



Case Report

Cervical and vaginal agenesis – A rare case report

Vijayalakshmi S, Mahendra G^{*}, Naga Prathyusha, Ravindra Pukale, Sekhar Durgam, Subrahmani

Department of Obstetrics and Gynaecology, Adichunchanagiri Institute of Medical Sciences, B.G. Nagara, Karnataka, India

*Corresponding author email: drmuba@gmail.com

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Abstract

Cervical agenesis is a very rare condition often associated with atresia of vagina. Clinical diagnosis is usually difficult before surgery. Transverse vaginal septum or vaginal agenesis is also a rare condition that results from incomplete fusion between vaginal components of the mullerian ducts and urogenital sinus. Here we presented case of 16 years old girl who presented with primary amenorrhea and cyclical lower abdominal pain for 6 months. Abdomino-pelvic scan showed hematometra with bilateral hematosalpinx with mild free fluid in pouch of Douglas with collapsed vagina and cervix. MRI pelvis confirmed USG findings giving differential diagnosis of high vaginal septum or cervical agenesis. She was operated for cervical recanalization and vaginoplasty.

Key words

Primary amenorrhea, Cervical agenesis, Vaginal agenesis, Vaginoplasty.

Introduction

Cervical agenesis is a very rare condition often associated with atresia of vagina. Clinical diagnosis is usually difficult before surgery. Transverse vaginal septum or vaginal agenesis is also a rare condition that results from incomplete fusion between vaginal components of the mullerian ducts and urogenital sinus.

Clinical presentation depends on whether it's partial or complete. With complete agenesis, menstrual blood accumulates and distends structures above it after puberty, resulting in hematoocolpos and hematometra. Such patients usually present with cyclical lower abdominal pain with primary amenorrhea and ultrasonographic findings of hematometra and



hematosalpinx. Occasionally a lower abdominal mass (hematometra) is palpable.

Case report

A 16 years old girl presented with primary amenorrhea and cyclical lower abdominal pain for 6 months. Patient was referred from Surgery Department after appendicectomy, as there was no relief of symptoms of pain in abdomen to Obstetrics and Gynecology Department. She had normal feminine features and general physical examination was normal. Her breasts were Tanner stage 3 with no masses. Her external genitalia were normal, on separation of labia majora, it appeared like imperforate hymen. On per rectal examination, vagina found to be absent and a tender globular firm smooth mass was noted. Abdomino-pelvic scan showed hematometra with bilateral hematosalpinx with mild free fluid in pouch of Douglas with collapsed vagina and cervix. MRI pelvis confirmed USG findings giving differential diagnosis of high vaginal septum or cervical agenesis. (Photo – 1, Photo - 2)

Photo – 1: Ultrasound showing hematometra.



Patient was posted for examination under anesthesia and diagnostic laparoscopy. Examination under anaesthesia showed imperforate hymen with absence of dimpling suggestive of vaginal agenesis. (Photo – 3) On

laparoscopy, uterus appeared bulky with bilateral hematosalpinx, minimal collection in pouch of Douglas, with endometriotic spots over the uterine surface, peritoneum, and pouch of Douglas. (Photo – 4, Photo - 5) Post operatively Inj. Depoprovera was given. 2 months later patient was posted for cervical recanalization and vaginoplasty.

Photo – 2: MRI pelvis showing absent cervix and vagina.



Photo – 3: Examination under anesthesia showing blind vagina.



Intra-operative findings

Hematometra with bilateral hematosalpinx was seen. Cervix was totally collapsed and seen as a thin fibrous band measuring 3.2 cm along with vaginal atresia of about 3 cm. Transverse incision was given over anterior surface of

uterus and approximately 400 ml of old menstrual blood was drained. **(Photo – 6)** A probe passed through uterine incision towards cervix ended blindly. Hysterectomy was done retaining both ovaries. Vagina was atretic and dissection was done in the loose areolar tissue or space between the bladder and rectum towards the cervical band. A full thickness skin graft was taken from right thigh and was used for construction of neovagina of about 6 cm depth via Mc Indoe technique. **(Photo – 7)** Prosthesis was kept under aseptic precautions to maintain the depth of the neovagina. **(Photo – 8, Photo - 9)** Post operative period was normal. **(Photo – 10)** Antibiotics and prosthesis maintained for 9 days. Following this period, prosthesis was replaced. Patient was discharged from the hospital with the indication of placing and keeping the prosthesis in the neovagina. Neovagina developed with proper squamous epithelium with a depth of 6 cm.

Photo – 4: Laparoscopy shows hematometra and hematosalpinx.



Discussion

Cervical agenesis is a congenital disorder of the female genital system that manifests itself in the absence of a cervix, the connecting structure between the uterus and vagina. One Mild form of condition in which cervix is present but deformed and non functional are known

as cervical atresia or cervical dysgenesis. Patients with cervical agenesis typically present in early adolescence, around the time of menarche, with amenorrhea and cyclic pelvic pain caused by the obstruction of menstrual flow from the uterus [1].

Photo – 5: Laparoscopy showing endometriotic spots.

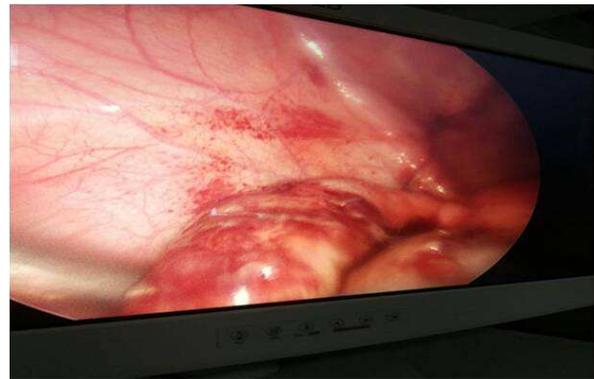


Photo – 6: Intraoperatively draining hematometra.



The diagnosis of cervical agenesis can be made by magnetic resonance imaging, which is used to determine the presence or absence of a cervix. Although MRI can detect the absence of a cervix (agenesis), it is unable to show cervical dysgenesis (where the cervix is present, but malformed). Ultrasound is a less reliable imaging study, but it is often the first choice by gynecologists to establish a diagnosis and can



identify a hematometra secondary to cervical agenesis [2, 3].

a vagina (vaginal agenesis), or a deformed and non functional vagina (vaginal atresia) [2, 3].

Photo – 7: Feeling for blind vagina and cervix.



Photo – 10: Post operative image showing neovagina.



Photo – 8: Prosthesis was used for creation of neovagina.



It is frequently associated with Mayer-Rokitansky-Küstner-Hauser (MRKH) syndrome, in which the most common result is an absent uterus in conjunction with a deformed or missing vagina, despite the presence of normal ovaries and normal external genitalia. It is also associated with cervical agenesis, in which the uterus is present but the uterine cervix is absent. Vaginal atresia is estimated to occur in 1 in 4000–5000 live female births. It is often unnoticed until adolescence, when pain and a lack of menstrual flow indicate the condition [4].

Photo – 9: Creation of neovagina.



Vaginoplasty is a reconstructive plastic surgery and cosmetic procedure for the vaginal canal and its mucous membrane, and of vulvo-vaginal structures that might be absent or damaged because of congenital disease (e.g. vaginal atresia) or because of an acquired cause (e.g. childbirth physical trauma, cancer) [1]. As such, the term vaginoplasty generally describes any such cosmetic reconstructive and corrective vaginal surgery, whilst the term neovaginoplasty specifically describes the procedures of either partial or total construction or reconstruction of the vulvo-vaginal complex [4, 5, 6].

Vaginal atresia is a birth defect or congenital abnormality of the female genitourinary system that manifests itself in the absence of



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