Case Report

Cervical Aplasia

OBS & GYN Section

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ABSTRACT

The incidence of genital malformations in the general population is 3-5%. Ours was a case of cervical aplasia. Our objective was to confirm the diagnosis and to manage the same surgically. It is essential that these patients receive psychological support as it has implications on their reproductive life. This case was referred to our tertiary care teaching hospital. After investigations and examination under anaesthesia, the patient underwent hysterectomy. Obstructive anomalies of the genital tract are the commonest among the mullerian duct malformations.

Key Words: Cervical aplasia, Mullerian malformation

INTRODUCTION

Müllerian agenesis is a congenital malformation in women which is characterized by a failure of the müllerian ducts to develop, resulting in an absent uterus, fallopian tubes and variable malformations of the upper portion of the vagina. It is the second most common cause of primary amenorrhoea after gonadal failure. Various modalities of treatment have been adopted, including anatomical reconstruction surgeries, IVF or embryo transfer to a gestational carrier.

CASE REPORT

A 30 year old nulligravida presented with primary amenorrhoea and cyclical abdominal pain for the past two years, which increased since three months. At the age of seventeen, she was investigated for primary amenorrhoea and was diagnosed to have a hypoplastic uterus with lower genital tract obstruction. She underwent a surgery for resection of the transverse vaginal septum, but continued to be amemorrhoeic. She got married a year later and the couple had no coital problems.

On examination, She was well built and had normal secondary sexual characters normal secondary sexual characters. Her thyroid and breast were normal. Her per abdomen examination revealed a suprapubic mass which arose from the pelvis, which corresponding to a 20 week sized gravid uterus. There was no other mass or free fluid in the abdomen. Her external genitalia appeared to be normal. Her vagina admitted two fingers and ended blindly and the cervix was not felt. A transverse septum was felt at the upper part of the vagina.

Her ultrasound scan revealed a large haematometra and her cervix was not visualized. The right fallopian tube was grossly distended and the right ovary was cystic. The left ovary was normal. Both her kidneys were normal.

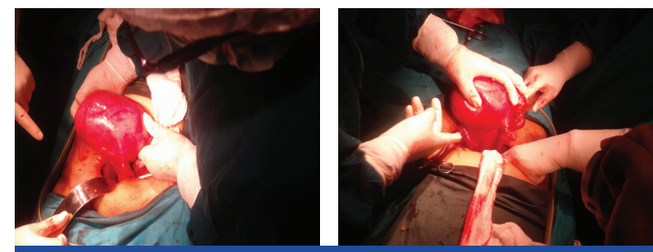
Hb was 12gm % and creatinine was 0.8. The patient was posted for EUA (examination under anaesthesia) and the per vaginal findings were confirmed. She underwent laparotomy which showed these per operative findings: the uterus was soft and cystic, which corresponded to 20 weeks of gestation. The lower part of the uterus ended as a dense fibrous band which was adherent to the apex of the vagina [Table/Fig-1]. The band was probed and was found to be non-canalized. The right tube showed haematosalphinx and the right ovary showed multiple cysts [Table/Fig-2]. The left adnexa was normal. The rest of the pelvis showed no evidence of endometriosis. In view of the above findings, it was decided to proceed with hysterectomy and right salphingoophorectomy. The uterus was delivered out and the vault was closed.

The histopathology report of the uterus showed features which were consistent with a haematometra. The dense fibrous band showed only muscle fibres and connective tissue, with no lining epithelium. The right tube lumen showed haemorrhage and pigment laden macrophages. The right ovary showed multiple haemorrhagic cysts. The post-operative recovery period was uneventful.

DISCUSSION

In the general female population, genital malformations occur with an incidence of 1%–5%, in groups of patients with infertility up to 6.5% [1-5]. The aetiology is possibly due to an aberrant developmental inhibition of the Mullerian ducts during embryogenesis. Various genetic factors have been investigated, but there is no conclusive evidence about the aetiology of any type of malformation [6-8]. The Vagina, Cervix, Uterus, Adnexa and Associated Malformation (VCUAM) classification which was designed for easy clinical use, makes it possible to record all the anatomic malformations and associated abnormalities separately. This is important particularly for the description of syndromes with genital involvement [9].

The congenital absence of the cervix is relatively infrequent and it occurs in 1 in 80,000 to 100,000 births. It is associated with partial or complete vaginal aplasia and renal anomalies [10]. Agenesis of the cervix presents a diagnostic challenge to the gynaecologist. Ultrasound and MRI (Magnetic Resonance Imaging) with pelvic



[Table/Fig-1 and 2]: Per-Operative

examination under anaesthesia help in confirming the diagnosis. An early diagnosis offers effective presurgical planning and preparation. Patient counselling should be initiated before the treatment.

Two basic categories of cervix anomalies have been observed.

1. The first type is cervical aplasia ie. lack of the uterine cervix , with the lower uterine segment which narrows to terminate in a peritoneal sleeve at the vaginal apex.

2. The second type of cervix dysgenesis is where the endocervical glands are seen on pathological examination. It has four subtypes

Many authors recommended hysterectomy for cervical agenesis with a functioning uterine corpus, to eliminate problems such as sepsis, cryptomennorrhoea, endometriosis and multiple surgeries [11,12].

With the recent advances in the reproductive medicine and laparoscopic surgical techniques, conservative surgery is a possibility and perhaps should be considered as the first-line treatment option [13]. Peritoneal grafts for the reconstruction of the cervix was reported by Alborzi S [14]. The canalization procedures are worthwhile in cases with adequate stroma to allow cervicovaginal anastamosis [15,16]. With the advances in reproductive medicine, pregnancy with IVF (in vitro fertilization) and embryo transfer is possible. However, the management modalities can vary from one institution to another, but we concluded that the treatment of choice should be hysterectomy, coupled with a psychological component.

The development of extreme anxiety about the patient's femininity and a distortion of her physical image can affect her self-esteem. The psychological adjustment and general attitude are also very important in deciding as to what procedure should be used and when it should be done as per the updated protocol by Lindenman. E et al [17].

Ultimately, the best surgical approach requires a discussion on the post-operative complications, the degree of cervical abnormality and the patient's desired treatment outcome. Whether the patient desires definitive treatment with a hysterectomy or whether she wants to pursue a patent outflow tract and the possibility of future childbearing when she is anatomically appropriate, evidence-based medicine must become the source for the surgical strategies [16].

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