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Laparoscopic Gonadectomy in Patients With Androgen Insensitivity Syndrome: Case Series

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Background: The gonadectomy in patients with androgen insensitivity syndrome (AIS) has been conventionally performed using laparotomic techniques. But currently several reports have suggested laparoscopic gonadectomy in these patients because of shorter hospital stay and minimal postoperative pain and hemorrhage.
Objectives: To report the laparoscopic gonadectomy in 11 patients suffering from androgen insensitivity syndrome
Patients and Methods: This study is a case series of 11 patients with AIS aged between 17 and 20 years who were referred to the Shiraz University of Medical Sciences affiliated clinics and private hospitals from February 2006 to December 2012 and underwent bilateral laparoscopic gonadectomy. The patient's outcome and results were reported in this study.
Results: Overall we operated 11 patients suffering from AIS with mean age of 18.6 ± 3.8 (17-20) years. All of the patients presented with primary amenorrhea. The serum levels of testosterone were in normal range of male population. All the patients had an uneventful laparoscopic gonadectomy and no complications occurred during the operation and in the postoperative period. The mean operation duration was 40.3 ± 5.9 (range 55 to 35) minutes. The hospital stay duration was 1.6 ± 0.9 days. All of the patients were followed for at least 6 months after operation.
Conclusions: Laparoscopic gonadectomy is a safe, simple and effective procedure for the removal of the testis in patients with AIS. We recommend utilizing this method for gonadectomy in patients with AIS where facilities are available.

Keywords: Laparoscopy; Androgen Insensitivity; Testicular Feminization

1. Background

The androgen insensitivity syndrome (AIS) is a rare X-linked recessive disorder recognized by XY genotype but female phenotype with an estimated prevalence of 1 in 20,000 living birth (1). The disease is caused by defective androgen receptors leading to resistance to systemic androgens (2). Clinically the disease is classified as complete (complete resistance to androgens) and incomplete (partial resistance to androgens). Patients suffering from AIS are at increased risk of gonodal cancer. Previous reports have shown that approximately 20-30% of patients with AIS with preserved gonads develop gonadal cancer. In order to prevent malignant change in the abdominal testicles, prophylactic bilateral gonadectomy is recommended for all the patients with AIS. However the operation is usually postponed till puberty in order to allow spontaneous feminization and avoid the need of hormone replacement therapy. In addition, the risk of malignant changes of testicles is minimal before the puberty (3, 4). The gonadectomy in patients with AIS has been conventionally performed using laparotomic techniques. But currently several reports have suggested laparoscopic gonadectomy in these patients because of shorter hospital stay and minimal postoperative pain and hemorrhage (3, 5, 6). However the technique is time consuming and requires qualitative surgical skills and laparoscopic equipment which are not available in all centers (7).

2. Objectives

In this study we describe the method and outcome of laparoscopic gonadectomy in 11 patients suffering from AIS.

3. Patients and Methods

This study is a case series of 11 patients with AIS aged between 17 and 20 years who were referred to the Shiraz University of Medical Sciences affiliated clinics and private hospitals from February 2006 to December 2012 and underwent bilateral laparoscopic gonadectomy. All the patients were diagnosed according to cytogenetic and hormonal investigations and underwent complete systemic and gynecologic examinations, including ultrasonography. Pelvic ultrasonography and karyotyping were done as diagnostic tests, and urinary tract abnor-

malities were ruled out by means of intravenous pyelography. Hormonal assays included estrogen, testosterone (T), luteinizing hormone (LH) and follicular stimulating hormone (FSH). The study protocol was approved by institutional review board and ethics committee of Shiraz University of Medical Sciences and all the recruited patients provided their informed written consents. Psychiatry consultation was requested for all the patients before the operation and the social and familial consequences were fully described to the patients. All the patients were found to have normal female phenotype. All the patients had normal female body shape, breasts and external genital appearance, but all the patients lacked ovaries and uterus. The gonads were found in different positions in the abdomen, from the entrance of inguinal canals in the abdomen to the abdominal cavity or inguinal canal. The serum levels of T and LH were higher than normal range for women while the FSH level was normal in all the individuals. The intravenous pyelography was normal in recruited patients and the karyotype was 46 XY in all patients.

Laparoscopic gonadectomy was performed in all the patients under general anesthesia by means of a three ports technique (8). All laparoscopic gonadectomy were performed by one laparoscopic gynecologist (S.A.). After carbon dioxide insufflation, a 10-mm trocar and sleeve were inserted infra-umbilically. The whole abdominal cavity was visualized with a 10-mm endoscope. Two 5 mm suprapubic trocar and sleeve were then inserted. Then the pedicle of the gonad, the spermatic cord, and the course of ureter were identified. The pedicle was coagulated with bipolar diathermy and cut with laparoscopic scissors. With taking into consideration the possibility of malignancy and to prevent spillage of tumor cells and contamination, the gonads were placed in an endobag and removed intact after extending the suprapubic port. In two patients with gonads in the inguinal canal, we applied a transinguinal approach for removing the gonads via laparoscopy. A long-term hormonal replacement treatment was initiated postoperatively using orally conjugated estrogens.

4. Results

Overall we operated 11 patients suffering from AIS with mean age of 18.6 ± 3.8 (17-20) years. The characteristics of the patients are summarized in Table 1. All of the patients presented with primary amenorrhea. Breasts were present in all the patients. The mean depth of vagina in these 11 patients was 2.9 ± 1.6 (range 0.5 to 7) cm. Most of the patients (81.8%) had intra-abdominal gonads while in 2 (18.2%) patients, one of the gonads was intra-abdominal and the other was in the inguinal canal. The serum levels of testosterone were in normal range of male population. All the patients had an uneventful laparoscopic gonadectomy and no complications occurred during the operation and in the postoperative period. The mean operation duration was 40.3 ± 5.9 (range 35 to 55) minutes. The hospital stay duration was 1.6 ± 0.9 days. All the patients had normal examinations in a 6 months follow-up period.

Table 1. The Characteristics of 11 Patients Suffering From Androgen Insensitivity Syndrome (AIS) Undergoing Laparoscopic Gonadectomy in Our Center

Variable	Value (n = 11)
Age, y, Mean ± SD	18.6 ± 3.8
Amenorrhea, No. (%)	11 (100)
Clitoromegaly, No. (%)	3 (27.7)
Normal breasts, No. (%)	11 (100)
Pubic/Auxiliary hair, No. (%)	8 (72.7)
Uterus, No. (%)	0 (0.0)
Vagina, cm, Mean ± SD	2.9 ± 1.6
Normal, No. (%)	2 (18.2)
Blind, No. (%)	4 (36.4)
Short, No. (%)	5 (45.4)
Gonads' location, No. (%)	
Intra-abdominal	9 (81.8)
Inguinal canal and intra-abdominal	2 (18.2)
LH, No. (%)	
High	11 (100)
Normal	0 (0.0)
FSH, No. (%)	
High	10 (90.9)
Normal	1 (9.1)
Testosterone, No. (%)	
Normal for Male	11 (100)
Normal for Female	0 (0.0)
Estrogen, No. (%)	
High	1 (9.1)
Normal	10 (90.9)
Progesterone, No. (%)	
High	2 (18.2)
Normal	9 (81.8)
Operation duration, min, Mean ± SD	40.3 ± 5.9
Hemorrhage, mL, Mean ± SD	46.8 ± 10.9
Hospital stay, days, Mean ± SD	1.6 ± 0.9

5. Discussion

This was a case series reporting laparoscopic gonadectomy in 11 patients suffering from AIS. We showed that the results and outcomes of laparoscopic gonadectomy in patients with AIS is acceptable and none of our patients developed intraoperative or postoperative complications and the examinations were favorable after 6 months of operation. The results of this study are in con-

sistence with previous reports of laparoscopic gonadectomy in these patients (6-12). The gonads in patients with AIS are at increased risk of developing gonadal cancer. Thus clinicians recommend prophylactic gonadectomy in all the patients (3). The operation should be postponed to the end of the second decade of life in order to let the patients develop the secondary female sex characters. Most of the authors believe that the most appropriate age for prophylactic gonadectomy is 16 to 18 years. In addition, in most of the patients the diagnosis is not made before this age because most of the patients present with primary amenorrhea (6). On the other hand, sometimes the gonadectomy is indicated because of virilization, hirsutism or clitoromegaly in younger age. In these cases, administration of high dose estrogen after gonadectomy is indicated to assist developing the secondary female sex characteristics.

Previously, laparotomic gonadectomy was considered the standard method of surgery in these patients. For this purpose, exploratory laparotomy followed by total abdominal hysterectomy with bilateral gonadectomy was performed. However, during the previous decade, the practice of laparoscopic gonadectomy in patients with AIS has increased dramatically (11-13). In this report we also removed all the intra-abdominal and inguinal gonads using laparoscopic technique. The results and outcome was favorable in all the patients. Laparoscopic gonadectomy encompass many advantages over laparotomic procedure including rapid recovery, minimal blood loss, shorter hospital stay and minimum psychological trauma (8-12). It is also easier for the surgeon to localize the gonads via laparoscopy due to the better visualization of the entire abdomen and pelvis. All these advantages make laparoscopy the method of choice for removing the gonads in AIS patients. However this point should be kept in mind that laparoscopic procedures need appropriate medical facilities and skilled surgeons that are not available in all the centers. In a study by Yalinkaya et al. (10), a laparoscopic technique for gonadectomy in patients with AIS was introduced. In this method, they applied a transinguinal approach for removing the gonads via laparoscopy (laparascopy-assisted transinguinal extracorporeal gonadectomy). It is similar to our procedure for two inguinal gonads that we had in our cases. This technique has several advantages over laparotomy and operative laparoscopy, including short operation time, simplicity, and cost-effectiveness (10).

In conclusion, laparoscopic gonadectomy is a safe, simple and effective procedure for the removal of dysgenetic gonads in patients with AIS. We recommend utilizing this method for gonadectomy in patients with AIS where facilities are available.

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Author's Contributions

Saeed Alborzi performed the laparoscopic gonadectomy; Zohre Tavana: concept and design, and manuscript preparation; Malihe Amini, Gathered the data and prepared the manuscript.

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