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Case Report

Hematometra Secondary to Cervical Agenesis with Didelphys Uterus with No Communicating: A Case Report

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Abstract

Introduction: Mullerian agenesis a rare uterine anomaly accounts about 3% of uterine anomaly. The prevalence of congenital cervical agenesis or dysgenesis ranges from 1/80,000 to 1/100,000. Previously, the mainstay of treatment for congenital cervical malformations was hysterectomy. Currently through improving surgical technical and growing experience in the field of reconstructive techniques, conservative surgical management has become an option. We reported a case of complete cervical agenesis in a 13-year-old girl who underwent a successfully utero-vaginal communication opened.

Case Report: A 13 years old nulligravida lady, who never saw her menses yet. She was referred from local primary Hospital with a diagnosis of Hematometra secondary to cervical agenesis for possible MRI and Urogynegyncology evaluation. Initially she was presented to that Hospital for abdominal swelling of 1-year duration then diagnosed to have hematocolpometra secondary to plus cystic abdominal mass, for this she was operated and huge abdominal cyst removed and hymenectomy done but still hematometra is not evacuated then referred on 42nd post-operative day. She also has history of abdominal swelling and pelvic fullness of 3 weeks duration after operation was done. She has cyclical lower abdominal pain of 3 months duration. On Physical examination, she is well looking with stable vital sign. She has 16 weeks abdominopelvic mass boggy non tender smooth surface, she well-formed external female external genital with tanner stage 3 pubertal development on speculum examination vagina is well formed with length 7 - 8 cm long no cervix visible. Transabdominal Ultrasound Index Hematometra secondary to? plus? Didelphys uterus plus congenital absence of right kidney. With final diagnosis of hematometra secondary to cervical agenesis plus? Didelphys uterus plus congenital absence of right kidney septum resection done.

Conclusion: Congenital cervical agenesis, is currently classified as type C4 congenital malformation according to the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE) classification system of female genital tract anomalies, previously categorized as type IB congenital anomalies according to the American Fertility Society/American Society for Reproductive Medicine (AFS/ASRM) classification. The main objectives of treatment were symptoms relief, Achievements are to relieve symptoms, regular menstruation and restoring fertility. As this case septum resected and she was relieved from symptom and started to saw her menses.

Keywords: *Cervical Agenesis; Didelphys Uterus*

Background

Mullerian anomalies appear to have a substantial impact on the reproductive potential of the affected women, and the obstructive forms may affect the woman's health. Mullerian anomaly prevalence to be between 4% and 6% of the general population. In many cases the treatment of these anomalies may necessitate complex surgical approaches of undetermined prognosis. Embryological mullerian ducts are differentiated, fused and canalized finally form fallopian tubes, uterus, cervix and upper vagina. These are fused with the uro-

genital sinus that forms the lower vagina. These processes start cranially and progress caudally. Arrest at any level leads to ''localized gynatresia". Cervical agenesis currently classified as type C4 Mullerian anomaly, according to the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE) classification system of female genital tract anomalies, previously categorized as type IB congenital anomalies according to the American Fertility Society/American Society for Reproductive Medicine (AFS/ASRM) classification. The prevalence of congenital cervical agenesis or dysgenesis ranges from 1/80,000 to 1/100,000 [1,2].

Cervical agenesis is obstructive anomaly if associated with functioning uterus, hematometra will occur [3,4]. It is estimated that only 4.8% of women with cervical agenesis have a functioning uterus [5]. Any pubertal female with primary amenorrhoea and a history of cyclic abdominal pain should raise the clinical suspicion of cervical malformations (with or without a normal vagina). These cases present with primary amenorrhea, well developed sexual characters and cyclic abdominal pain [6]. Hematocolpos secondary to imperforate hymen or a transverse vaginal septum should be differentiated.

The clinical examination should include inspection and palpation, ultrasound, and magnetic resonance imaging (MRI) to complete the diagnostic workup. Inspection of the genital area and the introitus can confirm the presence or absence of a vagina and might exclude hematocolpos.

In the diagnosis of female genital tract anomalies, 2D ultrasound has 67.3% sensitivity and 98.1% specificity [12] and 3D has 98.3% sensitivity and 99.4% specificity. MRI can correctly subclassify as many as 85.8% of the cases with female genital tract anomalies [7]. Surgical exploration sets the final diagnosis.

The main objectives of treatment were symptoms relief, achievement of regular menstruation and restoring fertility.

Several methods of reconstructive surgery have been developed to create an new canal to communicate utero vaginal canal [6]. To prevent closure of the surgically formed utero vaginal canal, it is recommended that to insert catheter stent is left for three to five weeks in utero vaginal canal formed [8-10]. Cyclic estrogen progestogen therapy such as combined contraceptive pills given postoperatively for 2 - 3 months promotes epithelialization of the surgically formed utero vaginal canal [8,9].

The conservative surgical treatment is simple technique might open the door for wider application of this therapeutic option with the possibility of restoration and maintenance of normal menstrual outflow tract, fertility preservation and also, improvement of the patient's psychic condition.

Case Presentation

A 13 years old nulligravida lady, who never saw her menses yet. She was referred from local primary Hospital with a diagnosis of Hematometra secondary to cervical agenesis for possible MRI and Urogynegyncologist evaluation. Initially she was presented to that Hospital for abdominal swelling of 1-year duration then diagnosed to have hematocolpometra secondary? to plus cystic abdominal mass, for that she was operated and huge abdominal cyst removed and hymenectomy done to evacuate hematometra but still hematometra is not evacuated then referred on 42nd post-operative day. She had history of abdominal swelling and pelvic fullness of 3 weeks duration after operation was done. She had cyclical lower abdominal pain of 3 months duration which was not relieved after operation.

She had no history of vaginal bleeding, no foul smelling vaginal discharge. She didn't start sexual intercourse. She had no history of cough, shortness of breath, orthopnea. No history of yellowish discoloration of eye.

On physical examination, she is well looking and with stable vital sign. On breast examination well-formed breast with tanner stage 4 Abdominal examination, there is 16 weeks sized abdominopelvic mass boggy smooth surface, non-tender. No sign of fluid collection or organomegaly. On Genitourinary system- no CVAT and SPAT. Per vaginal examination-there is well formed external genitalia with tannel

stage 4 pubic hair development vaginal length is 5 - 7 cm and no cervix palpable or visible during speculum examination. On transabdominal ultrasound- Partially full bladder, Uterus filled with hypoechogenic (fluid) measuring 7 x 8 cm with visible ovary on rt side (Figure 1) and there visible uterus with uterine slight on lt side (Figure 2), left kidney visible in place with echotexture and no visible rt kidney. No free fluid in the peritoneal cavity, paracolic gutters or culde sac. With above history, physical examination and ultrasound diagnosed - Hematometra secondary to cervical agenesis plus? Didelphys uterus plus congenital absence of rt kidney. She was prepared for laparotomy and vaginal approach and taken to Operation theater.

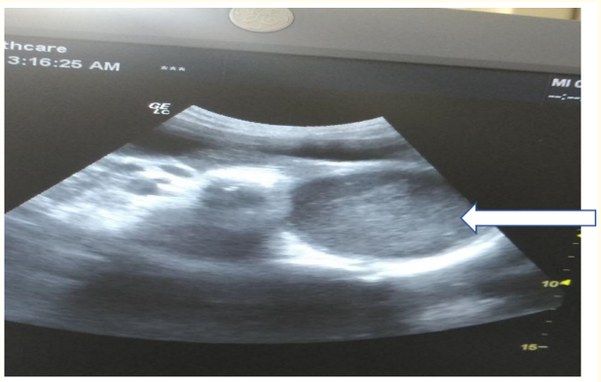

Figure 1: *Transabdominal ultrasound showing hematometra seen by arrow above.*

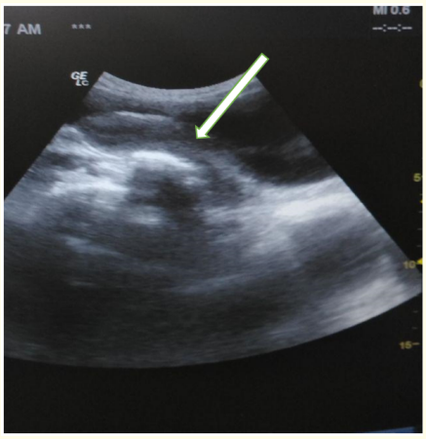

Figure 2: *Transabdominal ultrasound of another empty uterus with uterine slit. The arrow is uterus with endometrial slit.*

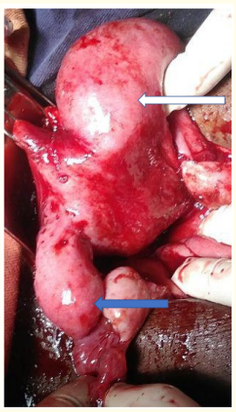

Figure 3: *Posterior view Intraoperative images of rt uterus with hematometra (white arrow), lt uterus (blue arrow).The arrow is uterus with endometrial slit.*

Written informed consent taken, patient prepared and transferred to OR and General anesthesia given, abdomen cleaned with povidone iodine, draped and entered through midline incision below umbilicus.

Intraop findings: There are two uterus with their tubes and their ovaries, Rt uterus is 16 week sized boggy uterus and lt uterus small, healthy looking with tube and ovary. There is no communication between them. There is health looking urinary bladder. Lt kidney is palpable but rt kidney is not palpable.

What was done: Vesical uterine peritoneum reflected down then about 5 cm longitudinal incision made over uterine isthmus then 500 ml hematometra evacuated then visualized no communication down to vagina. A third assistant insert speculum using sponge forceps push up upper vagina (thick septum), then using two allies septum was hold then incised between two allies. Now the uterine isthmus is directly opened to vagina. Uterine isthmus repaired to vagina. Foley catheter number 18 inserted to uterine isthmus and inflated with normal saline of 50 ml uterine isthmus repaired by two layer. Foley catheter stayed for 7 days, patient was smooth post operatively and discharged on 8th post op day. She was appointed after 1 month of operation then evaluated no complain and she visit her menses one cycle without any problem. She was counseled on the main surgery is to relieve symptoms and if she will be become pregnant advised to have prophylactic cerclage at 14 weeks.

Discussion

Cervical agenesis is a rare case in which the management is not easy because there are no many medical personnel exposed to this case [11]. So, this case report may give as referral for others.

The uterine cervix provides an outflow tract for the menstruation. Cervical agenesis hinders this function, it also maintains pregnancy during pregnancy affects the reproductive function and represents a therapeutic dilemma. Finally, if all treatment fail hysterectomy was the eventual treatment for cervical agenesis because of the common complications of recanalization of the cervix and the unlikelihood of a viable pregnancy [12]. Recent advances operation led to conservative surgery as the first line treatment [8].

The disorder is not life saving but it will make the patient feel uncomfortable, psychological problem and infertility. If it persists for a long time and not be managed properly, it may lead to complications such as kidney disorders and infertility problems [13].

Neocervix was made with the main objective to drain the menstrual blood and maintain sexual function. Placement of the largest intrauterine 24F-catheter is expected to prevent the portion newly created from closing or relapse.

Conclusion

The procedure promises to be a simple and reasonably effective method for the creation of a menstrual outflow tract, symptom relief and to become fertile for cases of cervical agenesis with functioning uterus. Treatment of cervical agenesis with minimally invasive therapy is strongly recommended for menstrual blood drainage and symptom relief and also for maintaining fertility in the future.

Consent

Written informed consent was obtained from the lady for publication of this case report and accompanying image.

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