Congenital Atresia of Uterine Cervix - A Rare Case Report

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PRESENTATION OF CASE

A 22-year-old woman, single, came to Radiodiagnosis Department of North Bengal Medical College and Hospital for ultrasonography examination of whole abdomen for evaluation of amenorrhea and vague cyclical lower abdominal pain. She had been treated outside the hospital for several years for above symptoms without any fruitful outcome. There was no history of any surgical management to this patient. Careful clinical examination of pelvis revealed an imperforate hymen. No other clinical signs were found except mild lower abdomen tenderness. Routine ultrasound was done with curvilinear probe with frequency of 5 MHz in GE LOGIQ P 9 model ultrasound sonography (USG) Machine. Cervical agenesis was suspected based on sonographic findings, non-visualization of the cervix with a uterus like structure (measuring approx. 30 x 36 x 30 mm.) in right adnexal region. Mild collection seen in pouch of Douglas. Both ovaries and bilateral adnexa were normal. Vagina showed no abnormal collection. Other abdominal organs like liver, gallbladder (GB), common bile duct (CBD), portal vein (PV), pancreas, spleen, both kidneys, and bladder appeared normal. Transvaginal examination as well as transvaginal sonography could not be performed as imperforate hymen. Therefore, the patient underwent magnetic resonance imaging (MRI) examination of whole abdomen which confirmed the ultrasonographic findings and the case diagnosed as congenital atresia of uterine cervix with imperforate hymen.

CLINICAL DIAGNOSIS

Main sonological findings of cervical atresia is non-visualization of uterine cervix in pelvis preferably in its normal anatomical position. Clinical examination helps in identifying lower genital tract anomalies like imperforate hymen or blind vaginal pouch but the distinction between cervical atresia and a high vaginal transverse septum is not possible.¹

In our case, USG revealed non-visualization of uterine cervix without any collection within vagina and uterus like structure in right adnexal region. Mild collection seen in pouch of Douglas. Bilateral ovary and adnexal region as well as other abdominal organs were normal. MRI was done to confirm these findings and there was no diagnostic discrepancy between ultrasonography and magnetic resonance imaging. Both the modalities diagnosed this case accurately.

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Primary amenorrhoea can be a presenting symptom for a broad spectrum of congenital uterine anomalies ranging from hypoplastic uterus to imperforate hymen. USG is the modality of choice to define the internal genital anatomy and help us to classify the level of obstruction or aplasia. In patients with cervical aplasia, may have upper vaginal pouch with atretic lower third of vagina, similarly some patients of under-developed cervix may have vaginal pouch with significant atretic segment of vagina. If the cervical dysgenesis occurs, there should not be any abnormal intravaginal accumulations of fluid or blood whereas in case of transverse vaginal septum or imperforate hymen with intact cervix, haematocolpos or hydrocolpos may develop. Ultrasonography, magnetic resonance imaging with detailed clinical pelvic examination can exclude this.

**DIFFERENTIAL DIAGNOSIS**

Past and present articles are descriptive and difficult to assess and compare due to difference in sample size and variation in surgical techniques. Grimbizis et al. published the success of end to end cervico-cervical anastomosis in 116 cases of transverse cervical defect whereas Rober et al. have outlined the importance of cervical anatomy. Although conservative surgeries with fruitful outcome reported, complications (especially obstructive) have not been fully documented. If vaginal atresia involved simultaneously, more difficulties arise to make a fistulous tract. Post-operative complications like infective, inflammatory, mechanical injuries to various organs, re-obstruction etc. have been reported. However, cervical reconstruction surgeries have been reported with successful outcome specially in obstructive type, but it has challenges. The goals of reconstructive surgery are to provide a conduit for menstruation, to relieve pain and to preserve reproductive potential. Patients with atresia or cervical fragmentation are usually poor candidates for canalization and total hysterectomy is the treatment of choice. Patients with either cervical obstruction or a fibrous cord may reasonably be considered for reconstruction. Controversies do exist in the treatment options and some authors describe uterovaginal anastomosis as the first line of management. However, evaluation is needed pre-operatively about the remaining part of cervix, so that risks and benefits of any procedure can be assessed.

**PATHOLOGICAL DISCUSSION**

Congenital cervical atresia is a rare clinical entity that was first reported by Ludwig in 1900. It is associated with acute or chronic abdominal or pelvic pain and reproductive problems. There is lack of uniformity in the literature regarding its classification and management. According to American Fertility Society, it is classified as type IB mullerian anomalies. Cervical atresia has been further classified into different types as follows: (i) The cervical body is intact with obstruction of the cervical os (ii) The cervical body consists of a fibrous band (iii) Fragmented portions of the cervix are noted (iv) The mid portion of the cervix is hypoplastic with a bulbous tip. Embryologically, female reproductive tract develops from paired mullerian ducts. Its complete formation and differentiation depends upon three phases of development-organogenesis, fusion (lateral as well as vertical) and seaptal resorption. Cervical atresia is considered as defect in the elongation of mullerian duct. At about 20 weeks of gestation, cervix is formed as a condensation of stromal cell at a specific site around the fused mullerian duct and its differentiation is a complex process which involves both mesodermal and endodermal tissue.

**DISCUSSION OF MANAGEMENT**

Based on non-visualization of uterine cervix without any other urogenital anomalies through careful clinical pelvic examination, ultrasonography as well as magnetic resonance imaging, the case was diagnosed as congenital atresia of uterine cervix with imperforate hymen.

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