CONGENITAL CERVICO-VAGINAL ATRESIA – A CASE REPORT

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ABSTRACT

Congenital cervico- vaginal atresia with a functional uterus, resulting in retrograde menstruation, leading to haematometra and in late cases haematosalpinx and pelvic endometriosis is a rare anomaly of the mullerian system. The main aim of management in a young patient with no severe haematometra, minimal pelvic endometriosis and adhesions is uterovaginal anastomosis / canalization to restore normal menstruation and fertility through assisted reproductive techniques. But in patients, whose chances of fertility would be markedly compromised by the presence of pelvic endometriosis, adhesions, haematometra, ovarian endometriotic cysts or advanced age, primary hysterectomy rather than conservative management should be considered.

Key words: *Congenital Cervical Atresia, Haematometra*

INTRODUCTION

Congenital cervical atresia with or without vaginal agenesis, but with functional uterus is a rare anomaly of the mullerian system (1,2). The presence of functional endometrium and functional ovaries may cause retrograde menstruation, leading to haematometra and in late cases haematosalpinx and pelvic endometriosis. The most common clinical presentation occurs after menarche in the form of primary amenorrhea, cyclic abdominal pain and abdominopelvic masses. Early diagnosis and reconstructive surgery is necessary to relieve the symptom related to haematometra and retrograde menstruation and to restore regular menstruation and fertility (1, 2). Many reconstruction procedures have been performed to relieve the symptoms, but eventually hysterectomy may be required in some complicated cases.

CASE REPORT

In January 2006, a 24 year old, unmarried female of low socioeconomic status presented to the gynae OPD of our hospital with complaints of primary amenorrhea and cyclic lower abdominal pain for 10 years. Since 2-3years she also felt a lump in lower abdomen. Tracing back her past history, there was a normal onset of thelarche and pubarche at 12 years of age. She had sought care at various medical centers over the past 8-10 years. A pelvic sonography was done at 14 yrs of age which showed a bulky uterus with haematometra and normal adnexae. She received symptomatic medical treatment by many doctors (analgesics, antibiotics and long courses of oral contraceptives). A repeat sonography at 18 yrs of age revealed an enlarged uterus (97mm x 66mm x 60 mm) with haematometra with right sided ovarian cyst (85mm x 76mm x 66 mm). She was advised surgical treatment by various doctors, but did not get any surgery done due to financial constraints.

Physical examination revealed a healthy looking girl, 164 cm tall and weighing 42 kg. The pubic and axillary hair was normal. Secondary sex characters were well developed. Her vital signs were normal. Abdominal examination revealed enlarged uterus (14 weeks pregnant uterus size) and a lump in left iliac fossa (4 x 6 cm), which was non-mobile, mildly tender. Pelvic examination revealed normal-appearing external genitalia but only a small shallow dimple at the vaginal introital site (2-3 cm). There was no evidence of a bulge or haematocolpos. On per rectal examination, a firm pelvic mass was felt 2-3 cm above the anal orifice through the anterior wall of rectum. Pelvic sonography showed a large fluid-filled uterus (16 cm in longitudinal diameter), consistent with a haematometra. No obvious uterine cervix was seen. Fibrous tissue was seen in upper part of vagina. There was a mass in right adnexa (8x6cm) that appeared to be contiguous with the uterus (haematosalpinx or tuboovarian mass). Left ovary was seen normally. No anomaly in urinary system or any other system was seen. Her haemogram, liver and renal function tests were normal. Serum concentrations of LH, FSH, oestradiol, prolactin and TSH were all within normal limits. Therefore, a provisional diagnosis of cervical and vaginal atresia associated with haematometra and right tubo-ovarian mass was made and laparotomy decided.

At laparotomy, dense adhesions were found between omentum, small and large intestines, uterus, bilateral tubes and ovaries forming frozen pelvis. All adhesions were separated carefully. Uterus was 14-16 weeks size with haematometra with right sided haematosalpinx with endometriotic cysts in right ovary. Left ovary was normal. Hysterectomy with right salpingo-oophorectomy was done. The post –

operative period was un-eventful. Gross pathological examination of the specimen showed uterus with blind cervix. On cut section, uterine cavity was filled with old clotted blood, cervix was solid, cervical os and cervical canal could not be visualized. Cut section of tubo ovarian mass showed cystic and hemorrhagic areas in the ovary and tube.

Microscopic Examination revealed functional endometrium of non-secretory type. Sections from cervical area revealed no epithelium, no cervical stroma and no cervical glands. The normal ovarian tissue was almost replaced by haemorrhage and cystic areas with occasional primordial follicle.

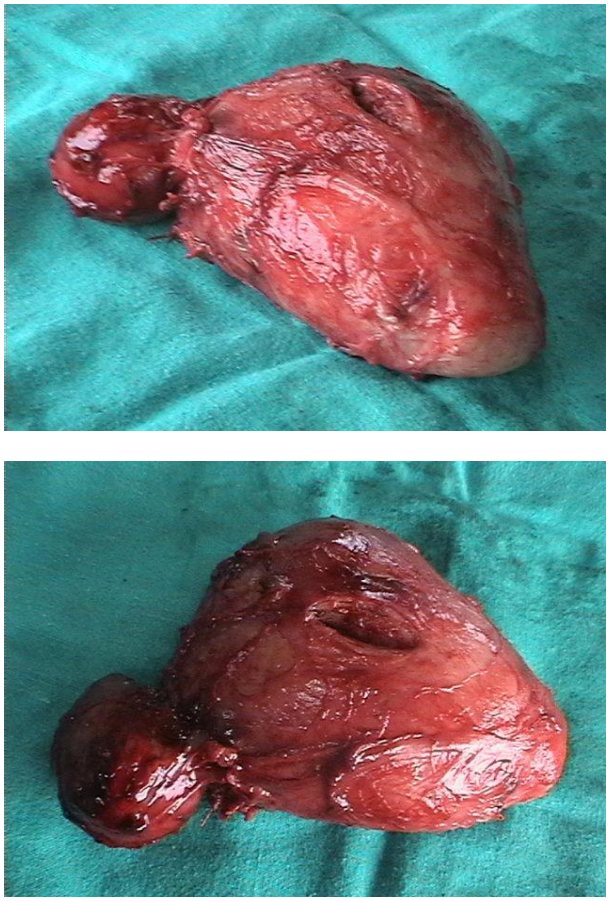

Fig. 1- Gross photograph of the uterus with cervical agenesis.

DISCUSSION

Congenital atresia of the uterine cervix with absence of cervical stroma and endocervical glands, is a very uncommon class 1B mullerian malformation associated in 50% of the cases with a vaginal aplasia (3,4). Any abdominal or pelvic, acute or chronic pain, in a pubescent girl must evoke suspicion of an obstructive genital syndrome. Transabdominal or transperineal sonography may specify the level of the obstacle, but are not very reliable for the diagnosis of uterine cervical atresia. MRI appears to be the most reliable diagnostic tool for uterovaginal malformations (3, 4).

Unlike most other mullerian anomalies, the initial surgical management of congenital cervical atresia remains controversial. The main aim of management is uterovaginal anastomosis / canalization to restore normal menstruation and fertility. Because of post-operative complications of uterovaginal canalization including intraabdominal infection, recurrent obstruction of uterovaginal neocanal due to canal stricture or stenosis and persistent infertility, a majority of clinicians view hysterectomy as the optimal primary surgical management in these patients (3, 4). Lack of endocervical glandular function and epithelium may be one of the most important contributing factors resulting in stenosis and infertility (1, 2, 3).

The majority of these patients have evidence of either pelvic endometriosis or pelvic adhesive disease or both at the time of laparoscopy/ laparotomy. The degree of pelvic endometriosis and adhesions present needs to be considered in recommending a surgical approach to the problem. It must be emphasized that patients, whose chances of fertility would be markedly compromised by the presence of pelvic endometriosis, pelvic adhesions, salpingitis, ovarian endometriotic cysts or advanced maternal age should be advised to consider primary hysterectomy rather than conservative management (3).

This case was unique in its late presentation with haematometra, haematosalpinx, endometriotic ovarian cysts and dense pelvic adhesions and chances of fertility appeared almost nil. Hence, hysterectomy and right sided salpingoo-oopherectomy was decided as the primary treatment to relieve her from her symptoms. Absence of cervical stroma and glands was substantiated by pathological examination.

The ideal candidate for canalization would be the patient of young age with no severe haematometra, minimal pelvic endometriosis and adhesions at the time of surgery. Amniotic membrane appears to be a good alternative endogenous material for epithelisation of neocervix in uterovaginal canalization procedures (2). It has been suggested that successful reconstruction is likely only in the presence of cervical stroma. Further, recent advancements in assisted reproductive techniques may afford patients, who successfully undergo canalization procedure, a better opportunity to achieve pregnancy in the future (1, 2, 3, 4).

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