Complete imperforate tranverse vaginal septum with septate uterus: A rare anomaly

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ABSTRACT

The isolated, complete, transverse vaginal septum is one of the most infrequent anomalies of the female genital tract, and when it coexists with a septate uterus, it is even rarer. This report describes a case of transverse vaginal septum with septate uterus. A 12-year-old girl sought medical assessment because of severe cyclic lower abdominal cramping and pelvic pain. Local examination revealed a blind vaginal pouch of 2 cm and on rectal examination a tender pelvic mass was noted. Radiological examination showed transverse vaginal septum in the lower vagina with bicornuate uterus. Surgical resection of the vaginal septum was done under laparoscopic guidance. Hysteroscopy revealed presence of uterine septum which was resected by a resectoscope. Post-operative dilatation of vagina was done to prevent restenosis. Laparoscopic guided abdominoperineal approach is better in such a case as multiple mullerian anomalies may coexist with each other.

KEY WORDS: Imperforate hymen, septate uterus, transverse vaginal septum

INTRODUCTION

The complete, imperforate transverse vaginal septum is one of the most infrequent anomalies of female reproductive tract. The largest series reported by Lodi states an incidence of three cases in 90,000 patients.[1] The estimated incidence is one per 30,000 to 8,4000 women. Transverse vaginal septum results from incomplete fusion between the vaginal components of the mullerian ducts and the urogenital sinus. Clinical presentation depends on whether it is complete or partial. With complete septum, the menstrual blood accumulates in the genital tract resulting in hematocolpos and hematometra. Such patients usually present with cyclic lower abdominal pain and occasionally lower abdominal mass (hematometra) may be palpable. Incomplete septum allows partial egress of menstrual blood and such patients complain of dysmenorrhea and dyspareunia.[2] We present a case of a 12-year-old girl who had a combination of transverse vaginal septum and septate uterus presenting with severe pain in lower abdomen and pelvis.

CASE REPORT

A 12-year-old girl sought medical treatment because of 1 year history of worsening cyclic, pelvic, and lower abdominal cramping that evolved into constant sharp pain. She experienced thelarche at 11 years of age and had never menstruated till date. Her general physical examination was normal. Her breasts were Tanner stage 3 with no masses. Her external genitalia were normal without any clitorimeglay. On local examination, a 2 cm blind vaginal pouch was seen. A tender, globular, firm, smooth mass was noted on rectal examination. On transrectal ultrasonography, echogenic fluid was evident in the uterus, cervix and vagina [Figure 1]. Magnetic Resonance Imaging (MRI) of pelvis confirmed the presence of small shallow lower vaginal segment connected to dilated cervix and uterus with normal appearing ovaries. Findings of bicornuate uterus with transverse vaginal septum were ascertained. Kidneys and pelvicalyceal system was seen normal. On the basis of clinical examination and radiologic studies, diagnosis of imperforate transverse vaginal septum was made and decision of septum resection under laparoscopic guidance was taken.

Intraoperative

Patient was placed in dorsal lithotomy position. Under general anesthesia (GA),

verres needle was introduced and pneumoperitoneum was created. Primary 10 mm intraumbilical trocar was put. One left paraumbilical 5 mm port was created. Uterus was seen as a heart shaped structure with two distinct horns and central depressing groove suggesting a bicornuate uterus [Figure 2]. Both tubes were grossly dilated and congested revealing gross hematosalpinx [Figure 3]. The ovaries were normal in appearance, size, and shape. There was a huge swelling below the uterine isthmus as if the uterus is sitting on a big ball. On local genital examination, blind vaginal pouch of 2 cm was seen. Transverse vaginal septum was identified and grasped with allis tissue holding forceps and stab incision was given. Thick old clotted blood collection of about 700 ml drained out and swelling below the uterus was seen reducing in size and finally disappeared [Figure 4]. The uterine contour altered and now had a broad flat fundus with a central depression suggesting a septate uterus. The vaginal septum was lifted with tissue forceps and separated from vaginal mucosa circumferentially with a scalpel. This approach was used to avoid post-operative stricture formation. Everting sutures were taken at 2, 5, 7, 10 o′ clock position. On hysteroscopy, intrauterine septum was seen and same was resected using resectoscope with monopolar electrode [Figure 5]. Pediatric Foleys no. 8 was inserted and balloon was inflated with 4 ml saline. Post-operative recovery was good and uneventful. Oral estradial valerate 8 mg per day was started to produce uniform endometrial lining. Patient was discharged on the second postoperative day. Weekly follow up was advised to the patient. The patient had vaginal bleeding for about 15 days. Foleys was removed after one month. Regular vaginal dilatation with a vaginal sponge covered with a condom was advised in view of preventing recurrence. Patient comes for regular follow up in every 3 months and is having regular menstrual cycles now.

DISCUSSION

Our case was a combined manifestation of resorption and vertical fusion defects of the mullerian duct in the form of

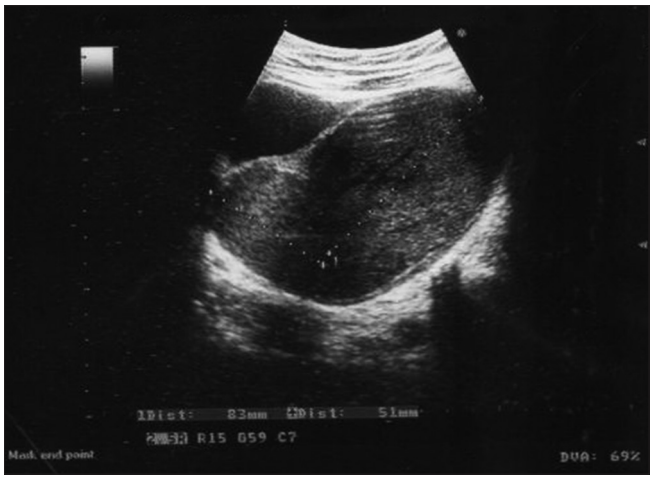

Figure 1: Ultrasonography showing echogenic fluid in the uterus, cervix, and vagina

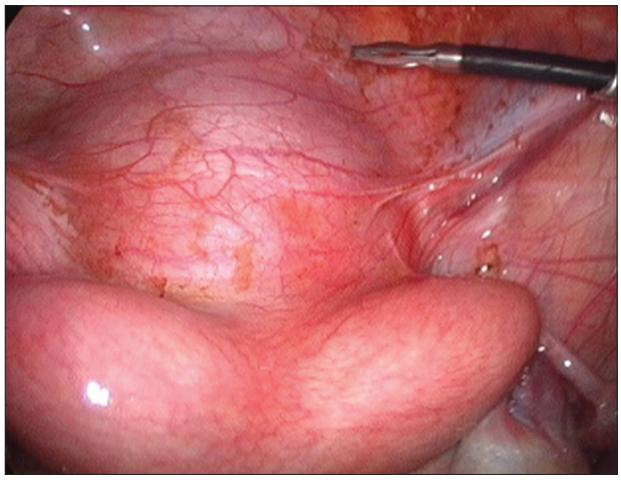

Figure 2: Laparoscopic view of internal genitalia suggesting bicornuate uterus with swelling below the isthmus

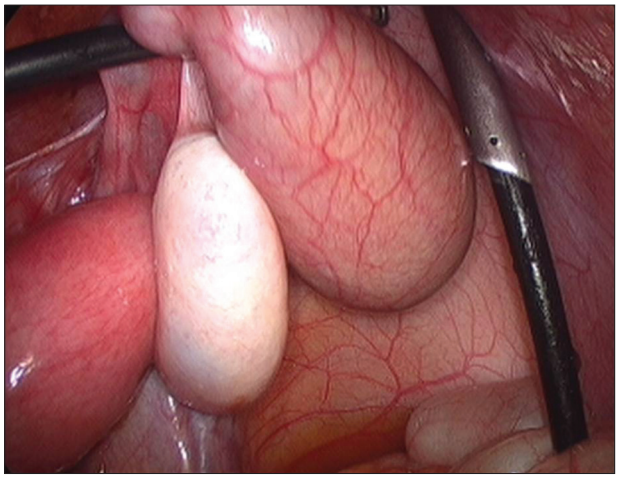

Figure 3: Laparoscopic view showing gross hematosalpinx with normal ovaries

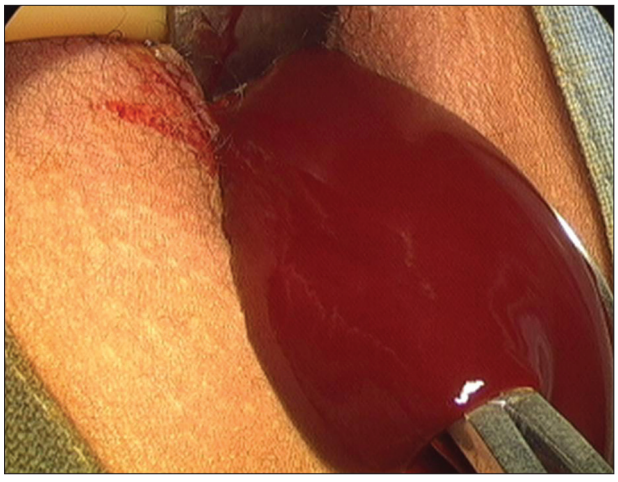

Figure 4: Hematocolpometra draining out

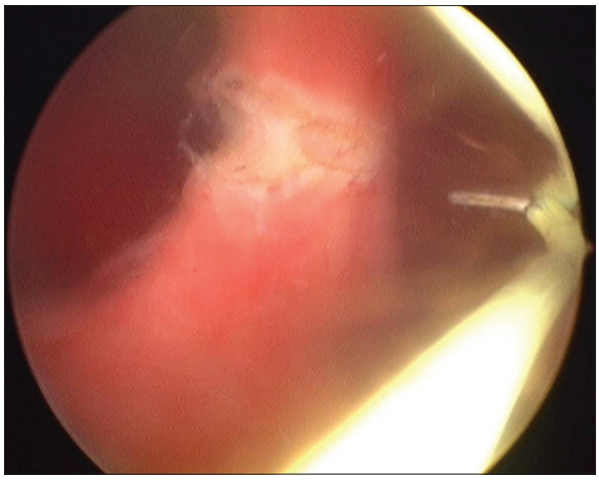

Figure 5: Hysteroscopic septal resection

septate uterus and transverse vaginal septum. Preoperative assessment of transverse vaginal septum includes elicitation of medical history and performance of physical examination. The family history of genetic abnormalities, secondary sexual characters, presence of pelvic mass, and appearance of internal and external genitalia should be noted. Radiologic studies that help in diagnosing transverse vaginal septum are pelvic ultrasonography, magnetic resonance, and computerized tomographic imaging. MRI is considered the criterion standard for imaging mullerian duct anomalies. It provides high resolution images of uterine body, fundus and internal structures.[3] Intrvenous pyelography should be done to detect associated anomalies of genitourinary tract. The differential diagnosis of transverse vaginal septum includes imperforate hymen, cervical dysgenesis, and vaginal agenesis. The most common is imperforate hymen, which accounts for the commonest anomaly of vaginal outflow tract with an incidence of 1 in 1,000 women.[4] Clinically, the vaginal septum is located between upper 1/3 and lower 1/3 of the vagina, while the imperforate hymenal membrane is present between the labia across the vaginal vestibule. In our case, the septum was present in lower 1/3 of the vagina creating a blind vaginal pouch of 2 cm. In Lodi's series,[1] 46% of the vaginal septums were upper, 40% were middle, and 14% were in the lower vagina. Restenosis and reaccumulation of fluid in transverse vaginal septum is more common than imperforate hymen, so one needs to make proper differentiation between the two entities. Treatment of septum is surgical. In our case, we followed a abdominoperineal approach that lead to a complete diagnosis of septate uterus with transverse vaginal septum. Complications include enterotomy, cystotomy, vaginal stenosis, and recurrence of the symptoms. Late sequelae include endometriosis, which is commonly associated with the upper vaginal septums. If patient is not sexually active, as in our case, vaginal dilatation is to be maintained to prevent recurrence.

CONCLUSION

Transverse vaginal septum is a very rare mullerian anomaly which may be associated with imperforate hymen, imperforate anus, ectopic ureter, vesicovaginal fistula, bicornuate uterus, and septate uterus.[5] All physicians should be aware of this entity in differential diagnosis of hematocolpos with abdominal pain and primary amenorrhea in early adolescent years. Early diagnosis could be based on premenarchal gynecological examination and could lead to correct management to avoid complications of endometriosis, dysmenorrhea, and infertility. Strict follow up should be emphasized because transverse vaginal septum carries a higher risk of re-occlusion leading to recurrence of symptoms. Laparoscopic guided abdominoperineal approach is better as multiple mullerian anomalies may coexist with each other.

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