THE JOURNAL OF OBSTETRICS AND GYNECOLOGY OF INDIA

The Journal of Obstetrics and Gynecology of India (May–June 2016) 66(3):196–198
DOI 10.1007/s13224-015-0716-7

CASE REPORT

Laparoscopic Gonadectomy and Hernia Repair for Complete Androgen Insensitivity Syndrome (CAIS): A Rare Cause of Primary Amenorrhea

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Received: 5 September 2014 / Accepted: 22 May 2015 / Published online: 11 June 2015

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Case Reports

Case I

A 26-year-old phenotypic sexually active female presented with primary amenorrhea and 5 years infertility. Patient's height was 171 cm and weight 70 kg. Physical examination showed normal development of breast (Tanner stage 2) with no pubic and axillary hairs and blind vaginal pouch of 2 cm.

MRI revealed a hypoplastic uterus with an absent cervix and upper vagina and two oblong enhancing soft tissue structures with small cysts in both inguinal canals adjacent

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the round ligament. Preoperative level of testosterone (830.09 ngm/dl) was high. Karyotype was 46XY. A working diagnosis of complete androgen Insensitivity syndrome (CAIS) was made. Informed consent was obtained for removal of gonads with bilateral inguinal hernia repair. Patient was counseled for hormone replacement therapy.

At laparoscopy, the left gonad appeared cystic. It was lying partially in the abdomen. The right gonad was totally in the inguinal canal. Laparoscopic bilateral orchidectomy with transabdominal preperitoneal (TAPP) hernia repair was performed (Fig. 1). Histopathology confirmed testicular tissue.

Case II

28-year-old unmarried female presented with primary amenorrhea and backache. Physical examination showed normal breasts, no axillary and pubic hairs, blind Vaginal pouch, and right labial soft tissue swelling.

Hormonal analysis showed FSH—79.98 IU, LH—33.86 IU, AMH—0.52 µgm/ml, total testosterone—730.12 ngm/dl, and XY Karyotype. Patient was counseled and informed consent obtained for gonadectomy with right-sided inguinal hernia repair.

At laparoscopy, right gonad was in inguinal canal which was removed (Fig. 2) and herniorraphy done. Left gonad was intraabdominal over left external iliac vessels removed with help of harmonic scalpel.

Discussion

CAIS is a X—linked recessive condition. Normally, testosterone acts on androgen receptors in the Wolffian ducts which develops into the epididymides, vasa deferentia, and seminal vesicles. In CAIS since the androgen cannot act on receptors either completely or partially,

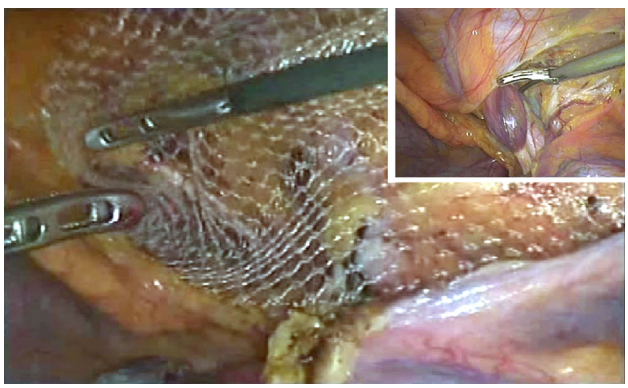

Fig. 1 Case 1—Hernia repair with mesh (*inset* Right gonad in inguinal canal)

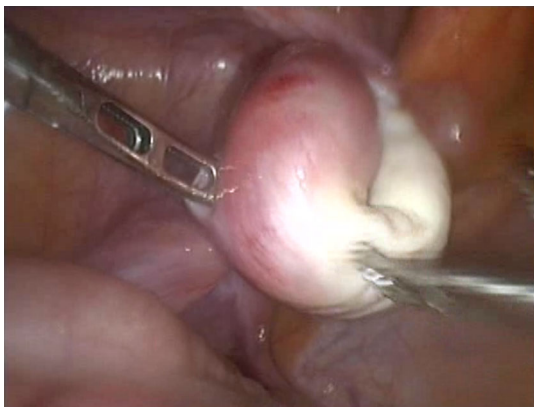

Fig. 2 Case II—Right gonad dissected from inguinal Canal

virilization fails to occur. Hence, a genetic male presents as a phenotypic female, with features that range from a mild defect to complete female habitus and female external genitalia (Quigley scale—1 through 7), despite the presence of a Y-chromosome.

The first medical report on androgen insensitivity syndrome (AIS) was published in 1953 by J. S. Morris, an American Gynecologist (Morris syndrome) [1]. The incidence of CAIS occurs in 1 out of 20,000 births and can be incomplete with various sexual ambiguities, or complete female phenotype [2]. It is 10 % in patients with primary amenorrhea and 1.1 % in females presenting with inguinal swelling. A CAIS patient usually presents with primary amenorrhea at puberty and infertility.

The risk of germ cell malignancy is relatively high in these patients, increasing with age. The incidence (dysgerminoma, gonadoblastoma) is as low as 0.8 % in CAIS and 5.5 % in AIS. Overall risk of malignancy increases markedly after puberty and reaches 33 % by the age of 50 years [3]. Gonadectomy is recommended after postpubertal period to help development of feminization during puberty. Malignant changes in the germ cells are relatively late [4]. Gonads may lie anywhere in the path of congenital descent. They are usually associated with inguinal hernia in 50 % of patients. Hernioplasty should be undertaken to prevent future obstructed hernia.

Laparoscopic approach to gonadectomy is an excellent minimally invasive technique. It provides good visualization of internal genitalia, and even small gonads and mullerian structures can be removed. Postoperatively, recovery is rapid with minimal discomfort to patient affording early mobilization.

Gonadectomy predisposes patients to osteoporosis and other menopausal symptoms; therefore, hormonal replacement therapy (HRT) is prescribed till the natural age of menopause to prevent the associated accelerated aging process of the whole body.

In our cases, both patients presented with primary amenorrhea and infertility. Ectopic gonads were in the inguinal canal. After explaining the risk of malignancy, laparoscopic bilateral orchidectomy with herniplasty was performed. Patient was started on HRT because of the associated risks of osteoporosis and cardiac problems.

Conclusion

CAIS should be excluded in patients with primary amenorrhea and primary infertility. Gonadectomy is recommended to prevent gonadal malignancies. Laparoscopic approach to gonadectomy is an excellent minimally invasive technique. Hormonal replacement therapy (HRT) should be given to prevent osteoporosis.

Compliance with ethical requirements and Conflict of interest There is no conflict of interest with any pharmaceutical company or hospital. There is no conflict of interest between authors. The authors have no financial disclosure to make.

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