Complete uterine septum involving cervix with unilateral external cervical os stenosis: diagnosis and management

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ABSTRACT

Background: Congenital external cervical os stenosis is a form of Mullerian duct abnormality which is rare in virginal adolescents.
Case: Pre-operatively, two non-sexually active adolescents of 12 and 14 years of age, with history of light menses and severe lower abdominal pain for 2 and 4 months respectively, were diagnosed with an obstructed, oblique vaginal septum syndrome. However, at operative vaginoscopy, a single cervix with a duplicated cervical os and unilateral cervical stenosis was discovered, confirming the diagnosis of a complete uterine septum involving the cervix and unilateral cervical stenosis. They were treated by expanding the affected external cervical os while leaving the hymen intact. A year following the treatment, follow up results found no recurrence of symptoms.
Summary and conclusion: Operative vaginoscopy using a hysteroscope is crucial and recommended for the diagnosis and treatment of congenital external cervical os stenosis in virginal adolescents.
Keywords: External cervical os stenosis, Vaginoscopic approach, Hysteroscope, Oblique vaginal septum, Complete uterine septum

INTRODUCTION

Congenital external cervical os stenosis is a rare Mullerian duct abnormality, characterized by the external cervical os having a diameter less than 4mm.[1] Patients normally present with hypomenorrhea associated with cyclic pelvic and/or lower abdominal pain [2] or primary dysmenorrhea.

In this present study, we report two unique cases of complete uterine septum involving the cervix with unilateral external cervical os stenosis, but only one cervix, in sexually inactive adolescents. Both the patients were treated with operative vaginoscopy using a hysteroscope. This case report is important because of the patient's presentation with a common symptom of dysmenorrhea caused by a rare congenital anomaly, unilateral cervical stenosis in a patient with a complete uterine septum involving the cervix, resulting in the need for a high level of suspicion to make the diagnosis. The mode of treatment, which is the use of minimally invasive process without disrupting the hymen, is also very rarely used.

CASE PRESENTATION

Case 1: A 12 year-old adolescent, who had experienced menarche 3 months prior to the date of admission, presented with severe lower abdominal pain and mild vaginal bleeding during menstruation for about 2 months. Pelvic magnetic resonance imaging (MRI) revealed septate uterus (Fig 1A), two cervical canals (Fig 1B) and a single vagina. B-ultrasound revealed the absence of her left kidney. During vaginoscopy using a hysteroscope (Karl-Storz Company, Germany) of 5.4 mm outer diameter, the vagina was found to be normal with no obvious protuberance seen in unilateral vaginal walls as usually seen in an oblique vaginal septum. On further inspection, a single cervix revealed a normal appearing external cervical os on the right with a slight bulging on the left side of the cervix (Fig 1C) due to the retained menstrual blood. Old retained menstrual blood was removed with double action 5-Fr forceps and the left stenosed external cervical os was identified. It was very narrow with a diameter of about 1mm and dark red liquid was seen flowing out through this narrow orifice since this procedure was performed during menstruation phase. The stenosed left external os was then expanded with a needle electrode and hemostasis was achieved using a roller-ball electrode. The hysteroscope was further introduced through each external cervical os which revealed normal cervical canals with each cervical canal opening into a uterine horn with a single tubal ostium and a medial septum. This confirmed the diagnosis of a unilateral cervical os stenosis in a case of complete uterine septum involving the cervix and excluded the diagnosis of oblique vaginal septum. At the end of this procedure, diameter of the left external cervical os was about 1cm, with no obvious bleeding. There was no disruption of the hymen and the patient was discharged the same day.

Case 2: 14 year-old adolescent was admitted to the Third Xiangya Hospital for dysmenorrhea for a period of 4 months. She had experienced menarche a year prior to the presentation. A pelvic MRI revealed a complete septum of her uterus (Fig 2A) and two cervical canals. B-ultrasound showed the absence of her left kidney. Vaginoscopy using a hysteroscope discovered similar findings to the first case. In this case too, the adolescent had a single cervix, two cervical canals and a stenotic left external cervical os

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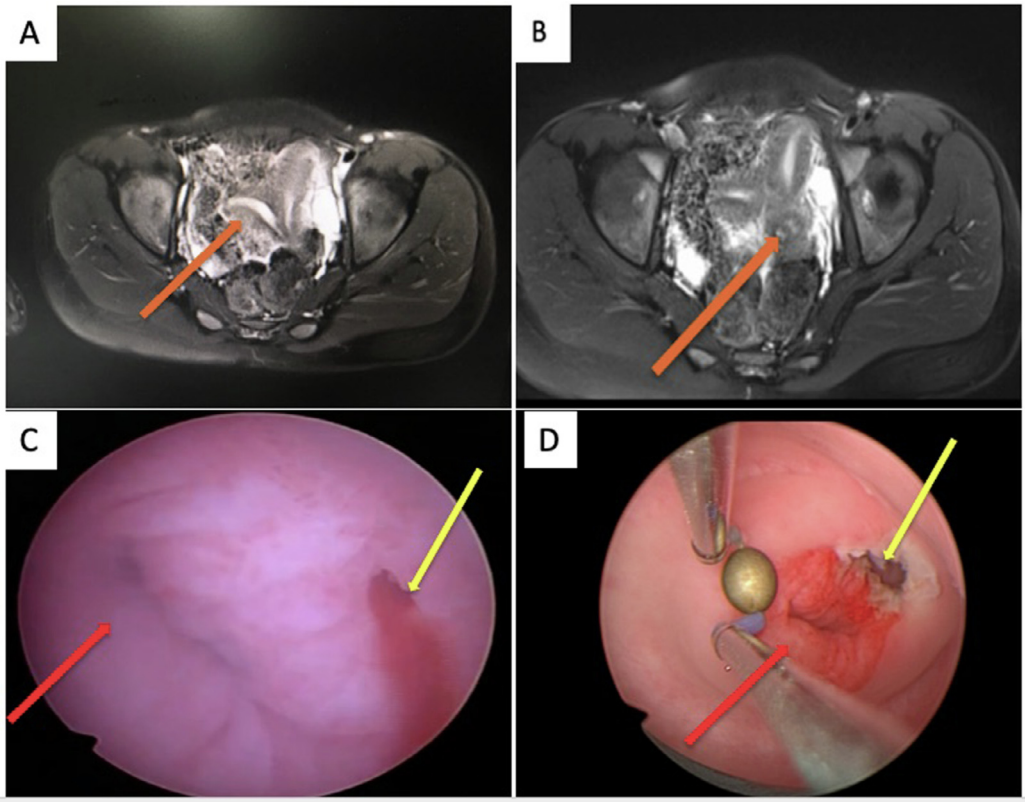

Figure 1. (A) Pelvic MRI showing septate uterus (orange arrow), (B) two cervical canals (orange arrow); (C) inspection of the cervix revealed a normal appearing external cervical os (red arrow) on the right with a slight bulging in left side of the cervix (yellow arrow) due to accumulation of old retained menstrual blood, seen at the beginning of vaginoscopy using a hysteroscope, (D) dilated cervical os at the end of the vaginoscopy combined with hysteroscope(yellow arrow; red arrow is the normal right cervical os).

(Fig 2B). On further exploration of the uterine cavity with a hysteroscope, after expanding the left stenosed external os, a complete septum was found. The mode of treatment was exactly the same as in first case by expanding the stenosed left cervical os with needle electrode.

A successful follow up using vaginoscopy combined with a hysteroscope, 2 months after the surgery, revealed the once stenosed left external os was normal (Fig 2D). Both the patients were on follow up for a period of 1 year and were found to remain free of any recurrence of symptoms.

SUMMARY & CONCLUSION

Using the AFS classification scheme, Mullerian duct anomalies (MDA) can be grouped into three major categories based on the stages of embryonic development: underdeveloped anomalies, non-fusion and non-degeneration.[3] The first group includes underdevelopment of the right and left fallopian tubes, uterus, cervix and upper 2/3rd of vagina. The second group includes non-fusion of midline that separates left and right uterus, cervix, and upper vagina. The third group is non-degeneration, which occurs due to an arrest in the third stage of embryonic development. As such, fused midline segments along the uterus, cervix and vagina degenerate or reabsorb. And thus failure of reabsorption results in septate uterus, cervical and vaginal canal. Complete septate uterus has two uterine cavities, two cervical and vaginal canals as the Mullerian ducts fuse along all of these segments and any fused portion may not degenerate.[4] However, it should be noted that both the cases mentioned in this case report have complete septate uterus with two uterine cavities, two cervical canals but only one cervix and vagina. This is a rare MDA, which is inconsistent with the current understanding of Mullerian development.

External cervical os stenosis, a rare form of Mullerian duct abnormality, is defined as an external cervical os less than 4mm in diameter. Stenosis of unilateral external cervical os, usually presents with primary dysmenorrhea. Prepubertal girls are commonly asymptomatic and only diagnosed incidentally until after menarche. Early diagnosis and management are critical to reduce the risk of hemotometra (accumulation of blood in the uterus),[5] pyometra,[6] and endometriosis (due to retrograde flow of menstrual blood into the pelvis region in premenopausal women).[6] If left untreated, infertility [6] may develop at the later stage of complication due to long-term obstruction of menstrual blood in the uterus owing to stenosed cervical os. Though there are various imaging techniques for the diagnosis such as pelvic MRI, color doppler ultrasound, abdominal X-ray, computerized tomography (CT), the diagnosis in sexually inactive adolescent requires a high degree of suspicion due to it's rarity as a cause of dysmenorrhea and because of the narrow undeveloped vagina and an intact hymen, which may hamper a routine vaginal examination using a speculum. Pelvic MRI is comparatively a better imaging method because of its detailed anatomical pictures. However, the

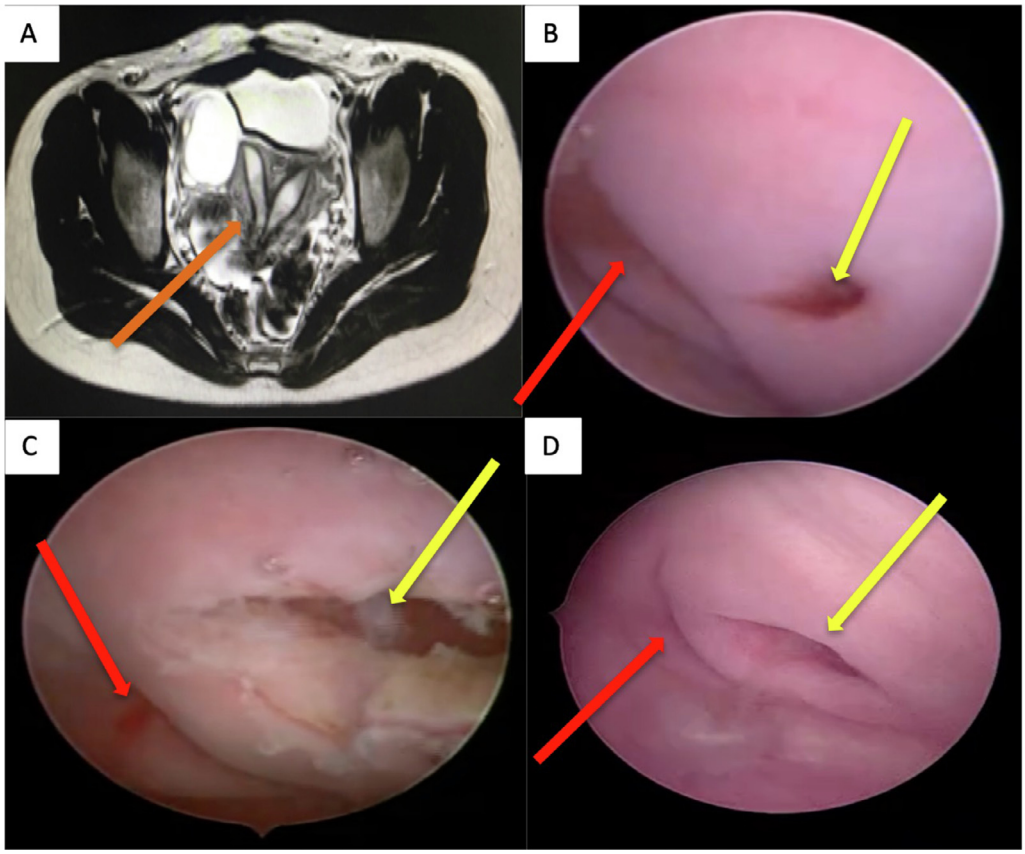

Figure 2. (A) Pelvic MRI depicting complete septate uterus (orange arrow), (B) vaginoscopic image with the hysteroscope showing two external cervical os, (C) image after the surgery, and (D) follow up image indicating both the external os as normal external cervical os (red arrow indicates normal right external os whereas yellow arrow indicates stenosed left external cervical os in Fig B,C and D).

best method to make an accurate diagnosis is operative vaginoscopy.

Both the patients in this case report were adolescent with an intact hymen. They experienced onset of symptoms just after menarche. A proper routine history was taken, and physical examination including external genitalia and recto-abdominal exam was performed. Routine lab tests, pelvic and renal ultrasonography, and MRI of the pelvis preoperatively diagnosed it as oblique vaginal septum (variant of obstructed hemivagina and ipsilateral renal anomaly). Oblique vaginal septum syndrome mimics our cases, as patients are asymptomatic until menarche, present with primary dysmenorrhea, have old blood clots in unilateral uterine cavity, ipsilateral renal agenesis, and similar imaging findings. Then again, our cases are different from oblique vaginal septum due to the vaginal anatomy as well as the typical anatomical cervical canal structure with presence of normal cervical mucous, seen during vaginoscopy using a hysteroscope. The final diagnosis of unilateral cervical os stenosis with a complete uterine septum was confirmed during the operative vaginoscopy. This procedure also confirmed the presence of an unobstructed normal vagina, a single cervix and two cervical canals. The management of our cases was minimally invasive because vaginal exploration was limited due to immature genital tract and intact hymen. Thus, surgical intervention was intended via vaginoscopy combined with a hysteroscope using vaginoscopic approach, in order to create a wider opening of stenosed external cervical os without disrupting the hymen. However, the resection of the uterine septum was not necessary at that moment. The advantage of this day care surgical technique is no post-operative discomfort and pain, and therefore, the patient can resume their normal daily routine just after the discharge. Also, there is no retraction of the vaginal wall in this procedure. Such an approach is crucial for the diagnosis and treatment of these cases because of its minimally invasive nature, which is effective in preventing the disruption of hymen and tearing of the undeveloped vagina.

AUTHOR CONTRIBUTIONS

DX designed the study conception. AZ and NK recruited patients and collected information, analyzed images, drafted and revised the article. DX critically revised the manuscript for important intellectual content. The final version of the manuscript was approved by all the authors. All authors agree to be accountable for all aspects of the work.

Declaration of Competing Interest

The authors have no conflicts of interest.

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REFERENCES

1. Barbieri R.L.: Stenosis of the external cervical os: An association with endometriosis in women with chronic pelvic pain. Fertil Steril 1998. doi:10.1016/S0015-0282(98)00189-7.
2. Zhu X., Xu D., Allornuvor G., Gao F., Xue M.: Hysteroscopic Management of Congenital External Cervical Os Stenosis Using a "No-Touch" Technique in an Adolescent. J Pediatr Adolesc Gynecol 2015; 28(2):e23–6. doi:10.1016/j.jpag.2014.07.007.
3. CROSBY W.M., HILL E.C.: Embryology of the Mullerian duct syytem. Review of present-day theory. Obstet Gynecol 1962; 20:507–15.
4. Robbins J.B., Broadwell C., Chow L.C., Parry J.P., Sadowski E.A.: Müllerian duct anomalies: Embryological development, classification, and MRI assessment. J Magn Reson Imaging 2015; 41(1):1–12. doi:10.1002/jmri.24771.
5. Rezai S.: Hematometra and Hematocolpos, Secondary to Cervical Canal Occlusion, a Case Report and Review of Literature. Obstet Gynecol Int J 2017; 6(3):1–3. doi:10.15406/ogij.2017.06.00208.
6. Moramazi F., Roohipoor M., Najafian M.: Association between internal cervical os stenosis and other female infertility risk factors. Middle East Fertil Soc J 2018; 23(4):297–9. doi:10.1016/j.mefs.2018.02.002.