CASE REPORT

Bicornuate Uterus with Complete Cervico-Vaginal Agenesis and Skeletal Deformity: A Case Report

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Received: 10 November 2017 / Accepted: 1 March 2018 / Published online: 15 March 2018
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Introduction

Mullerian duct anomalies are one of the most complex female genital tract disorders encountered by obstetricians and gynecologists. The mullerian ducts are differentiated, fused and then canalized to develop fallopian tubes, uterus, cervix and the upper vagina, while the lower part of vagina develops from urogenital sinus. The disruption in the development of mullerian system can lead to a wide variety of uterine and vaginal abnormalities. The American Fertility Society (AFS) classification was previously the most widely accepted for categorizing mullerian duct abnormalities and included seven classes of mullerian anomalies. In 2013, ESHRE consensus introduced the CONUTA classification which classifies the mullerian anomalies into uterine (U), cervical (C) and vaginal (V) abnormalities [1]. This classification has now replaced the AFS classification. Functioning uterus with cervical and vaginal agenesis can lead to the development of hematometra, hematosalpinx,

Fig. 1 a Figure showing associated skeletal deformity. White arrow shows hypoplastic thumb. Black arrow shows hypoplastic toes. **b** MRI image showing widely apart bilateral uterine horns with absent cervix and vagina

endometriosis and even chronic pelvic pain. The prevalence of congenital vaginal agenesis is 1 in 4000–5000 live births [2], whereas the prevalence of congenital cervical agenesis is 1 in 80,000–100,000 live births [3]. Only 7–8% of patients with vaginal agenesis have a functional uterus.

Case Report

A 13-year-old girl presented in our gynecology outpatient department with complaints of abdominal pain for the past 10 months which was cyclical, occurring every month. Initially, pain was relieved with oral analgesics, but later required injectables. Menarche was not attained. On general physical examination, she was average built and secondary sexual characters were age appropriate (breast and pubic hair Tanner IV and axillary hair present). Her bilateral upper and lower limbs had deformity in the form of hypoplastic bilateral thumb, great and little toe (Fig. 1a). Systemic examination did not reveal any cardiac, CNS or spine abnormality. On X-ray, bilateral middle phalanx of fingers were hypoplastic; bilateral capitate and hamate bones were fused. Her karyotype was 46 XX, and hormonal profile was normal.

On per abdominal examination, abdomen was soft with no palpable mass; local examination revealed a patulous urethra with blind vagina. She had an MRI report which showed two functional uterine horns, appearing divergent in hemipelvis. The cervix and vagina were not visualized and showed only T2 hypointense soft tissue. Bilateral ovaries appeared normal (Fig. 1b). There was no renal

abnormality detected on imaging, and her liver function and kidney function tests were within normal limits. A diagnostic laparoscopy was done which revealed findings of bicornuate uterus with enlarged uterine horns separately placed in bilateral iliac regions without any intervening connecting tissue. Besides hematometra, there was 100 ml of hemoperitoneum. The cervix was completely absent. Bilateral tubes and ovaries were normal (Fig. 2). The parents were explained about the condition and were counseled regarding the need for hysterectomy. Bilateral uterine horn excision was done laparoscopically with the preservation of bilateral tubes and ovaries. Cut section of uterus showed two blind endometrial cavities. Postoperative period was uneventful. Histopathological examination confirmed the secretory phase of endometrium. In the postoperative period, she has been in regular follow-up and is pain free.

Discussion

The occurrence of bicornuate uterus with functioning endometrium along with complete cervico-vaginal agenesis is a rare finding. Management in these cases is a challenge for gynecologists, and the decision needs to be individualized as it not only has a huge psychosocial impact but also affects the future fertility of the patient. The treatment options include utero-vaginal anastomosis along with metroplasty or excision of rudimentary horn and in rare cases straightforward hysterectomy. The present case was symptomatic in terms of cyclical pain,

Fig. 2 a Laparoscopic view of widely divergent bilateral uterine horns (black arrows) along with hemoperitoneum. **b** Schematic diagram of intraoperative finding

and laparoscopy revealed the findings of functional bilateral uterine horns which were far apart in iliac fossa on both sides along with complete cervico-vaginal agenesis (U5aC4V4). So, the options of cervico-vaginoplasty and metroplasty were unrealistic in our case. The decision of hysterectomy was taken, and the parents were counseled regarding the need of vaginoplasty at a later stage at least 3 months prior to her marriage. Also, to the best of our knowledge, the skeletal deformity present in this case is rare and has not been reported in the literature earlier.

On reviewing the literature, we could find very few cases of bicornuate uterus with cervico-vaginal agenesis. Mostly, the approach in these cases was of hysterectomy followed by vaginoplasty. A similar case has been reported in 2014 where a 15-year-old girl had unicornuate uterus with nonfunctioning rudimentary horn with cervico-vaginal agenesis with rectovaginal fistula (RVF) [4]. She underwent total abdominal hysterectomy with RVF repair. Another case of a 15-year-old girl with pelvic pain and primary amenorrhea was found to have didelphys uterus with complete cervical and vaginal agenesis. She was

managed with laparotomy and excision of horns followed by the creation of neovagina later on [5]. In our case, we could manage with minimally invasive surgery and laparoscopic excision of bilateral horns was performed.

Though there have been case reports where utero-vaginoplasty has been done, results described are generally with short-term benefits [6]. It can be done either in the same sitting with vaginoplasty or in the second sitting. Malecot's catheter, Pigtail catheter or even Foley's catheter can be placed through the anastomosis over which fibrosis and epithelialization occur resulting in neocervix formation. This procedure retains the uterus, and hence, there is the possibility of pregnancy either naturally or through assisted reproductive techniques. Very few cases of pregnancies have been reported in cases of cervico-vaginal agenesis. Acien et al. [7] in a case report described the successful pregnancy outcome after McIndoe's vaginoplasty, cervico-isthmus resection and implantation of uterine corpus in neovagina. However, in our case it was not feasible due to widely separated horns.

Conclusion

Bicornuate uterus with cervico-vaginal agenesis is a rare entity, and it poses challenge for the gynecologists as the management varies with each patient. Thorough evaluation is needed in such cases as it can be associated with renal or skeletal malformations. Traditionally, the treatment has remained hysterectomy, but it carries great psychosocial impact on the adolescent girl and her family. Therefore, uterine-preserving surgeries in the form of metroplasty and

utero-vaginal anastomosis followed by vaginoplasty are one of the options, but the results are not very promising. So, individualized approach is required after the proper counseling of the family.

Compliance with Ethical Standards

Conflict of interest The authors report no conflict of interest.

Informed Consent Informed written consent was obtained from the patient.

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