CASE REPORT

Haematometrocolpos and acute pelvic pain associated with cyclic uterine bleeding: OHVIRA syndrome

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SUMMARY

Obstructed hemivagina and ipsilateral renal agenesis syndrome is a complex urogenital malformation usually presenting with obstructed menses and pelvic pain during female adolescence. The diagnosis can be established preoperatively with relative certainty by MRI or ultrasonography, and outcomes are usually satisfactory following surgical resection of the septal portion of the obstructed hemivagina. Such cases are best managed in referral centres with expertise in anatomical disorders of the female genital tract.

BACKGROUND

Outflow anomalies involving the female genital tract have been considered traditionally to result from developmental aberrations in the müllerian duct, urogenital sinus or both.¹ The syndrome of obstructed hemivagina and ipsilateral renal agenesis (OHVIRA), also known as Herlyn-Werner-Wunderlich syndrome, is a specific urogenital malformation that is associated with uterus didelphys, absence of one kidney and ipsilateral outflow obstruction secondary to a blind hemivagina. In most cases, these obstructive problems present in adolescent girls shortly after menarche with pelvic pain and dysmenorrhoea secondary to a haematometrocolpos.¹ Prompt and appropriate diagnosis and treatment are facilitated by proper preoperative imaging and surgical expertise.^{2,3}

CASE PRESENTATION

A 13-year-old virginal girl was referred to the reproductive medicine clinic with a vaginal mass associated with severe midline pelvic pain throughout the previous month. Her mother did not recall use of any potential teratogens during pregnancy. Since menarche, she reported six menses lasting 3–5 days requiring 6–7 tampons or pads per cycle. Initially, dysmenorrhoea was managed satisfactorily with acetaminophen, but she developed deep, persistent pelvic pain 1 month prior to consultation. She had been known to have a sole left kidney discovered on a voiding cystourethrogram performed at age 1 due to a febrile urinary tract infection. The referring physician performed an abdominal ultrasound and noted a 5 cm upper vaginal mass. No comment was made about a potential uterine anomaly. The physician performed an examination under general anaesthesia with only a bivalve vaginal speculum

and the preoperative diagnosis of ‘suspected micro-perforate hymen’. The surgical note reported only a large vaginal mass with no visible cervix. The patient was awakened from anaesthesia without further exploration or resolution of the vaginal mass. Postoperatively, the patient was referred immediately to our institution.

INVESTIGATIONS

A pelvic MRI was obtained which revealed a didelphic uterus with haematometrocolpos of the right horn and vagina secondary to an obstructing hemivagina. The vaginal mass measured 6×6×5 cm. The left horn appeared normal without haematometrium although MRI did not clearly demonstrate a vaginal canal on the left side (figure 1). A single left kidney was verified by MRI. The obstructing septal thickness measured approximately 7 mm. Findings were consistent with OHVIRA syndrome with a symptomatic haematometrocolpos, and a discussion was held with the patient and her family concerning surgical options and long-term expectations.

TREATMENT

At her second surgical procedure, vaginoscopy with a paediatric cystoscope revealed a bulging mass on the right side of the vagina (figure 2) with a normal cervix located at the apex of the left upper vagina (figure 3). Incision and resection of the right-sided oblique septum produced about 500 mL of old unclotted blood (figure 4). Vaginoscopy confirmed the presence of a cervix on the right with blood draining from the haematometrium into the hemivagina (figure 5). Following surgical resection of the vaginal septum (figure 6), the vaginal mucosa was reanastomosed with 000 poliglecaprone absorbable suture. We elected not to perform laparoscopy to assess endometriosis. A Mileyx silicone vaginal dilator (Cooper Surgical, Trumbull, Connecticut, USA) with dimensions 78×14 mm was fitted in the operative suite for postoperative use to assist in the maintenance of functional vaginal integrity. The vaginal dilator was recommended to prevent postoperative vaginal adhesions given the thickness of the vaginal septum (7 mm) that required a few interrupted absorbable sutures to reanastomose the vaginal margins. Often, a vaginal dilator is not used following resection of a thin oblique septum but can be considered on a case-by-case basis.¹

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Figure 1 (A) MRI (T1 weighted) demonstrating uterus didelphys, haematometrium and haematocolpos. (B) Right haematometrium emptying into right haematocolpos (arrow). (C) Single left kidney with right renal agenesis.

OUTCOME AND FOLLOW-UP

Her pelvic pain had largely resolved on postoperative day 1. The patient tolerated nightly use of the vaginal dilator and discontinued insertion at the 4-week postoperative visit. Her first menses postoperatively was associated with only modest dysmenorrhoea, and at 4 months, she continues to do well.

DISCUSSION

The presentation of this case and the diagnostic and surgical manoeuvres used are similar to other reports in the literature.^{13–8}

Figure 3 Normal cervix at the apex of the left hemivagina (vaginotomy). This finding was not visualised at the prior operation with a vaginal speculum.

Mullerian anomalies occur in up to 4%–7% of reproductive age women, with approximately 40% of those cases associated with renal anomalies.¹²⁴ The mullerian duct develops then fuses with the urogenital sinus between the 6th and 12th gestational weeks. According to one of the widely accepted hypothesis, OHVIRA syndrome is caused by a developmental arrest at about 8 or 9 weeks of gestation.^{5–79}

Two hypotheses speculating on the mechanism resulting in OHVIRA syndrome appear in the literature. One hypothesis argues that the mullerian (paramesonephric) ducts from the upper vagina fail to fuse fully with the urogenital sinus.⁴ This

Figure 2 Haematocolpos within the right hemivagina (vaginotomy). Notice bulge into the left hemivagina.

Figure 4 Haematometrium is drained (gross).

Figure 5 Right cervical os draining blood from haematometrium following incision into the right obstructed hemivagina (vaginotomy).

theory does not adequately explain the associated agenesis of the kidney and ureter. A different hypothesis argues that the wolffian (mesonephric) duct forms the entire vagina, and unilateral developmental arrest of the wolffian duct and mesonephros would explain simultaneous renal and ureteral agenesis as well as vaginal occlusion.^{1-3 10} (The mesonephros outpouches to form the metanephric diverticulum which eventually develops into the kidney.) Although unilateral renal agenesis usually occurs on the ipsilateral side of the obstructing vaginal septum, there are case reports of contralateral renal agenesis to the obstructed hemivagina, the presence of both kidneys with uterus didelphys and unilateral vaginal obstruction, and degrees of variability in vaginal septum.^{3 8} Together, it appears that müllerian outflow anomalies are probably multifactorial in nature with polygenic causes, environmental insults and teratogenic drugs all implicated.³

Imaging modalities to consider include transabdominal ultrasonography of the pelvis and renal system (transvaginal ultrasound may not be tolerated by the young patient), MRI (gold standard) or transvaginal three-dimensional ultrasonography with renal imaging in an adolescent who is older or who has initiated sexual intercourse.^{1 3 9}

Treatment of OHVIRA syndrome inevitably ends in surgery, with medical management (ovarian suppression) typically used

Figure 6 Portions of excised hemivagina (gross).

Patient's perspective

It was frustrating to learn that my daughter's first trip to the operating room did not resolve her acute medical issue. We were relieved that a second opinion undertook proper imaging before surgery and had an organized approach to her care.

Learning points

- Persistent and progressive pelvic pain beginning with menarche should prompt investigation for an obstructive outflow anomaly.
- In cases of complex genital anomalies, comprehensive imaging of the renal and reproductive systems by MRI or ultrasound should precede any surgical exploration or treatment.
- Complex genital malformations are best addressed in referral centres experienced in diagnosing and treating such anomalies.

to bridge patients until the surgical resection for the vaginal septum can be safely performed. As such, the end goal of medical management is complete menstrual cycle suppression with continuous combination oral contraceptives, gonadotropin releasing hormone (GnRH) agonists or depot medroxyprogesterone acetate.¹ Medical treatment may be discontinued following successful surgery.

The role of laparoscopy to assess for peritoneal endometriosis remains debated. In most cases, it appears that endometriosis resolves once the obstructive anomalies have been addressed, making the addition of laparoscopy for diagnosis and excision unnecessary.^{3 8} Laparoscopy (or prolonged ovarian suppression with combination oral contraceptives) could be considered subsequently in those with persistent and problematic dysmenorrhoea. Postoperative use of vaginal dilators may be useful to maintain cavity integrity.¹

Complex genital anomalies are frequently misdiagnosed and treated incorrectly in general gynaecological practice. This case illustrates the importance of a correct diagnosis initially to avoid inappropriate or unnecessary surgery. In most cases, referral to centres familiar with diagnosis and management of complex genitourinary anomalies is wise.^{2 3}

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Reminder of important clinical lesson

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