Case report:

Unilateral Functional Uterine Horn with Non Functioning Rudimentary Horn and Cervico-Vaginal Agenesis: Case Report

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ABSTRACT:

Developmental anomalies involving Mullerian ducts are one of the most fascinating disorders in Gynaecology. The incidence rates vary widely and have been described between 0.1-3.5% in the general population. We report a case of a fifteen year old girl who presented with primary amenorrhea and lower abdomen pain, with history of instrumentation about two months back. She was found to have abdominal lump of sixteen weeks size uterus. On examination vagina was found to be represented as a small blind pouch measuring 2-3cms in length. A rectovaginal fistula (2x2 cms) was also observed. Ultrasonography of abdomen revealed bulky uterus (size 11.2x6 cm) with 150 millilitre of collection. A diagnosis of hematometra with iatrogenic fistula was made. Vaginal drainage of hematometra was done which was followed by laparotomy. Peroperatively she was found to have a left side unicornuate uterus with right side small rudimentary horn. Left fallopian tube and ovary showed dense adhesions and multiple endometriotic implants. Both cervix and vagina were absent. Total abdominal hysterectomy was done and rectovaginal fistula repaired. The present case is reported due to its rarity as it involved both mullerian agenesis with cervical and vaginal agenesis along with disorder of lateral fusion. This is an asymmetric type of mullerian duct development in which arrest has occurred in different stages of development on two sides.

Key Words: Mullerian agenesis, unicornuate uterus, rectovaginal fistula, cervical agenesis, Vaginal agenesis

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Introduction:

Developmental anomalies involving Mullerian ducts are one of the most fascinating disorders in Obstetrics and Gynaecology. The incidence rates vary widely and have been described between 0.1-3.5% in the general population[1,2,3]. Class I of American Fertility Society[4] as well as Buttram and Gibbons[5] classification involves agenesis or hypoplasia of vagina, cervix, fundus or tubes or any combination. Ib involves agenesis of cervix, which is a rare anomaly. It results from failure of mullerian duct canalization or increased local epithelial proliferation after canalization. A functioning uterus with cervical agenesis leads to formation of hematometra and endometriosis and chronic pelvic pain. Disorders of lateral fusion are included in Class II-III. Presence of rudimentary horn with well developed one side horn is Class II anomaly but is quite common.

Case History:

A fifteen year old girl presented in Gynaecology Out-patient department with complaints of primary amenorrhea, pain in lower abdomen along with progressive abdominal swelling for the last one year. She had periodic exacerbations in the intensity of pain and was treated with anti-spasmodics during the episodes. There was history of some instrumentation about two months back at a local private hospital, the details of which were not available.

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She had normal secondary sexual characteristics; normal breast development and, pubic and axillary hair distribution. On abdominal examination, lump size found to be measure sixteen weeks fundal height of pregnancy which was firm, mobile and non tender. On local examination a blind pouch of vagina approximately 2-3cms in length was observed. Per-rectal examination revealed a tense, firm, tender lump anteriorly. A rectovaginal fistula, approximately 2x2cms in measurement, was also observed. Ultrasonographic examination revealed hematometra with a bulky uterus of size 11.2 x 6cm, with 150cc of collection. Sonologically both ovaries appeared to be normal. A diagnosis of primary amenorrhea with hematometra with iatrogenic rectovaginal fistula was made.

Vaginal drainage of hematometra was done and the procedure followed by laparotomy was performed. On laparotomy, a left sided unicornuate uterus with right side small rudimentary (1 cm approx) was found. Both horns were connected by a fibrous band (1 cm approx). Right fallopian tube and ovary was normal. Left fallopian tube and ovary showed dense adhesions, with multiple endometriotic implants on ovary. The ovary itself was normal in size. Both cervix as well as upper vagina was found to be absent. Abdominal hysterectomy was performed preserving the both ovaries. Repair of rectovaginal fistula was performed simultaneously. Post operative recovery was uneventful. On follow up, the patient was symptomatically relieved and doing well.

Discussion:

The normal development of female reproductive tract involves a series of highly orchestrated and complex interactions which direct the differentiation of Mullerian duct and urogenital sinus. A vast array of structural anomalies may result from interruption of mullerian duct development.

Cervical and vaginal agenesis is characterized by the absence or hypoplasia of the uterus and proximal vagina. This anomaly is termed as Mullerian aplasia and is classified in Class I of American Fertility Society (AFS) classification. Incidence of Class I Mullerian aplasia reported to be 1 in 5000. Congenital cervical agenesis, Class Ib, is a rare anomaly; only 58 cases have been reported till now. Fujimoto et al[6] reported 7 cases and reviewed 51 previously reported cases. They have concluded that half of the cases with cervical absence or atresia had normal vagina while half had complete of partial atresia. Combined vaginal and cervical agenesis is an extremely rare anomaly[7].

The management of cervical agenesis with functional uterus is difficult as surgical repair has not been found to be generally successful for such cases. Hysterectomy with preservation of ovary is the only answer for such patients. There have been case reports where attempts were made to re-construct the cervical canal. However, short term benefits have also been observed, with temporary restoration of menstruation. Even one pregnancy has been reported[8]. However, this iatrogenic endometrial-vaginal fistula is a risk favouring ascending infections; even a few deaths due to sepsis have been reported[9]. Besides, conception is considered unlikely due to absence of cervical mucus. Therefore, the conservative approach in the management of such cases to restore fertility had little success [10-11].

The present case is difficult to classify according to AFS and Buttram and Gibbons classification because it involves both mullerian agenesis with cervical and vaginal agenesis along with disorder of lateral fusion. This is an asymmetric type of mullerian duct development in which arrest has occurred in different stages of development on two sides. Goluda et al[12] reported a case of primary amenorrhea which had bicornuate rudimentary uterine functional horn with complete cervical vaginal agenesis coexisting with ovarian endometriosis. They have performed resection of rudimentary functioning horn with bilateral salpingo opherectomy.

Conclusion:

Mullerian anomalies are comparatively rare entities. The diagnosis as well as management of such cases remains a challenge to the gynaecologists due to the varied combinations and presentations. The present case is unique, as cervical agenesis per se is an extremely rare entity, and, to the best of our knowledge, the combination with vaginal agenesis and unicornuate uterus with a functioning endometrium as well as a rudimentary non functional horn and an iatrogenic rectovaginal fistula has never been reported in the literature.

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