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CASE REPORT

Transumbilical laparoendoscopic single-site gonadectomy for Turner's syndrome with Y-chromosome mosaicism

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Abstract Laparoendoscopic single-site surgery (LESS), a minimally invasive procedure, is gaining widespread acknowledgment in pediatric urology. We report the case of a 7-year-old girl with Turner's syndrome with 45,XO/46,XY mosaicism, for whom bilateral prophylactic gonadectomy using LESS was performed. Histopathological findings revealed bilateral streak gonads. The surgical and cosmetic outcome was excellent. Diagnostic and therapeutic laparoscopy is essential for accurate clinical management of patients with disorders of sex development. Although this is only a single case report, it supports the theory that LESS is an exceedingly practical and superior technique for such children, since it provides excellent magnification, as well as allowing normal psychological development owing to the concealed scar. Further studies and long-term follow-up are required to evaluate the benefits and limitations of applying LESS in pediatrics.

Introduction

Turner's syndrome (TS) is a relatively common chromosomal disorder, affecting 1 in 2000 live female births [1]. Because more than 30% of patients with a Y-chromosome risk developing gonadoblastoma, the prophylactic gonadectomy of streak gonads is recommended [2].

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The minimal invasive nature, absence of scars, efficacy and low morbidity make laparoscopy an optimal modality for the evaluation and treatment of patients with disorders of sex development (DSD). Recently, laparoendoscopic single-site surgery (LESS) has been performed in pediatric urologic surgery, accompanied by modifications to laparoscopic devices [3,4]. In the present case, we performed single-site transumbilical laparoendoscopic gonadectomy for a girl with TS with Y-chromosome mosaicism.

Case report

A 7-year-old girl of short stature was presented to the pediatric outpatient department of our hospital. She was 103.6 cm (−2.5 SD) tall and weighed 16.0 kg (−1.5 SD). The external genitalia and psychological examination did not reveal any abnormality. Endocrine evaluation showed a high follicle-stimulating hormone level (70.5 mIU/mL; normal female 2.0−8.3 mIU/mL). G-banding showed a 45,XO/46,XY karyotype, and abdominal ultrasonography and MRI revealed a rudimentary uterus but no gonads. There were no responses of serum testosterone and estradiol after hormonal stimulation. For an accurate diagnosis, we performed LESS to evaluate gonad maturation and identify internal duct derivatives.

Under general anesthesia, a 25-mm skin incision, cephalad to the umbilicus, was made and a SILS™ port (Covidien, Mansfield, MA, USA) was placed (Fig. 1A). Observation of the inner abdominal cavity using a 5-mm 30° laparoscope revealed a properly maturing uterine structure and two Fallopian tube like structures, leading to bilateral streak gonads. Because these gonads were underdeveloped and were not expected to function normally, bilateral gonadectomy was performed using a 5-mm Roticulator Endo Dissect™ and LigaSure™ vessel sealing system (Covidien) via the SILS™ port (Fig. 1B−D). Histopathological examination of the bilateral gonads showed interstitial ovarian tissue, but without ovarian follicles or malignant cells (Fig. 2). Thus, the patient was diagnosed with TS with Y-chromosome mosaicism. There were no postoperative complications, and the incision scar was almost indistinguishable from the umbilicus after 2 months (Fig. 3).

Discussion

We performed a bilateral gonadectomy with LESS in a case of TS with Y-chromosome mosaicism and obtained a clinically excellent outcome. LESS offers the advantage of reducing the number of abdominal incisions to one. While caring for patients with DSD, special attention should be given to their psychological development. Patients with DSD often have psychiatric vulnerabilities, such as anxiety or mood disorders, and suffer from gender identity disorder or gender dissatisfaction [5]. Because hospitalization, examination procedures and surgeries stress children of all ages, we should strive to reduce the burden. From this standpoint, we consider LESS the most feasible procedure for pediatric DSD patients.

In the present case, we performed the surgery safely by using flexible laparoscopic instruments and endoscope, without any postoperative complication. Moreover, since this patient achieved an excellent cosmetic appearance, she and her parents are satisfied with the postoperative

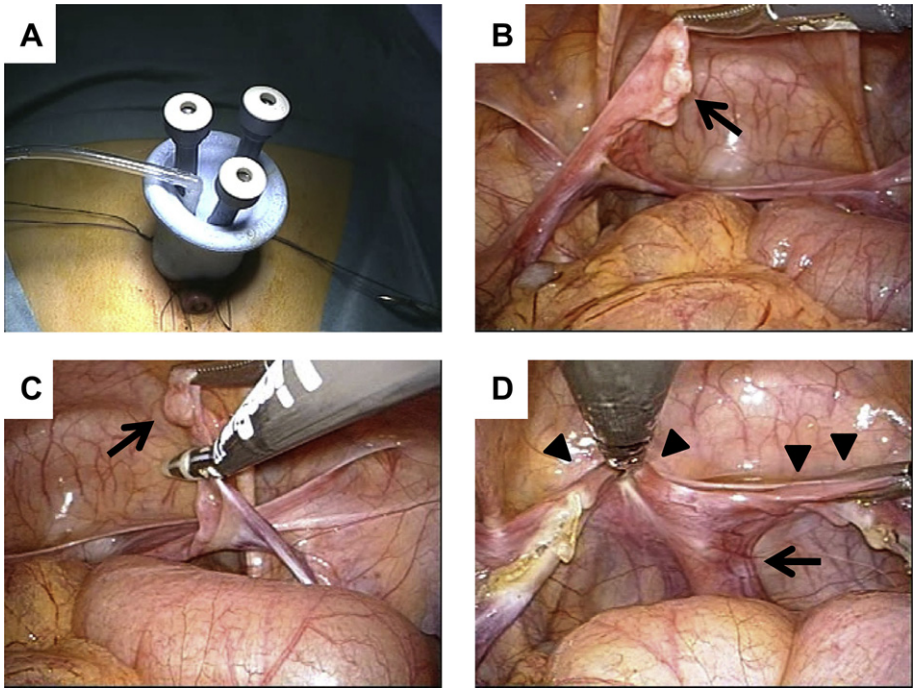

Figure 1 Laparoendoscopic single-site gonadectomy. (A) A SILS™ port containing gas insufflation channel and three individual 5-mm cannulas. (B) Left gonad lined by Fallopian tube and vessels (arrow). (C) Right gonad (arrow). Bilateral gonads were removed using 5-mm LigaSure™ vessel sealing system. (D) Bilateral sealing stump, Fallopian tube (arrowheads) and uterine structure (arrow).