

Case Report

Transverse Vaginal Septum: A Rare Cause of Primary Amenorrhea: Case Report

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Abstract: Genital outflow tract obstruction is a rare cause of primary amenorrhea. A transverse vaginal septum is a defect of vertical fusion between the para-mesonephric ducts and uro-genital sinus. If the septum is complete, the menstrual flow will be obstructed causing primary amenorrhea. The septum is basically a membrane of fibrous connective tissue with both vascular and muscular components formed anywhere along the vagina during embryological development. We present a case of 14-year old female who presented with intermittent, colicky abdominal pain. There was no history of attainment of menarche. The clinical examination revealed a small, blind ending lower vagina with a tough transverse membrane separating the lower portion from the upper genital tract and a firm, tender, bimanually palpable mobile mass in the suprapubic region. The ultrasound examination revealed an hourglass shaped collection with internal echoes within the endometrial cavity extending into the upper vagina. The magnetic resonance imaging of pelvis confirmed the presence of haematometra with hematocolpos in upper 2/3rd of the vagina.

Keywords: Transverse vaginal septum, menarche, MRI, Urogenital sinus

INTRODUCTION

Transverse vaginal septum is a rare congenital anomaly that results from incomplete fusion of the vaginal components of the Mullerian ducts and the urogenital sinus. The septum can vary in thickness and may be located anywhere along the vagina, although most are found at the junction of upper and mid-vagina. Clinical presentation depends on whether it is complete or partial.

With complete septa, menstrual blood accumulates and distends structures above the septum after puberty, resulting in hematocolpos and haematometra. Such patients usually present with cyclic lower abdominal pain and ultrasonic findings of haematocolpometra. Occasionally a lower abdominal mass (haematometra) is palpable. Incomplete septa allow partial egress of menstrual blood and such patients usually complain of crypto-menorrhoea, dysmenorrhoea and dyspareunia [1].

Imaging has an indispensable role in distinguishing transverse vaginal septum from the more common imperforate hymen and agenesis or severe hypoplasia of the cervix.

CASE REPORT

A 14 year-old female presented with history of lower abdominal pain of 2-month duration, the pain was localized to supra-pubic region, intermittent, colicky and associated with nausea. There was no history of attainment of menarche. Physical examination revealed a tender midline mass in the supra-pubic region and normal external genitalia. There was no bulging of hymen. On rectal examination, there was bulging of upper vagina towards the rectum. Breast development was of Tanner stage IV, pubic hair development Tanner stage II. The patient was referred to radiology department for ultrasound abdomen and pelvis. The ultrasound examination revealed an hourglass shape sonolucent collection, measuring 7.9x7.4 cm (craniocaudal by transverse), within the upper vagina extending into the endometrial cavity (Fig. 1). Internal echoes were noted within the collection. Both ovaries were well visualized and were normal. On the same day, the pelvic examination done under anesthesia revealed intact, normal appearing hymenal opening. There was a small, blind ending lower vagina with a tough transverse membrane separating the lower portion from the upper genital tract. The MRI examination of

the pelvis confirmed the presence of haematometra with hematocolpos confined to upper 2/3rd of the vagina. The collection displayed hyperintense signals on T2-weighted (Fig. 2), T1-weighted (Fig. 3). and short tau inversion recovery sequence images indicative of sub-acute hemorrhage (Fig. 4). There was a 5 mm thick septum about 3cm away from the vaginal introitus completely separating the lower 1/3rd of the vagina from the upper portion (Fig. 2). The patient underwent incision and drainage under general anesthesia. A stab incision was made in the septum and 300ml of hemorrhagic fluid was drained. Post op ultrasound revealed near total disappearance of collection.



Fig. 1: Ultrasonography of pelvis in a 14-year old girl. Sagittal section in the midline showing collection with internal echoes in endometrial cavity (arrow) and upper portion of vagina (arrowhead)



Fig. 2: Magnetic resonance imaging of the pelvis in a 14-year old girl. The midline sagittal T2-weighted image shows hyperintense collection in the endometrial cavity (arrow), upper two thirds of the vagina (arrowhead) and a thick septum separating upper two-thirds of vagina from lower 1/3rd (circular arrow).



Fig. 3: Magnetic resonance imaging of the pelvis in a 14-year old girl. The midline sagittal T1-weighted image shows hyperintense collection in the endometrial cavity (arrow), upper two thirds of the vagina (arrowhead).



Fig. 4: Magnetic resonance imaging of the pelvis in a 14-year old girl. Short tau inversion recovery (STIR) sequence. The midline sagittal image shows hyperintense collection in the endometrial cavity (arrow), upper two thirds of the vagina (arrowhead).

DISCUSSION

Transverse vaginal septum is a defect of vertical fusion during embryogenesis of the vagina with an incidence of 1:30,000 to 1:84,000 women [2]. Transverse vaginal septum is classified as class I anomaly by Digwani and Falcone Classification system for vaginal anomalies [3]. Embryologically, the upper two-third of the vagina develops from the Mullerian duct and the lower third of the vagina is formed from the ascending sinovaginal bulb which fuses with the Mullerian duct. Transverse vaginal septum develops due to a failure of resorption of the tissue between the vaginal plate, originating from the urogenital sinus and the caudal aspect of the fused mullerian ducts around the 5th month of gestation. So, this septum divides the vagina into two segments, reducing its functional length and causing outlet tract obstruction. These septa are commonly located in the upper vagina (46% of cases)

but can also be located in the mid (40%) or lower (14%) vagina [3].

A transverse vaginal septum is either perforate (incomplete) or imperforate (complete) and results from varying degrees of failure in resorption of the tissue between the vaginal plate and the caudal aspect of the fused Mullerian ducts. In our case, the premenarchal nulligravid female presented with cyclic pelvic pain and primary amenorrhea. This can be explained by the obstructed menses. The tender midline mass in the suprapubic region suggests the possibility of a complete obstruction. Pelvic examination revealed normal external genitalia. There was no bulging of hymen. Clinical examination of the vulva was normal suggesting that the septum is in the mid- or upper vagina. In the case of a complete septum, patients commonly present with primary amenorrhea and cyclic pelvic pain. Incomplete (perforated) septum may manifest later in life, with dyspareunia and dysmenorrhea. The membrane, when visible will be thick and pink and will not allow transillumination, unlike an imperforate hymen. Patients with a history of imperforate transverse vaginal septum generally have a normal upper genital tract and therefore do not have reduced fertility. While it may occur in isolation it is often combined with other Mullerian duct anomalies like Uterus didelphys. The membrane, when visible will be thick and pink and will not allow transillumination, unlike an imperforate hymen [4].

In some cases where the position of the septum is very low, there is a possibility of a clinical misdiagnosis of an imperforate hymen. However, after excision, histology will show the presence of mullerian duct (mesodermal origin) tissue in the septum [5]. The concurrent occurrence of transverse vaginal septum and an imperforate hymen in a single individual is unique [6]. If the patient has normal secondary sexual characteristics like development of the breasts and pubic hair, ultrasonography or magnetic resonance imaging is indicated. And, if the patient has normal uterus, an outflow tract obstruction should be considered. In this case, trans-abdominal ultrasound scan confirmed that the palpable mass in the suprapubic region as an hourglass shaped sonolucent collection with diffuse low level internal echoes within the endometrial cavity distending it suggestive of a hematometra. This collection was seen extending into the upper vagina suggesting of hematocolpos. And the fact that this collection was mainly within the upper vaginal cavity excludes the possibility of cervical agenesis. Both the ovaries were well visualized and were normal. Both the kidneys and the urinary bladder were normal. In Mayer Rokitansky Kuster Hauser syndrome, there is agenesis of uterus and most or the entire vagina with unilateral renal agenesis due to dysplasia of the mullerian ducts [7].

However, the vaginal septum could not be confirmed on ultrasound. On Ultrasound, a solid appearance and a completely anechoic collection have also been documented. The thick myometrial wall and milder distention of the uterine cavity, as compared with the thin and often imperceptible wall of the vagina aid in differentiating the uterus from the cervix. The lower uterine segment is funnel shaped as it is narrowed with hematometra in the absence of hematocolpos in cervical agenesis [4]. HSG has little role in the evaluation of transverse septum in view of the inherent inability to catheterize the cervix. So, an MR study was indicated. MR imaging helps in depicting the pelvic anatomy and in determining the thickness of the vaginal septum. Identification of the cervix on MR images is crucial for differentiating a high transverse vaginal septum from congenital absence of the cervix [3]. On MRI, there was a collection within the distended vagina which was hyperintense on T1-weighted (T1W), T2-weighted (T2W) and short tau inversion recovery (STIR) images. . On MR images, there is clear delineation of hematometra given the preserved zonal anatomy of the uterus, as are the cervix and endocervical canals, when present. Also, MRI helps in easily identifying associated additional anomalies in view of its multiplanar capability and is critical in helping delineate complex anomalies with marked secondary distortion of the uterovaginal anatomy [4].

CONCLUSION

Transverse vaginal septum is a rare congenital anomaly that can present anytime with a wide spectrum of complaints. Whenever a female in the premenarchal age presents with cyclic, pelvic pain and a suprapubic tender mass, a possibility of transverse vaginal septum should always be borne in mind. Ultrasound aids in localization and characterization of the lesion while MRI will help in confirming the vaginal septum including its thickness and relation to the adjacent structures because of its better delineation.

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