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Case Report

Cryptomenorrhea due to Agenesis of Upper Part of Vagina Treated by Vaginoplasty with Amnion Graft and Cervicovaginal Anastomosis: a Case Report

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Abstract:

Cryptomenorrhea due to vaginal agenesis is a rare developmental anomaly of Mullerian duct. It may be complicated by retrograde menstruation, endometriosis and acute or chronic pelvic infection. We report a case of a 12-year-old girl who presented with a history of severe cyclical lower abdominal pain for 2 months. Her clinical examination revealed well developed secondary sexual characteristics, slightly bulged lower abdomen with a palpable tender mass and a short blind vagina. Ultrasound imaging showed hematometra, hematocervix and vaginal agenesis. She was treated by vaginoplasty with amnion graft and cervicovaginal anastomosis with serial vaginal dilatation using plastic vaginal mold. The patient was symptom free with regular menstruation on follow up visit at 1, 3 and 9 months. The Uterus was normal in size with empty cavity and well-defined endometrial lining on ultrasound.

Key words: Cryptomenorrhea, Vaginal agenesis, Hematometra, Vaginoplasty, Amnion graft.

Introduction:

Cryptomenorrhea is a gynecological abnormality where menstruation occurs but is not visible due to outflow tract obstruction[1]. The most common cause of cryptomenorrhea is imperforate hymen which is a rare mullerian anomaly. But cryptomenorrhea due to vaginal agenesis is an extremely rare mullerian anomaly[2]. The clinical presentation is primary amenorrhea and cyclical lower abdominal pain which may be complicated by retrograde menstruation, endometriosis and acute or chronic pelvic infection. We present a case of a 12-year-old girl with severe lower abdominal pain because of upper vaginal agenesis resulting in cryptomenorrhea.

Case Report:

A 12-year-old adolescent girl presented with a history of severe cyclical abdominal pain for 2 months. She developed secondary sexual characteristics at around 11 years of age, but she did not attain menarche till date. Suddenly she developed monthly lower abdominal pain, which was initially severe, incapacitating, lasting for 3-4 days, then gradually subsided, and persisted as a dull aching pain for 2 months. With this complaint she consulted a gynecologist and was diagnosed as a case of cryptomenorrhea and referred to BSMMCH.

On examination, she had a normal female curvature and voice with well-developed secondary sexual characteristics (Tanner stage 4). Per abdominal examination revealed a soft, tender mass in the hypogastric region, about 18 weeks of pregnant uterus size and lower limit could not

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be reached. On per vaginal examination under general anesthesia, external genitalia looked normal; vagina was short, blind, and admitted tip of index finger only. Digital rectal examination revealed a soft mass felt through upper part of anterior rectal wall.

A Pelvic ultrasound was done which showed an enlarged uterus with thinning of wall, cavity containing collection of fluid and distended cervix. Vaginal slit could not be outlined. Both kidneys and ureters were normal. Her routine investigations such as CBC, urine analysis, renal function test, and liver function tests were normal.

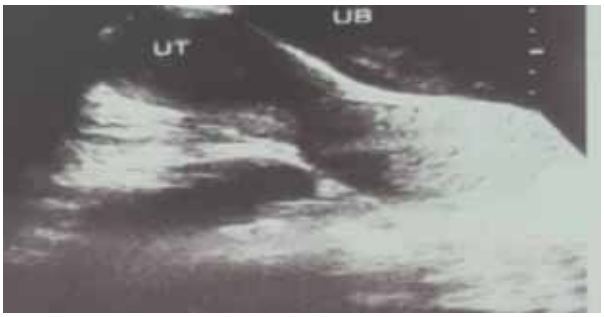

Figure 1: Ultrasound picture shows dilated uterus and cervix with menstrual blood and site of vaginal atresia.

Screening for HBsAg and HIV was normal. The patient and her guardians were counselled about the diagnosis and treatment options. It was planned for vaginoplasty with drainage of hematometra and hematocervix creating cervicovaginal anastomosis through abdomino-perineal approach after consultation with plastic surgeon. Pre-anesthetic checkup was done.

After giving general anesthesia, the patient was placed in lithotomy position. A transverse incision was given at middle part of the blind vagina to separate the bladder and rectum. Both sharp and blunt dissection were carried out to create neo-vagina. When about 6 cm of vaginal depth was created, the lower limit of hematocervix was reached, identified, and made an incision over there. The accumulated menstrual blood was coming out and allowed free drainage. Foley's catheter could not be introduced to maintain the patency of cervix as the opening was difficult to identify during drainage. Per vaginal discharge stopped after 7 days. The patient was re-evaluated by pelvic examination and ultrasound. On examination, cervical opening seemed to be stenosed. There was still a presence of small collection of fluid within the uterine cavity. Then the decision was taken to insert vaginal mold with amnion graft under general anesthesia. After 3 weeks of amnion grafting, the patient was discharged on request with advice for regular use of vaginal dilator. The third and final procedure was done two months after grafting. Abdomen was opened by lower transverse incision. A small incision was made on the lower segment of the uterus and collected blood was sucked out. Then a cervical opening was made at the level of external os through which a Foley's catheter was introduced per vagina and kept in situ. The patient was discharged with catheter in situ on 5th postoperative day without any complication. The patient was followed up after 1 and 3 months. She was symptom free and regularly menstruating. The Uterus was normal in size and endometrial cavity was empty. The catheter was removed after 3 months. Her next follow- up was after 9 months and 12 months of operation. She had regular menses with no complications.

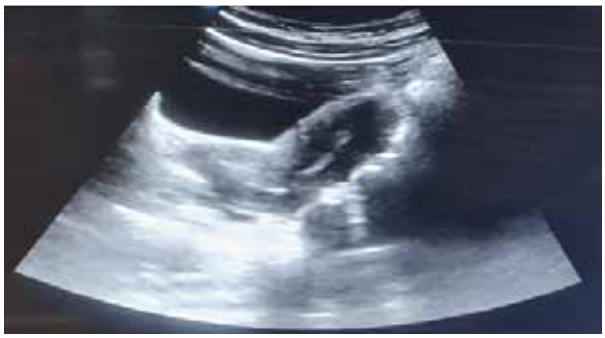

Figure 2: Postoperative ultrasound imaging shows normal size empty uterus at follow up visit.

Discussion:

Vaginal agenesis is a rare mullerian malformation. Usually occurs as a part of Mayer- Rokitansky-Kuster-Hauser syndrome where uterus and mullerian part of vagina is absent[3]. Vaginal agenesis is caused by underdevelopment of the mullerian duct where upper vagina is replaced by fibrous tissue. It is often present with urological and skeletal abnormalities[4].

Patients with vaginal agenesis have a normal female karyotype, functional ovaries and well developed secondary sexual characteristics. Primary amenorrhea and cyclical abdominal pain in adolescent girls are the common clinical features. Diagnosis should be suspected if there is absence of bulging from introitus. The diagnosis can be confirmed by ultrasound examination. Magnetic resonance imaging may be useful to diagnose associated anomalies[5].

This 12 years old girl had severe lower abdominal pain for short duration and primary amenorrhea despite well-developed secondary sexual characteristics. Diagnosis of cryptomenorrhea due to vaginal agenesis was made by presence of tender suprapubic mass with a small blind vagina and ultrasound imaging. Our case highlights that vaginal agenesis should be kept in differential diagnosis of cryptomenorrhea especially where there is no bulging from introitus.

Once the diagnosis of cryptomenorrhea is confirmed, the management is usually surgical. Surgical interventions for simple mullerian duct malformation such as imperforate hymen, transverse vaginal septum and cervical atresia have been performed without complications[6-9]. Creation of neo vagina for vaginal agenesis is a more complex operation which is associated with high morbidity and may require hysterectomy[10]. Several procedures have been successfully used for creation of neovagina such as McIndoe vaginoplasty. But it has the disadvantages of hair growth and numbness in vagina[11]. Amniotic membrane as a graft and vaginal mold has been successfully used in vaginoplasty[12]. After creating neovagina, uterovaginal anastomosis is done preferably in separate settings[13]. This principle was followed in the case described. Chakravarty et al. reported 18 adolescent girls with congenital cervicovaginal atresia who was treated with laparotomy followed by uterovaginal anastomosis and vaginoplasty with a plastic mold and fresh amnion graft. Two of the girls became pregnant spontaneously and delivered at term by cesarean section[14].

Conclusion:

Vaginal agenesis should be suspected in adolescent girls presenting with primary amenorrhea and cyclical lower abdominal pain. Management of vaginal agenesis is a challenge due to its complexity and fertility concern. We recommend abdominoperineal approach for vaginoplasty and uterovaginal anastomosis in the same setting to reduce repeated surgery and anesthesia related complications.

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