Primary Amenorrhoea with Transverse Vaginal Septum Presented with Pyometra of 24 Weeks Size

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INTRODUCTION

Transverse vaginal septum is a congenital anomaly of the female genital outflow tract. It most likely remains unrecognized till adolescence and manifests as primary amenorrhoea with cyclic lower abdominal pain in background of appropriate Tanner staging secondary sex characteristics. Diagnosis is done by clinical examination and imaging, usually Magnetic Resonance Imaging. Early surgical management by vaginal or abdominal approaches improves the outcome. A 16 years adolescent with appropriate Tanner staging secondary sex characteristics presented to gynecology (OPD) of Kathmandu Model Hospital with complaints of primary amenorrhoea and palpable abdominal mass of 24 weeks size with cyclic lower abdominal pain. Ultrasound showed features suggestive of hematometra. Under spinal anaesthesia, drainage and excision of the transverse vaginal septum was done followed by placement of vaginal mould post-operatively. We present here a case of primary amenorrhoea with pyometra secondary to transverse vaginal septum.

Keywords: primary amenorrhoea; transverse vaginal septum; hematocolpos.

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CASE

A 16 years girl from Rupandehi district presented at the Obstetrics and Gynaecology Out-patients with complaints of cyclical pain over lower abdominal region since three years. This pain used to occur for around 3-4 days and got repeated each month. It was moderate in intensity and sometimes severe enough which required hospital emergency visits. She had not been sexually active yet. She had undergone Examination under Anesthesia (EUA) twice in her hometown with inconclusive results and was then referred to our center.

At our center, findings of general physical examination were normal with appropriate Tanner staging secondary sex characteristics. Abdominal examination revealed tender abdominal mass (24 weeks size), soft to firm in consistency with a smooth regular surface, lower margin couldn’t be assessed. Gross Examination of external genitalia revealed normal findings. There was a small vaginal-introital opening, without obvious bulging of the hymen. Her ultrasound of abdomen and pelvis revealed predominately echogenic collection (volume: 800 ml) in the lower uterine cavity and cervix, which was continuous with upper uterine cavity, suggestive of hematometra. Urine examination revealed urinary tract infection. Parenteral antibiotics were administered for 48 hours as she was febrile too. Spinal anaesthesia was given and examination revealed blind vagina, around two cm in length. Left lateral episiotomy was given and bulge was felt at the upper end of the vagina, which was then pricked with a needle. A small opening was made and around 1500 ml of foul smelling pus mixed with old blood was drained (Figure 1). Repeated lavage of the uterus was done with betadine and saline. Excision of the vaginal septum was done. A 24 F indwelling catheter was inserted and balloon inflated for continuous drainage of pyometra. Vaseline gauze was packed in the vagina. Pus for culture was sent. Then, artificial
mould was made and placed in the vagina, which was continued for five days. Post-operatively, parenteral antibiotics were continued for 48 hours followed by oral antibiotics. On one week follow up patient was doing well and had no fresh complaints.

Figure 1. Picture showing drainage of pus mixed with old blood.

COMMENTS

Primary amenorrhoea is absence of menses at age 13 years when there is no visible development of secondary sexual characteristics or age 15 years in the presence of normal secondary sexual characteristics. When evaluating a patient with primary amenorrhea, the pathophysiology can be attributed to numerous sources. Chromosomal abnormalities associated with gonadal dysgenesis are the most common, accounting for 40% of cases. It is then followed by hypothalamic hypogonadism at 30%. A transverse vaginal septum/imperforate hymen represent only 3-5% of cases. Thus, a genital outflow tract obstruction is an uncommon cause of primary amenorrhoea.

A transverse vaginal septum may result from a failure of complete canalization of the primordial vaginal plate or from lack of union, which represents the junction of the urogenital sinus and the paramesonephric duct contribution. It may be obstructive, with mucus or menstrual blood accumulation in the vagina or uterus described as hematocolpos or hematometra, respectively or nonobstructive, with mucus and blood egress. It can occur at any level within the vagina but is more common in the upper portion, which corresponds to the junction between the vaginal plate and the caudal end of the fused mullerian ducts. Septum is most commonly imperforate and manifest as primary amenorrhoea, abdominal pain, and hematocolpos in adolescence. Perforate septum has a more variable presentation, usually associated with the foreshortened vagina and can present in adolescence or young adulthood.

Diagnosis is either confirmed by sonography or MRI. MRI is most helpful prior to surgery to determine the septal thickness and depth. Surgical repair technique is dependent upon the septal thickness. Smaller septa may be removed by excision followed by end-to-end anastomosis of the upper and lower vagina. Often, a large segment of vagina is absent, making anastomosis difficult which may require skin graft to cover the defect. Furthermore, postoperative vaginal dilatation is necessary to prevent stenosis at the anastomosis site. The Grünberger modification of the Garcia Z-plasty method is a convenient and effective technique for the surgical treatment of transverse vaginal septa.

In patients with a high vaginal septum, only a small portion of vagina is present, and retrograde menstruation occurs early after the onset of menstruation and may predispose to the development of endometriosis, which could well explain the reduced fertility in them. Thus, prompt diagnosis and surgical correction to drain accumulated blood may preserve reproductive capacity by preventing this sequel. Sanfilippo recommends laparoscopy concurrently with TVS excision because of the high rate of endometriosis. Long-term complications include vaginal stenosis and re-obstruction, which may require repeat surgery. There is limited data available on reproductive outcomes, but vaginal births have been reported.

CONCLUSIONS

An adolescent female with primary amenorrhoea should be evaluated keeping in mind its multifactorial etiologies. Proper physical examination and imaging helps in diagnosis of congenital obstructive anomalies and timely surgical intervention is necessary to prevent further complications. However, re-surgeries are sometimes required if there is re-stenosis or re-obstruction.
REFERENCES


