CASE REPORT

Laparoscopic hysterectomy in a case of male pseudohermaphroditism with persistent Müllerian duct derivatives

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We describe laparoscopic diagnosis and treatment for a case of dysgenetic male pseudohermaphroditism with persistent Müllerian ducts. The patient, a 32 year old man, with a history of surgery for hypospadias and cryptorchidism during childhood, was referred because of anejaculation. He was of short stature, with male external genitalia composed of a small penis and hypoplastic testis (1 ml right, 6 ml left side). Plasma follicle stimulating hormone (FSH) was high (17 mUI/ml), testosterone low (1.9 ng/ml), and his karyotype was 46,XY. Pelvic ultrasound, nuclear magnetic resonance (NMR) and genitography disclosed a uterine-like structure with cavity communicating with the urethra. Laparoscopy and urethrocystoscopy confirmed the presence of a 4 cm uterus, which was removed endoscopically at the same time. A biopsy of the left gonad was also performed. The uterus contained endometrial tissue and was fibrotic. Histology of the left gonad showed spermatocytic arrest. We diagnosed dysgenetic male pseudohermaphroditism. Laparoscopy, in our opinion, is an optimal tool to diagnose and treat abnormal sexual conditions.

Key words: dysgenetic male pseudohermaphroditism/hysterectomy/laparoscopy/male pseudohermaphroditism

Introduction

The management of intersex states includes medical, surgical, psychological and legal issues. Combined surgical and medical treatment is often used to correct the genitalia in concordance with the patient’s desire (Josso, 1981). Surgical correction of internal and/or external genitalia is for cosmetic and functional goals, and also to prevent degenerative changes in the vestigial remnants of gonads or internal primitive ducts (Wolf or Müller ducts) (Scully, 1981). The ablation of Müllerian duct derivatives (uterus, tubae) is indicated in intersex cases reared as males. Usually in these cases hysterectomy is carried out by laparotomy, even if a laparoscopical approach has been demonstrated to be effective and safe in gynaecological practice (Gomel, 1995). Laparoscopy has been considered an ideal diagnostic technique for patients with sexual ambiguity (Minozzi et al., 1975; Sinisi et al., 1987). There are very few reports of laparoscopic surgery for intersex problems (Wilson et al., 1992; McDougall et al., 1993). Here we report a case of dysgenetic male pseudohermaphroditism with persistent Müllerian ducts diagnosed and treated by an endoscopic approach (laparoscopy and urethrocystoscopy).

Case report

The patient, a male of 32 years old, was referred to the Institute of Endocrinology owing to infertility due to anejaculation. The family history was uninformative except for non-consanguinity of parents and the presence of a normal brother and sister. He had a history of repeated surgery during childhood for perineal hypospadias and cryptorchidism, with apparently normal puberty and sexual activity.

The male was 142 cm high, weighed 60 kg, with an adult male pattern of pubic hair, a stretched penile length of 3.5 cm with a normal site of urethral ostium and both testes were in the scrotum, the right being highly hypotrophic, 1 ml in volume, the left 6 ml in volume. There was no gynaecomastia. His hormonal profile was: follicle stimulating hormone (FSH) 17.5 mUI/ml (normal 2–11); luteinizing hormone (LH) 6 mUI/ml (normal 1.5–9); testosterone 1.9 ng/ml (normal 2.4–8.0); oestradiol 21 pg/ml (normal 15–60); cortisol 180 ng/ml (normal 80–150).

Pelvic ultrasound and nuclear magnetic resonance (NMR) showed the presence of a uterus-like structure (5.6×1.4 cm), the absence of ovaries, and a prostate-like organ. The genitography showed normal kidneys and urinary tracts. The karyotype was 46,XY in 200 Giemsa Trypsin Leishman (GTL)-banded metaphases from peripheral lymphocytes and skin fibroblast cultures.

With the informed consent of the patient, we decided to perform laparoscopy and, if necessary, surgical removal of the internal female structure. Moreover, to prevent high-risk neoplastic degeneration of the gonads we planned a right gonadectomy and biopsy of the left testis. The patient consented to left biopsy only.

At laparoscopy a small uterus (4 cm in length) was visible on the median line; two Fallopian-like tubes coursed bilaterally from the uterine fundus through the internal inguinal orifices. Uterosacral ligaments and gonads were not seen on either side. The uterine–bladder fold overlayed the uterine fundus, whereas the anterior uterine wall was extraperitoneal. Contem-
poraneous urethrocystoscopy revealed the presence of a stoma at the upper third of the urethra (Figure 1), providing opening to a recess with at its end a structure similar to a uterine cavity. This structure was catheterized for about 4 cm. The upper third of the ‘vagina’ had a stoma in the proximal urethra as an opening. The absence of prostate and seminal vesicles was confirmed. The uterus was removed by laparoscopy (Figure 2). This followed the incision and detachment of the bladder–uterine fold using forceps, and dissection of the tubes, round ligaments and uterine artery with scissors and bipolar tweezers. The organ was detached from the vaginal recess by means of endogya and removed in an endoscopic sac through an enlarged ancillary channel. The urethrocystographic check confirmed that the suture of the vagina had held. At the end of laparoscopy, a left testicular biopsy was carried out using the classical technique. Pathological examination revealed a normal uterus with bilateral Fallopian tubes and atrophic endometrial tissue. The left gonad was diagnosed as testis with spermatogenesis arrest at the spermatocyte stage. A search for carcinoma in situ (CIS) was negative.

Discussion

There are three main conditions of intersex characterized by persistent Müllerian duct derivatives: persistent Müllerian duct syndrome (PMDS), mixed gonadal dysgenesis (MGD) and dysgenetic male pseudohermaphroditism (DMP) (Zah et al., 1975; Brook, 1981; Rappaport and Forest, 1993). The short stature, the presence of abnormal genitalia and retained Müllerian structures in our subject excluded the condition of PMDS. Laparoscopic examination of internal gonads revealing dysgenetic streak gonads on one side suggests the diagnosis of MGD; however, histological sections of the biopsy revealing a combination of both immature hypoplastic testicular tubules and persistent stromal tissue characteristic of ovarian stroma but lacking primary ovarian follicles, classifies the gonad as a dysgenetic testis and suggests the diagnosis of DMP (Brook, 1981). The laparoscopic exploration excluded, in our case, the presence of a streak gonad on the right side. The lack of evidence of primitive dysgenetic stromal tissue within the left gonad biopsy specimen did not preclude the diagnosis of DMP in our case. In fact, it underlines that the histological differentiation between a dysgenetic testis and a normal cryptorchid testis may be very difficult, if the entire dysgenetic testis has not been examined (Brook, 1981). Moreover, our patient did not consent to biopsy and histological examination of the right testis, which appeared more hypoplastic, but consented only to biopsy of the left testis and hysterectomy.

Surgical removal of gonadal or internal genitalia is recommended for the prevention of neoplastic transformation, bleeding or infectious complications. The patient did not present bleeding because of endometrial atrophy probably due to the absence of oestrogenic stimulation. Absence of CIS following histology and the scrotal position of both gonads did not indicate a total lack of neoplastic risk. Continuous clinical, sonographic and laboratory surveillance should be performed.
regularly to reduce, but not eliminate, the risk (Scully, 1981; Savage and Lowe, 1990). Laparoscopy in our patient provided an appropriate visualization of internal genitalia and a simple and minimally invasive therapeutic approach. In fact abdominal incision was avoided and after 2 days the patient was discharged. To date, there have been few reports of laparoscopic surgery for intersex problems. In both of the cases described in the literature laparoscopy was undertaken to perform prophylactic gonadectomy (Wilson et al., 1992; McDougall et al., 1993). Laparoscopic hysterectomy has been practised since 1989 in gynaecological surgery (Gomel, 1995). To our knowledge, there are no cases of males with persistent Müllerian duct remnants treated with laparoscopy. This procedure resulted in a brief and uncomplicated postoperative course.

References

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