Thai Journal of Obstetrics and Gynaecology
March 1999, Vol. 11, pp. 49-52

CASE REPORT

Laparoscopic Gonadectomy in Gonadal Dysgenesis : a case report

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Abstract

46 XY pure gonadal dysgenesis is a syndrome characterized by disorder of sexual differentiation. We report a case of 46xy gonadal dysgenesis phenotypical girl, whose gonads were successfully detected and removed by laparoscopy. It is clearly shown that laparoscopic gonadectomy is an alternative to laparotomy to minimize patient's complications.

Key words : gonadal dysgenesis, laparoscopy

Introduction

46xy gonadal dysgenesis, first described by Swyer and Phil in 1955[1] and found to be 1:65000 male,[2] is a syndrome characterized by disorder of sexual differentiation and reversal of external sexual appearance. The patients will be phenotypic females with a 46XY karyotype and hypoplastic gonads. The syndrome is governed by the abnormality in the sex determining region on the y chromosome (SRY)[5] Most common presentations are primary amenorrhea with varies degrees of secondary sex characteristic deficiencies. The Muerellian system, e.g. uterus, upper one-third of vagina and fallopian tubes will show varies degree of atrophic appearance while the gonads become streak and associates with increasing risk of germ cell tumor, dictating early prophylactic gonadectomy. Traditionally the procedure has been performed by laparotomy.[3,4] Open laparotomy is usual the method of choice but laparoscopy becomes the competitive alternative. The advantages of laparoscopy over laparotomy are shorter hospital stay, shorter recover period, excellent cosmetic result, minimal pain and reduced need for analgesics.

A case report

A 15-year-old phenotypic girl was referred to Bangkok Christian hospital for ongoing investigation and treatment. She presented with primary amenorrhea. Her mother received no abnormal medication during pregnancy. The course of maternal pregnancy, delivery, her neonatal period and childhood were also uneventful. There was no family history of primary amenorrhea and delayed sexual development. Physical examination revealed short, slight obese female patient with 33-Kg body weight 128 cm. arm span and 132.3 cm. height Normal body hair distribution and no web neck. The breasts and pubic hair were Tanner stage I. The external genitalia were infantile (hypoplastic labia and clitoris). The 2- cm.-long vagina exhibited estrogen deficiency by a lack of

significant rugation. Pelvic ultrasound examination failed to demonstrate uterus, ovaries and any abdominal or pelvic mass. Chromosome study from peripheral leukocyte culture consistently showed 46 XY normal karyotype and intravenous pyelogram showed normal kidney and pyelocalyxial system. A laboratory examination revealed high levels of FSH and LH, 56 mIU/ml and 16 mIU/ml respectively. Estrogen secretion was low (under 20 pg/ml) and low testosterone level was also noted (0.9 ng/ml). She was counseled to undergo a bilateral gonadectomy and laparoscopy was scheduled along with intraoperative ultrasonogram to search for possible intraabdominal tumor.

A 10 mm. diagnostic laparoscope attached to a video system was introduced through an intraumbilical incision. Grasping forceps and a bipolar forceps were introduced into the abdomen through separate 5- mm. sites. Both streak, whitish shiny oval gonads, 5 x 20 mm. and 4 x 15 mm. on the left and right side respectively, were found to attach to fallopian tubes. Hypoplastic uterus and absent of ascites were also noted. The fallopian tubes with middle part communicated were inverted U shape. Total length was approximately 11 cm. Both ends looked fimbria-like. Round ligament, about 3 cm. long each looked like an arc with normal general appearance. Retroperitoneal space was thoroughly explored and both kidneys looked normal. No detectable abdominal lymph nodes. Light illumination from laparoscope and intra-operative ultrasonogram in the area of inguinal canal was done to search for possible muellerian remnants and revealed no abnormal mass. The bipolar cautery forceps were applied to the infundibulopelvic ligament and parietal peritoneum to which the gonads were attached and these sites were cauterized and divided by laparoscopic scissors. The procedure was carried out further until the proximal tube and utero-ovarian ligament was reached and these sites were then cauterized and resected. Bleeding point was checked and the specimens were removed through the 10 mm. sheath by removing the laparoscope through the sheath and processed for frozen histological examination to ensure the correct gonadal tissues. Both gonads consisted of fibrous streaks with ovarian stromal tissue without primordial follicles or testicular seminiferous tubules. No malignant alteration is shown. Estimated blood loss was about 30 cu.mm. The patient recovered well postoperatively, the wound healed completely and no analgesic was needed. The patient was discharged without complication on the 3rd postoperative day after adequate rest and observation.

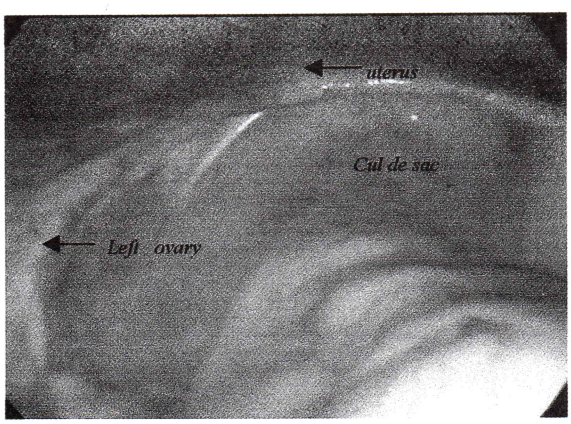

Fig. 1. Internal genital organ

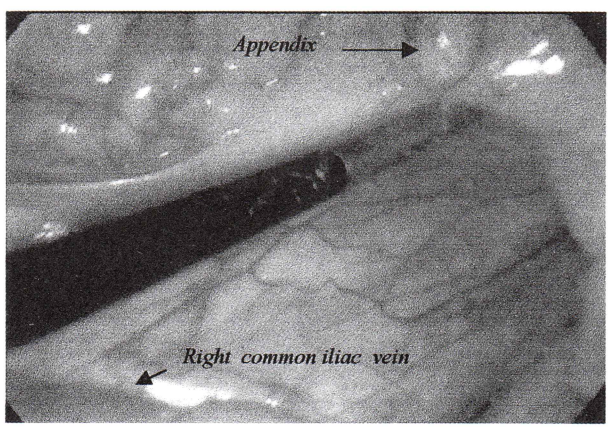

Fig. 2. Exploring ureter

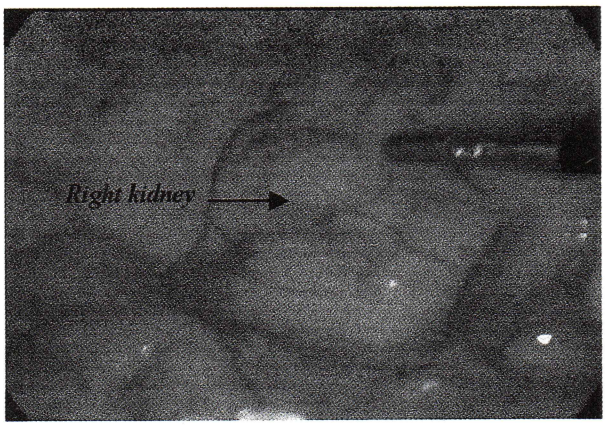

Fig. 3. Exploring right kidney

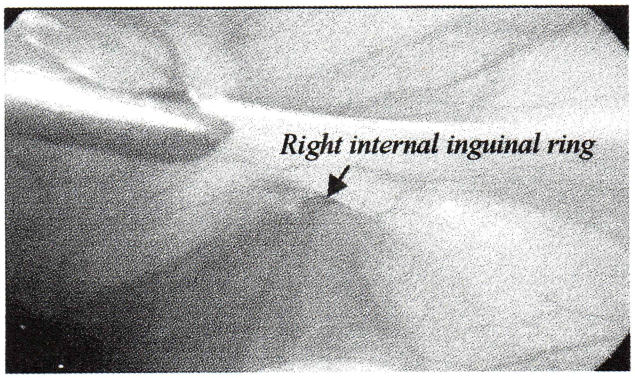

Fig. 4. Probing right internal inguinalring

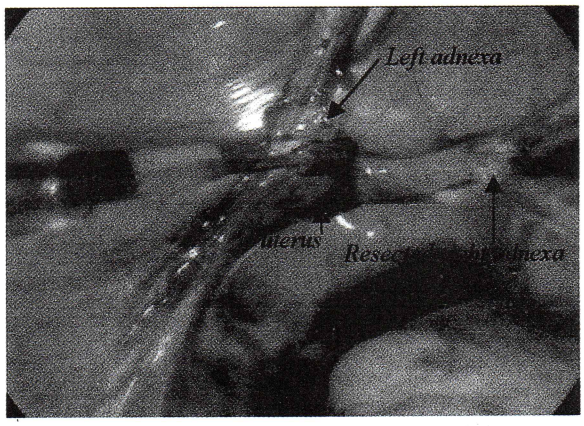

Fig. 5. Left adnexectomy

Discussion

Gonadal dysgenesis, also known as Swyer syndrome,[1] consists of a spectrum of disorders with persistent Muellerian duct structures. The diagnosis of 46 XY pure gonadal dysgenesis is accomplished by combination of features: female phenotype, absent of somatic sign of Turner's syndrome, absent of follicles in the streak gonads and presence of fallopian tubes and uterus. The syndrome is governed by the abnormality in the sex determining region on the Y chromosome (SRY). However patients with apparently normal SRY can also be found.[5,6] The risk of gonadal dysplasia is very high, estimated to be 25% [7,8] dictating early prophylactic removal of these dysgenetic gonads. Gonadoblastoma and dysgerminoma are the most frequently reported malignancy.[5]

Purtuondo et al reported using laparoscopy to perform ovarian biopsy followed by conventional laparotomy and gonadectomy.[9] With progression of endoscopic procedures, laparoscopic gonadectomy becomes increasing alternative to conventional laparotomy.[6,10,11]

Laparoscopic gonadectomy can not only minimize operative time, the extent of surgery thus good cosmetic result, peri- and postoperative drug needs, leads to much rapid postoperative recovery, but also minimizes hospital stay and expense and enhances doctor and patients' satisfaction in comparision with conventional laparotomy.

Acknowledgments

The authors are grateful to Dr.Bunasit Boonitt, the former director of Bangkok Christian Hospital and Dr.Panthip Viriyapanich for their fruitful discussion and thank to Mr.Chumchet Panichijiwasan from IVS Industry Vision Co., Ltd. and Mr. Naruecha Suewaja from Sony Thai Co.,Ltd. for their sophisticate instruments provided.

References:

1. Swyer GIM, Phil D. Male pseudohermaphroditism: A hitherto undescribed form.Br Med J 1955, 2: 709
2. DeWhurst J. Genetic and congenital sexual disorders. In; Gynecological therapeutics. Hawkins DF, editor. London: Bailliere Tindall, 1981:1
3. MacMahon RA, Cussen J ,Walters WAM. Importance of early diagnosis and gonadectomy in 46XY females. J Pediatr Surg 1980; 15:642
4. Jagiello G, Atwell JD: Prevalence of testicular feminization. Lancet 1962;1:329
5. Coutin AS. Hamy A. Fondevilla M. Savigny B. Paineau J. Visset J. Pure 46XY gonadal dysgenesis Journal de Gynecologie, Obstetrique et Biologie de la Reproduction 25(8): 792
6. Tsutsumi O. Iida T. Taketani Y. Laparoscopic surgery and DNA analysis in patients with XY pure gonadal dysgenesis. Journal of Obstetrics & Gynecology. 21: 67-74, 1995 Feb.
7. Schllhas H. Malignant potential of the dysgenetic gonad. I. Obstet Gynecol 1974; 44: 298
8. Troche V. Hernandez E. Neoplasia arising in dysgenetic gonads. Obstet Gynecol Surv 1988; 41: 74
9. Portuondo JA, Neyro JL, Barral A, Gonzalez-Gorospe F, Benito JA: Management of phenotypic female patients with an XY karyotype. J Reprod Med 31: 611, 1986
10. Droesch K. Droesch J. Chumas J. Bronson R. Laparoscopic gonadectomy for gonadal dysgenesis Fertil Steril 1990; 2:360
11. Arici A. Kutteh W H . Chantilis S J. Johns D A. Carr B R. Laparoscopic Removal of gonads in women with abnormal karyotypes J Reprod Med 1993;7:521