteral endometriosis often needs aggressive surgery [28], the extrinsic form may be treated with conservative techniques.

Conclusions

Multifocal development of endometriosis with involvement of the ureter causing severe obstruction, the bowel, the ovaries and the peritoneum is an uncommon condition and the counseling physician's awareness is important. A detailed study of the urinary tract based on US findings and urographic examinations in combination with MRI for the extent of endometriosis, is absolutely essential for all patients suffering from pelvic endometriosis.

An individual therapy plan depending on the patient's age, desire for children and the extent of the endometriosis should always be attempted. Collaboration between gynecologists and urologists is essential for the correct therapeutic procedure of these uncommon cases.

References

- [1] Snesky T.E., Liu D.T.: "Endometriosis: associations with menorrhagia, infertility, and oral contraceptives". *Int. J. Gynaecol. Obstet.*, 1980, 17, 573.
- [2] Cramer D.W.: "Epidemiology of endometriosis in adolescents". In: Wilson E.A. (ed.). *Endometriosis*, New York, Alan Liss, 1987, 5.
- [3] Giudice L., Kao L.: "Endometriosis". *Lancet*, 2004, 364, 1789.
- [4] Vinatier D., Orazi G., Cosson M., Dufour P.: "Theories of endometriosis". *Eur. J. Obstet. Gynecol. Reprod. Biol.*, 2001, 96, 21.
- [5] Halme J.A., Hammond M.G., Hulka J.F., Raj S.G., Talbert L.M.: "Retrograde menstruation in healthy women and in patients with endometriosis". *Obstet. Gynecol.*, 1984, 64, 333.
- [6] Fuji S.: "Secondary Müllerian system and endometriosis". *Am. J. Obstet. Gynecol.*, 1991, 165, 219.
- [7] Seli E., Berkkanoglu M., Aydin A.: "Pathogenesis of endometriosis". *Obstet. Gynecol. Clin. N. Am.*, 2003, 30, 41.
- [8] Heinig J., Von Otte S., Greb R., Kiesel L.: "Molecular mechanisms contributing to the pathogenesis of endometriosis". *Gynecol. Endocrinol.*, 2002, 16, 493.

- [9] Lebovic D.I., Mueller M.D., Taylor R.N.: "Immunobiology of endometriosis". *Fertil. Steril.*, 2001, 75, 1.
- [10] Christodoulakos G., Augoulea A., Lambrinoudaki I., Sioulas V., Creatsas G.: "Pathogenesis of endometriosis: The role of defective immunosurveillance". *Eur. J. Contracep. & Reprod. Health Care*, 2007, 12, 194.
- [11] dos Reis R.M., de Sá M.F., de Moura M.D., Noqueira A.A., Ribeiro J.U., Ramus E.S., Ferriani R.A.: "Familial risk among patients with endometriosis". *J. Assist. Reprod. Genet.*, 1999, 16, 500.
- [12] D'Hooghe T., Debrock S., Meuleman C., Hill J., Mwenda J.: "Future directions in endometriosis research". *Obstet. Gynecol. Clin. North Am.*, 2003, 30, 221.
- [13] Chapron C., Chopin N., Borghese B., Foulot H., Dousset B., Vacher-Lavenu M.C. *et al.*: "Deeply infiltrating endometriosis: pathogenetic implications of the anatomical distribution". *Hum. Reprod.*, 2006, 21, 1839.
- [14] Vercellini P., Pisacreta A., Pesole A.: "Is ureteral endometriosis an asymmetric disease?". *Br. J. Obstet. Gynecol.*, 2000, 107, 559.
- [15] Chapron C., Chiodo I., Leconte M., Amsellem-Ouazana D., Chopin N., Borghese B., Dousset B.: "Severe ureteral endometriosis: the intrinsic type is not so rare after complete surgical exeresis of deep endometriotic lesions". *Fertil. Steril.*, 2010, 93, 2115.
- [16] Marcelli F., Collinet P., Vinatier D., Robert Y., Triboulet J.P., Biserte J. *et al.*: "Ureteric and bladder involvement of deep pelvic endometriosis. Value of multidisciplinary surgical management". *Prog. Urol.*, 2006, 16, 588.
- [17] Ghezzi F., Cromi A., Bergamini V., Bolis P.: "Management of ureteral endometriosis: areas of controversy". *Curr. Opin. Obstet. Gynecol.*, 2007, 19, 319.
- [18] Antonelli A., Simeone C., Frego E., Minini G., Bianchi U., Cosciani-Cunico S.: "Surgical treatment of ureteral obstruction from endometriosis: our experience with thirteen cases". *Int. Urogynecol. J.*, 2004, 15, 407.
- [19] Fu C.W., Zhu L., Lang J.H.: "Terminal ileum perforation: a rare complication of intestinal endometriosis". *Ch. Med. J.*, 2007, 120, 1381.
- [20] Jubanyik K., Comite F.: "Extrapelvic endometriosis". *Obstet. Gynecol. Clin. North Am.*, 1997, 24, 411.
- [21] Yantiss R.K., Clement P.B., Young R.H.: "Endometriosis of the intestinal tract: a study of 44 cases of a disease that may cause diverse challenges in clinical and pathologic evaluation". *Am. J. Surg. Pathol.*, 2001, 25, 445.
- [22] Dimoulis P., Koutroubakis I., Tzardi M., Antoniou P., Matalliotakis I., Kouroumalis E.: "A case of sigmoid endometriosis difficult to differentiate from colon cancer". *BMC Gastroentero*, 2003, 3, 18.
- [23] Keckstein J., Wiesinger H.: "Deep endometriosis, including intestinal involvement-the interdisciplinary approach". *Min. Inv. Ther. & All Techn.*, 2005, 14, 160.
- [24] Yohannes P.: "Ureteral endometriosis". *J. Urol.*, 2003, 170, 20.
- [25] Balleyguier C., Roupret M., Nguyen T., Kinkel K., Helenon O., Chapron C.: "Ureteral endometriosis: the role of magnetic resonance imaging". *J. Am. Ass. Gynecol. Laparosc.*, 2004, 11, 530.
- [26] Kinkel K., Frei K., Balleyguier C., Chapron C.: "Diagnosis of endometriosis with imaging: a review". *Eur. Radiol.*, 2006, 16, 285.
- [27] Nezhat C., Nezhat F., Nezhat C.H., Seidman D.S.: "Severe endometriosis and operative laparoscopy". *Curr. Opin. Obstet. Gynecol.*, 1995, 7, 299.
- [28] Horn L.C., Do Minh M., Stolzenburg J.U.: "Intrinsic form of ureteral endometriosis causing ureteral obstruction and partial loss of kidney function". *Urol. Int.*, 2004, 73, 181.

Address reprint requests to:
 A. KONDI-PAFITI, M.D.
 Pathology Department
 Areteion Hospital
 76 Vas. Sofias Ave
 Athens, 11528 (Greece)
 e-mail: akondi@med.uoa.gr