

A Rare Case Report of a Congenital Imperforate Hymen Causing Obstructive Uropathy and Constipation in an 11-Year-Old Girl

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Abstract: Imperforate hymen is a rare congenital anomaly that can cause mild to severe complications, which leads to an increase in morbidity rates. The incidence is 1:1000, and there was often a delay in diagnosis since the symptoms were not specific until it caused several complications in other organ systems. We reported a case of obstructive uropathy and constipation in an 11-year-old girl who presented with huge hematocolpos due to imperforate hymen. The main symptom was severe lower abdominal pain with a visual analogue scale of 8. This case shows the significance of a physical examination in diagnosing, increasing efficiency in terms of time and cost, and supporting an earlier diagnosis. A corrective hymenectomy, blood drainage, and hymenoplasty were performed as the definitive management. The patient was able to urinate and defecate within seven hours after surgery. The patient also had her period three weeks after surgery.

Keywords: imperforate hymen, congenital anomaly, vulvovaginal anomaly

Introduction

The genital system is inert throughout the fifth and sixth weeks of fetal development.¹ During this period, the mesonephric (Wolffian duct) and paramesonephric (Müllerian duct) pairs of genital ducts are present. The lack of anti-Müllerian hormone (AMH) and the sex-determining region Y (SRY) gene in females causes the Wolffian ducts to recede and the Müllerian ducts to progressively differentiate.² The urorectal septum, which divides the rectum from the urogenital sinus, occurs in the seventh week of gestation. The Müllerian ducts descend to the urogenital sinus during the ninth week of pregnancy, creating the uterovaginal canal and implant. The Müllerian ducts unite to form a primitive uterovaginal canal during 11–12 weeks of gestation.^{2,3} The fetal hymen forms from the growth of the sinovaginal bulbs (where the Müllerian ducts meet the urogenital sinus) in the fifth month of gestation, when the vaginal canalization is finished. The hymen perforates typically at approximately 22 weeks of gestation. Failure of perforation during neonatal development causes imperforate hymen.

The hymen is a squamous epithelium-thin membrane that generally circumscribes the opening of the vagina with a small circular opening in the center.⁴ Imperforate hymen is a congenital anomaly in which the hymen completely covers the vaginal opening, with a prevalence of 0.1% of female births.^{4,5}

Even if the case is rare, it is considered the most common congenital malformation of the female genital tract.⁶ Delays in diagnosis could cause mild to severe complications in various organ systems.⁷ The symptoms also usually do not occur until the children reach their menarche age. They often remain unaware of the cyclic abdominal pain and amenorrhea complaints. The accumulation of menstrual blood in the vagina (hematocolpos), or uterine cavities (hematometra), or both (hematocolpometra) may cause severe complications in other organ systems.^{8,9} This report describes the case of an 11-year-old girl with an imperforate hymen presenting with obstructive uropathy and constipation.

Case Report

Patient Consent

The parents gave verbal and written informed consent for the publication of this case, including the patient's details and clinical images.

Case Presentation

An 11-year-old girl presented to our emergency department with severe lower abdominal pain. The pain lasted for approximately four days and progressively worsened. Past medical history revealed urinary retention symptoms, presenting with a small amount of urine output in an effortful urination. Additional symptoms included dysuria, post-void residue, suprapubic fullness, dark-yellow urine, flank pain, and low-grade fever over the past two days. The patient also complained of constipation. There was no hematuria or hematochezia.

The patient was born from a spontaneous vaginal birth with a weight of approximately 3200 grams at 38 weeks gestational age at the time of delivery. There was a perinatal history of four months of breastfeeding, appropriate growth and developmental milestones, and complete immunization or vaccine records. The patient did not experience her first menarche.

Investigation

Vital signs showed normal blood pressure (110/70 mmHg), respiratory rate (23 bpm), and temperature (36.2 Celsius degree), with tachycardia (124 bpm) and a visual analogue scale of 8. The patient was defined as overweight with a body mass index of 29.7 from the calculated ratio formula of 66 kilograms of body weight and 149 cm of body height.

Physical examination revealed abdominal distension, suprapubic tenderness, secondary sex characteristics, and complete external genitals. There was a bulging hymen showed as a bluish-shiny mass at the vaginal introitus (Figure 1A).

Further investigation revealed that she had been experiencing lower abdominal pain periodically, with a time interval in between, for the past five months. She did not recognize the pain because it initially perceived as a common abdominal discomfort, which did not significantly impact her quality of life. The patient denied having a family history of congenital conditions. There was a history of complete abortus of her mother and incomplete abortus of her aunt. No history of seizure, antibiotics or cytotoxic drug use, any herbal consumption, or radiation exposure during pregnancy.

Ultrasonography examination revealed a huge hypoechoic fluid-filled mass in the vaginal lumen, indicated as hemato-colpos, an accumulated blood-filled dilated vagina (Figure 2A). There was also a typical compression of the vesica urinaria (VU) due to the mass. The uterus was normal. Hematological investigations revealed leukocytosis (WBC $14.25 \times 10^3/\mu\text{L}$) with a high neutrophil (78%) and low lymphocyte (16.6%). The rest were within the reference range (Table 1).

Final Diagnosis

The patient was diagnosed with an imperforate hymen. Overweight was also established based on nutritional status.

Treatment

Hymenectomy with cross-incision method was performed under general anesthesia (Figure 1B). Approximately 600 mL of dark blood was obtained from the drainage (Figure 1C). Hymen reconstruction, or hymenoplasty, was carried out at the end.

The patient was treated with cefazolin (2x1 grams) and ketorolac (3x30 milligrams) intravenously for the pre-operative treatment. Cefadroxil (3x500 milligrams) and etoricoxib (2x90 milligrams) were prescribed perorally for five days for the take-home medication. The patient was advised to avoid excessive manipulation of the post-operative wound, maintain genital hygiene, follow a healthy diet, increase physical activity, and consult the pediatricians for optimal growth and development.

Outcome and Follow-Up

Seven hours after surgery, the patient felt mild pain in the post-operative wound. She was able to urinate and defecate. A post-surgery ultrasonography revealed an empty urinary bladder after a complete urination (Figure 2B). Within seven days, the wound showed a substantial improvement with no pain. Complete wound healing was observed two weeks after surgery (Figure 1D). The patient also had no complaints and reported her period three weeks after surgery.



Figure 1 (A) Bulging hymen showed as a bluish-shiny mass at the vaginal introitus; (B) Hymenectomy performed with cross incision method in an imperforate hymen; (C) Drainage results of a hematocolpos showed a large volume of dark color blood; (D) Complete wound healing of two weeks post-hymenectomy with a form of vaginal opening (black arrow).

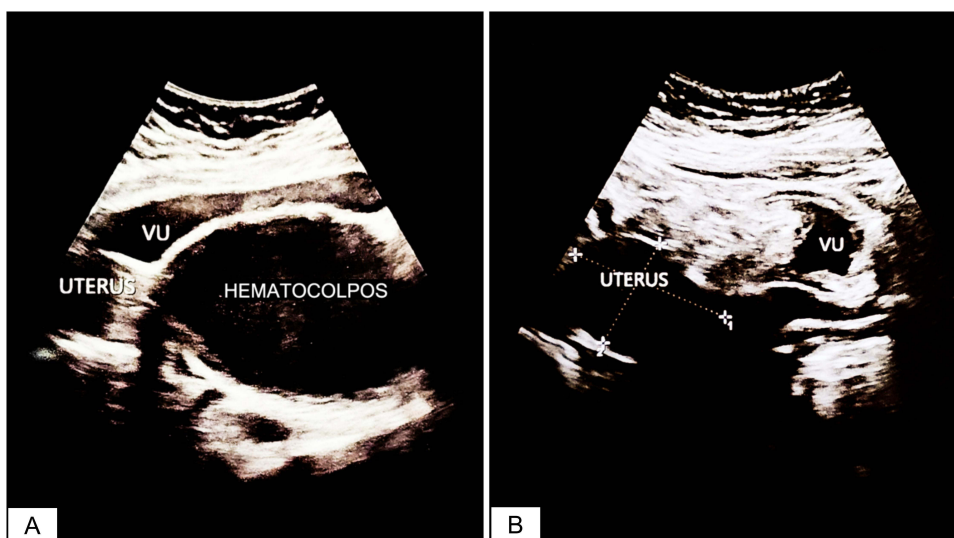


Figure 2 (A) Hematocolpos showed as hypoechoic mass fulfilled the vagina in an ultrasonography; (B) Post-hymenectomy ultrasonography revealed an empty urinary bladder after a complete urination.

Table 1 Hematological Investigations Results

Parameter	Value	Reference Range	Measurement Unit	Interpretation
WBC	14.25	4.1–11.0	$\times 10^3/\mu\text{L}$	High
Neutrophil	78	32–52	%	High
Lymphocyte	16.6	30–60	%	Low
Eosinophil	0.2	0–4	%	Normal
Monocyte	5.1	2–8	%	Normal
Basophil	0.1	0–1	%	Normal
RBC	5.07	4–5.3	$\times 10^6/\mu\text{L}$	Normal
Hb	12.7	11.8–15	g/dL	Normal
HCT	38.2	35–45	%	Normal
PLT	353	150–450	$\times 10^3/\mu\text{L}$	Normal
BT	2	1–5	Minute	Normal
CT	9	5–15	Minute	Normal
HBsAg	Non-Reactive	–	–	–
HIV	Non-Reactive	–	–	–

Abbreviations: WBC, White Blood Cell; RBC, Red Blood Cell; Hb, Hemoglobin; HCT, Hematocrit; PLT, Platelet; BT, Bleeding Time; CT, Clotting Time; HBsAg, Hepatitis B Surface Antigen; HIV, Human Immunodeficiency Virus.

Discussion

A bulging hymen, characterized by a bluish-shiny mass in the vaginal introitus, does not always occur in an early case.¹⁰ In a late complication, bladder compression can lead to urinary retention, hydronephrosis, or kidney injury. Mechanical obstruction in the urinary tract disrupts the urine outflow, further manifesting urinary tract diseases such as stones, infection, and hydronephrosis, leading to acute kidney injury.^{11,12} In more severe cases, the mass may obstruct the intestinal tract, leading to constipation. Diagnosis in cases of imperforate hymen with complications can be a challenge due to a lack of knowledge or an incomprehensive examination. Therefore, the wide range of differential diagnoses necessitates a precise and conscientious physical examination.

Imperforate hymen is rarely diagnosed during the neonatal period and is usually present later in puberty as cyclic abdominal pain, primary amenorrhea, or accumulating as a pelvic mass. The complications of these congenital anomalies can actually occur from the perinatal period, such as fetal hydrometrocolpos (HMC).¹³ Although it is a rare complication, HMC can occur due to cervical or endometrial mucus.^{13,14} In a further period of age, a congenital vaginal obstruction can cause infertility and is associated with endometriosis.^{15–17}

Education related to risk factors for imperforate hymen is also important to minimize reoccurrences in subsequent pregnancies. Even though the evidence-based etiology is still unclear, several things can still be attempted, including avoiding radiation exposure, cytotoxic drug usage, and non-standardized herb consumption.^{18,19} Seeking optimal nutrition during pregnancy is also necessary.²⁰

Surgical correction is a definitive medical management for imperforate hymen. Post-operative wound observations should be carried out periodically to evaluate the healing process. Consider installing a plug to prevent wound closure after the surgery. Other congenital anomalies are often associated with other congenital vaginal obstruction. A comprehensive organ screening also needs to be performed, considering the possibility of other congenital abnormalities in different body parts.

There were several limitations in this report. First, the pre-operative size of hematocolpos was not measured in this report, whereas the data could be important to determine the minimum size or volume of blood accumulation that can cause urinary and gastrointestinal obstruction. Second, the follow-up data in this case was not conducted over a long period, whereas there could be a possibility of post-operative obstruction recurrence of cases with no post-operative dilatator.

Conclusion

Imperforate hymen is a condition that needs attention and consideration as a differential diagnosis in pre-menarche or menarche-age children with complaints in the urinary tract or intestines, as well as post-menarche-age children with complaints of primary amenorrhea, especially if both are accompanied by cyclic abdominal pain. Physical examination of

the genitals is simple yet often forgotten, whereas it can be crucial for diagnosis. Check for anatomical abnormalities and congenital screening at an earlier age to prevent a late diagnosis and further complications.

Abbreviations

VU, vesica urinaria; AMH, anti-Mullerian hormone; SRY, sex-determining region Y.

Disclosure

The author(s) report no conflicts of interest in this work.

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