

Case Report: Transverse Vaginal Septum in A Down's Syndrome Patient

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ABSTRACT

Transverse vaginal septa are relatively rare anomalies. It has been shown to occur at different depths within the vagina. It results from vertical fusion or canalization disorder between the vaginal components of Müllerian ducts and urogenital sinus during embryological development of the vagina and can present with or without obstruction. The condition was initially described by Delaunay in 1877.

The authors describe a rare case of a middle-aged female with Down's syndrome, presented with secondary amenorrhea for 3 years. Initial examination and imaging investigation revealed the presence of a transverse vaginal septum. The diagnosis was facilitated using a combination of abdominal and transvaginal ultrasound scanning and CT. And it has been revealed that she has a high rising microperforate transverse vaginal septum. Surgery and follow up was also done.

Keywords: Transverse vaginal septum, Hypothyroidism, Down's syndrome.

INTRODUCTION

A vaginal septum results from incomplete fusion of the separating tissue between the fused Müllerian ducts and the urogenital sinus^[1]. This is estimated to occur to 1 per 30,000-84,000 women^[2]. It occurs in any portion of the vagina and can be transverse, longitudinal, or oblique.

Transverse vaginal septum is considered as one of the most rare female urogenital tract congenital abnormalities, and a low septum is even rarer. The etiology is unknown and is as a result of a female sex limited autosomal recessive transmission^[3]. And it is known as a fibrous membrane of connective tissue with vascular and muscular components, It can occur at almost any level of the vagina; superior, mid or inferior vagina.

The external genitalia appear normal, but the vagina is shortened and blocked resulting in cryptomenorrhea. Women with this condition may suffer pain during intercourse (dyspareunia).

Numbers are suitably vague, because it can go unnoticed if the septum doesn't trap menstrual blood behind it in a complete obstruction. Generally, these septa are not associated with other reproductive abnormalities but it may be combined with other Müllerian duct anomalies such as uterus didelphys. Clinical presentation depends on whether it is complete or partial.

Treatment for a transverse vaginal septum is generally surgery, where it can be removed. Postoperative complications, such as vaginal stenosis and reobstruction can occur, especially when the septum is thick.

CASE PRESENTATION

A 45-year-old woman, a known case of Down's syndrome who has normal secondary sexual characteristics, is nulliparous, obese, has bronchial asthma, and hypothyroidism for which she is taking L-thyroxine, she was referred from Rabigh Hospital, Saudi Arabia to Maternity and Children's Hospital, Jeddah. After complaining of secondary amenorrhea for 3 years and acute urinary retention for which a catheter was fixed. On per vaginal examination, it was found that she has a short vagina of 2 cm in length with a mass obstructing it that the fingers can't go around. No adnexal masses and no abnormalities were detected in pouch of Douglas.

Abdominal ultrasound showed a normal sized empty uterus about 55x30x38 mm, soft tissue and normal endometrium of 5 mm thickness with a cystic mass below it. There was collection of blood inside this cyst which measured 5.9 x 5 cm [Fig.1].

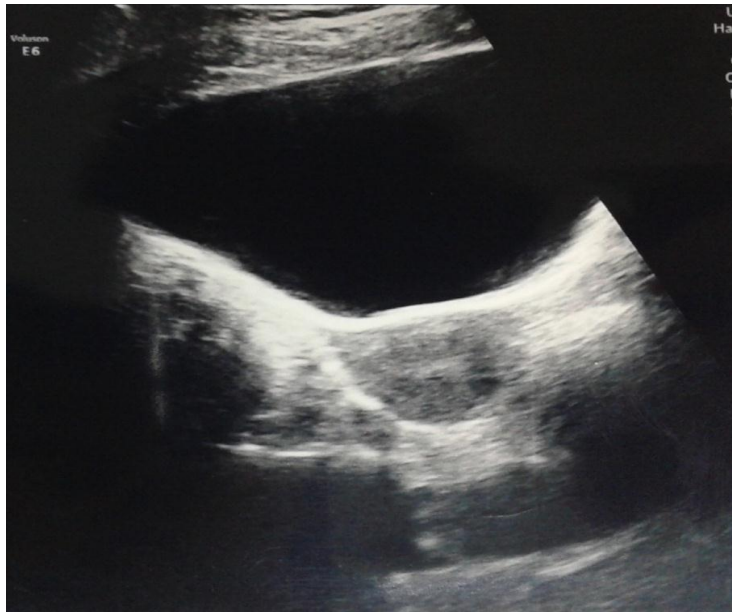


Fig.1 cystic mass measuring 5.9x5 cm

By transvaginal ultrasound, she had a transverse vaginal septum measuring 5.7x 4.7 cm. Later, CT showed a thick fluid collection below the uterus (in upper vagina) continuous with uterine cavity expanded by fluid.

She was prepared for surgical intervention and a transverse vaginal septum was excised and hematocolpos drainage was done.

In clinical follow-up, she complained again of amenorrhea and lower abdominal pain in the two months following surgery. Therefore, she was admitted for investigation and possible surgery. Abdominal ultrasound showed empty uterus measuring 46x24x26 mm and still blood in vagina measuring 31x 22 mm and the view of ovaries was suboptimal due to a bladder that wasn't full so we scheduled for another ultrasound appointment. Two days later U/S was repeated with full bladder and the following was found: Uterus was A/V. Size: 50x30x32 mm of a normal texture. Endometrium was normal and about 6mm thick, uterine cavity was empty. Nothing abnormal detected, Minimal bleeding in cavity near cervix, blood inside the vagina is less than previous time (2.8 x 2.6 cm). Left ovary was found to be normal in size and shape; measuring 18 x 14 mm. Right ovary was not seen due to lots of gases.

Seven days into admission, patient was prepared for surgery to resect the vaginal septum as much as possible.

Surgery was done under low spinal anesthesia in lithotomy position under aseptic technique. With U/S guidance, Foley's catheter was inserted.

Lower vagina was obstructed and scar tissue was dilated. Hematocolpos was drained through an

incision in transverse vaginal septum. The septum was excised as much as possible and endometrium sample was taken and sent for histopathology exam. Foley's catheter inserted into vagina with 30cc of normal saline and vagina was packed with 3 gauzes. Blood loss: 150 ml. Foley's catheter showed clean urine

In all 3 outpatient follow ups scheduled for her, clinical examination revealed a patent vagina with no adhesions or abnormal discharge.

RESULTS AND DISCUSSION

In all 3 outpatients follow ups scheduled for her, clinical examination revealed a patent vagina with no adhesions or abnormal discharge.

Transverse vaginal septum is a rare mullerian congenital malformation presenting with dysmenorrhea, dyspareunia and primary infertility in adult patient but it can present a diagnostic challenge, because symptoms are often not gynecologic in nature. Vaginal septa vary in location and thickness. The exact location is difficult to assess clinically unless we do vaginal and/or recto abdominal examination. 46% of vaginal septa are located in the upper third of the vagina, 40% in the middle third and 14% in the lower third ^[4]. The most common site of the septum is in the upper aspect of the vagina which can be confused with partial aplasia of the vagina ^[6]. The septum may be complete or may present with a small tract allowing escape of menstrual blood.

A transverse vaginal septum also presents management challenges. Treatment options vary from simple surgical excision ^[5] ^[6]. Z- Plasty

technique, various flaps and Hysteroscopy resection^[7]. First, the septum is examined vaginally and rectally to palpate the location of the bulge, it is commonly evaluated by trans-perineal ultrasound or MRI before attempting its resection, It is important to preoperatively assess transverse vaginal septum by asking about medical history and performance of physical examination. The family history of genetic abnormalities, presence of pelvic mass, and appearance of internal and external genitalia should be noted. C/T or MRI may be used for the diagnosis specially in cases with high vaginal transverse septum and can be helpful for planning surgical treatment^[8]. MRI is considered the criterion standard for imaging Müllerian duct anomalies and provides high resolution images of uterine body, fundus and internal structures. Ultrasound may be helpful to visualize a hematocolpos and the presence or absence of a uterus and/or septum. In our case, hematocolpos was suspected by ultrasonography. We decided to use an MRI to determine the etiology and select the type of treatment.

The exact surgical technique used depends on the thickness of the septum. Early accurate diagnosis should be followed by excision of the obstructing vaginal septum and relief of the symptoms.

The approach is dependent on the thickness of the septum; a thin septum (Less than 1 cm) should be resected as widely as possible to reduce the risk of postoperative stenosis, it can be repaired easily with an end-to-end anastomosis of vaginal mucosa^[5], especially if located low in the vagina. Using a vaginal mold or dilator after surgery can decrease the risk of vaginal stenosis^[9] and placement of a Foley catheter to drain the bladder is recommended because injuries to bladder and bowels are the most common intraoperative complications.

Thicker septa on the other hand, may require undermining the vaginal mucosa to allow the lower vagina to reach the upper vagina which is complicated postoperative by scar constriction and vaginal stenosis requiring further surgery or dilatation^[10]. In case of the patient being unable preoperatively to dilate the vagina, skin graft may be required to reanastomose the lower and upper vagina. In all cases, laparoscopy is performed to identify the endometriosis pathology due to the vaginal obstruction, and endometriosis is histologically confirmed. The use of a Foley's catheter appears to be a safe and effective method for resection of vaginal septa and with few complications.

CONCLUSION

High awareness is essential to make an early diagnosis. This case demonstrates the possibility of total occlusion of a perforate TVS likely resulting from septal scarring from gradually retained blood and tissue. Treating a case of transverse vaginal septum needs great expertise, as incomplete resection of a septum can put the patient at risk of repeated surgeries.

Complete resection of septum by identifying the extent of cicatrization and reunion of upper and lower vagina by overlapping sutures can out risk the patient from repeated failures. Z plasty technique or use of various flap techniques is the good alternative.

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