

CASE REPORT

Haematocolpos and Secondary Hematometra Due to Imperforate Hymen: Diagnosis and Treatment

Hematokolpos dan Hematometra Sekunder Akibat Himen Imperforata: Diagnosis dan Tata Laksana

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ABSTRACT

Background

Imperforate hymen is an uncommon congenital anomaly of the female genital tract and is rarely diagnosed in newborns. An imperforate hymen is a cause of primary amenorrhea.

Case Description

A 12-year-old girl was admitted to the pediatric outpatient clinic with cyclic abdominal pain for eight days. She had lower abdominal pain with dysuria and constipation for the previous six weeks. There is a bulging mass on the perineal that appears when straining. Fever, nausea, vomitus, abdominal enlargement, and leucorrhea were denied; breast enlargement since 12 years old with no history of menarche. The patient vital signs were normal. On physical examination obtained, lower abdominal pain and tender suprapubic mass were palpable. The pubertal status was normally secondary sexual characteristics. An interlabial mass was found protruding from the introitus vagina and the hymen intact on inspection of the external genitalia. Laboratory and urinalyses revealed normal. Hematometra and hematocolpos were found on abdominal ultrasonography, which led to the diagnosis of imperforate hymen. Therefore, a hymenectomy was performed and evacuated 520 ml of dark red blood. The patient's condition was good, and she had regular menstruation.

Conclusions

Imperforate hymen is an easy diagnosis, but sometimes clinicians are oblivious to including this in the differential diagnosis of lower abdominal pain in pubescent girls with primary amenorrhea. Also, a lack of awareness about the importance of external genitalia examination as part of routine physical examination in children and adolescents makes this diagnosis easily missed or delayed.

Keywords: Imperforate hymen; primary amenorrhea; cyclic lower abdominal pain; hematocolpos, hematometra

ABSTRAK

Latar Belakang

Himen imperforata merupakan kelainan kongenital genital perempuan yang langka dan jarang didiagnosis pada masa neonatal. Himen imperforata menjadi penyebab amenorhea primer.

Deskripsi Kasus

Anak perempuan berusia 14 tahun datang ke Poliklinik Umum Ilmu Kesehatan Anak (IKA) dengan keluhan nyeri perut yang berulang sejak 8 hari yang lalu. Nyeri perut bagian bawah disertai nyeri saat buang air kecil dan kesulitan buang air besar sejak 6 minggu lalu. Tampak benjolan di daerah kemaluan bila mencedakan. Demam, mual, muntah, pembengkakan perut, keputihan disangkal. Payudara pasien mulai tumbuh sejak usia 12 tahun namun belum pernah menstruasi. Pemeriksaan tanda vital dalam batas normal. Pemeriksaan fisik terdapat nyeri tekan perut bagian bawah dan massa suprasimfis. Status pubertas anak menggambarkan karakteristik seks sekunder normal. Pada genitalia eksterna terlihat massa interlabial yang menonjol dan himen yang tidak mempunyai lubang. Hasil pemeriksaan laboratorium dan urin dalam batas normal. Pemeriksaan ultrasonografi abdomen menunjukkan hematometra dan hematokolpos sehingga didiagnosis himen imperforata. Eksisi jaringan himen segera dilakukan dan terdapat darah berwarna merah kehitaman sebanyak 520 cc. Kondisi pasien membaik dan menstruasi teratur.

Kesimpulan

Diagnosis himen imperforata sangat mudah, namun sering terlupakan oleh klinisi sebagai salah satu diagnosis banding untuk pasien remaja perempuan dengan keluhan nyeri perut bawah dan amenorea primer. Kurangnya kesadaran pentingnya pemeriksaan genitalia eksternal sebagai pemeriksaan rutin pada pasien remaja perempuan membuat kesalahan dan keterlambatan diagnosis.

Kata Kunci: Himen imperforata, amenorea primer, nyeri perut bawah berulang, hematokolpos, hematometra.

INTRODUCTION

Imperforate hymen is a congenital disorder of the female genitalia caused by the failure of the final process of recanalization of the vagina during the embryonic period so that the connective tissue that forms the hymen does not have holes.^{1,3} Embryologically, the hymen originates from the endoderm of the urogenital sinus epithelium in the form of a thin mucous membrane that perforates during the embryonic period to connect the vaginal lumen and the vestibule. If the hymen is imperforated, total obstruction of the vaginal introitus occurs due to the accumulation of secretions in the vagina stimulated by maternal estrogen.² In addition to congenital malformations, the imperforate hymen can occur due to scar tissue due to genital trauma during sexual abuse or infection.^{4,5}

The incidence of imperforate hymen is very rare, ranging from 0.05–0.1% and most cases diagnosed in adolescence are rarely diagnosed in the neonatal period.² Some young adolescents are asymptomatic until menstruation begins, namely symptoms of lower abdominal pain and urinary retention.¹ In neonates, there is a case report of spontaneous rupture of the imperforate hymen at 4 and 7 days of age.⁶

Imperforate hymen is often overlooked as a differential diagnosis of complaints of lower abdominal pain in young girls and lower transverse vaginal septal anomaly.^{1,2,7} Imperforate hymen is easily diagnosed by careful external genital examination and confirmed with a cystic mass in the

pelvic ultrasonography of the abdomen.⁸ Delay in detection and diagnosis will increase the morbidity rate of infection, endometriosis, subfertility, hydronephrosis and kidney failure.

The best management in cases of imperforate hymen is in the incision and excision of the hymen. A good surgical technique can prevent recurrence or hole closure.^{1,8} The prognosis, in this case, depends on when it was detected, the presence of infection, surgical technique and accompanying anomalies.¹ This case report describes the case of a 14-year-old girl with a diagnosis of hematocolpos and secondary hematometra to an imperforate hymen presenting with the chief complaint of recurrent lower abdominal pain.

CASE DESCRIPTION

A 14-year-old girl came to the Pediatrics General Polyclinic (IKA) Cipto Mangunkusumo Hospital (RSCM) with complaints of severe abdominal pain eight days before coming to the hospital. Abdominal pain has been felt since one and a half months ago, accompanied by painful urination (b.a.k) and difficulty defecating (b.a.b). On the day the patient came to the polyclinic, there was lower abdominal pain that did not radiate and b.a.k and b.a.b pain. The patient notices a lump in the genital area when straining. The patient did not have fever, nausea, vomiting, abdominal swelling, vaginal discharge or bleeding. The patient had never menstruated, her breasts began to grow at the age of 12, and the patient's genitals and armpits had grown hair. The patient's mother and older sister experienced their first menstruation at the age of 12 years. The patient's growth and development seem normal.

Upon arrival, the patient's consciousness is fully conscious (*compos mentis*). His pulse is 96 beats per minute, respiratory rate is 20 times per minute, and temperature is 36°C. Clinically and anthropometrically (CDC-NCHS 2000 curve), the impression of malnutrition was found. Genetic potential height between 136 and 153 cm with a mid-parental height of 154 cm. Puberty status of the Tanner A3M3P3 patient.

Abdominal examination showed a palpable suprasymphysis mass of 10 x 12 cm and cystic consistency; periumbilical tenderness in the right and left hypochondrium. On the external genitalia visible mass (Figure 1). Routine blood tests and urinalysis within normal limits. Abdomen ultrasound examination according to hematocolpos and hematometra *et causa* hymen imperforate.



Figure 1. A bulging interlabial mass on the external genitalia is pink in color with a soft consistency, and no pubic opening is found

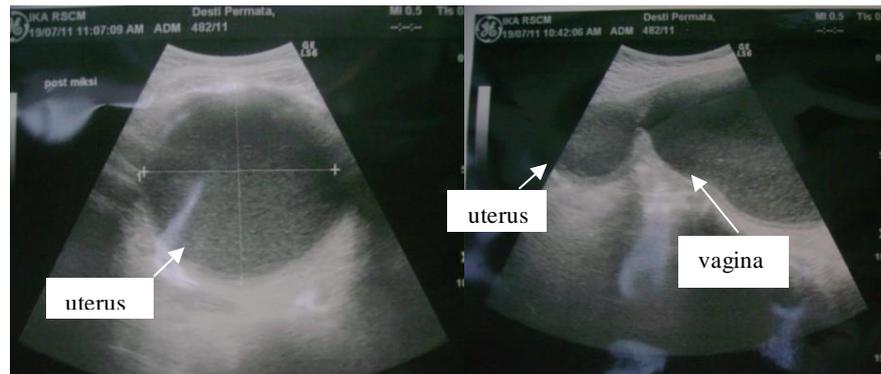


Figure 2. Transverse (left) and sagittal (right) abdominal ultrasound section. Hypoechoic mass in the inferior bladder is 14.2 x 8.5 x 8.8 cm. There is a second, smaller, uterine-shaped mass superior to the first. The vaginal cavity is filled with fluid, measuring 13.23 x 6.89 cm. Both ovaries are within normal limits.

Based on the examination, the diagnosis of imperforate hymen with hematocolpos and hematometra was established. The patient was then consulted by the Department of Obstetrics and Gynecology to have a hymen excision carried out under general anesthesia. Approximately 520 cc of red-black blood was obtained during the evacuation of hematocolpos and hematometra. The patient's condition after the procedure was good, so the patient was sent home 5 hours after the procedure was finished with antibiotics and oral analgesics.

One week after the procedure, control patients found a hymenal patent hole, dry excision wound, and no signs of inflammation or infection. Vaginal bleeding stopped on the sixth day after the procedure. The patient menstruates regularly on subsequent monitoring.

DISCUSSION

Imperforate hymen is the most common obstructive disorder of the female genital tract, although the overall incidence is small, ranging from 0.05% to 0.1%. In infancy, the imperforate hymen manifests as a protrusion of the introitus.^{9,10} This abnormality can be detected easily at any age by inspection of the external genitalia but is often only detected at the onset of menstruation at the age of 12-15 years. Accumulation of menstrual blood in the vagina (haematocolpos), uterus (hematometra) and fallopian tubes (hematosalping) cause frequent misdiagnosis as an intrapelvic mass.^{2,11-13} In this case, it was only diagnosed when there was already a hematocolpos and hematometra, which caused recurrent complaints of pain in the abdomen. The lower part, difficulty urinating and constipation, suprasymphysis masses and not menstruating despite normal pubertal development marked secondary sexual maturation according to Tanner 3. Non-specific symptoms, such as nausea, vomiting or diarrhea, can also be found.³ Hematocolpos and hematometra cause mechanical pressure on the urethra, bladder, digestive tract, and pelvic blood vessels resulting in symptoms of urinary retention, constipation, or leg edema.¹ Table 1 describes some clinical manifestations of the imperforate hymen during puberty.

The diagnosis of imperforate hymen is easily enforced by inspecting the pathognomonic signs in the external genitalia; namely, there is a protrusion of the bluish membrane caused by pink hematocolpos if the hymen is thick enough.^{1,2} The diagnosis of the imperforate hymen can be easily missed because genital examinations are not routinely carried out; therefore, a high level of accuracy is needed when dealing with patients like this.^{4,7} Menarche is an advanced stage in puberty, and most girls have reached secondary sexual maturation, according to Tanner 3 at menarche. If there is a gap between the time of secondary sexual maturation and the absence of menarche, one of the causes to be suspected is imperforate hymen.^{3,7}

This case was not familial, and no other congenital abnormalities were found. Other anomalies that may accompany it include polydactyly, bifid clitoris, ureteral duplication, ectopic ureters, hypoplastic kidneys, multicystic dysplastic kidneys, urethral membranes, imperforate anus and low-lying anorectal anomalies.^{10,11,14-16}

Table 1. Clinical manifestations of imperforate hymen at puberty ¹³

Recurrent abdominal pain Primary amenorrhea Abdominal mass Acute urinary retention Constipation Peritonitis Acute abdomen Bilateral hydronephrosis Back pain Bladder perforation

Differential diagnosis of primary amenorrhoea, lower abdominal pain, and pelvic mass in a pubertal girl between the transverse vaginal septum, longitudinal vaginal septum, and vaginal agenesis.¹ Imperforate hymen can be differentiated from a low-lying transverse vaginal septum by the Valsalva maneuver.⁸ In vaginal agenesis, there is a normal hymenal structure with vaginal dimples, and no uterus is visible on ultrasound. High-lying vaginal atresia and transverse septum are almost always accompanied by other gastrointestinal or urogenital anomalies, whereas other congenital anomalies rarely accompany imperforate hymen and low-lying septum.¹⁰ If there is any doubt about the diagnosis, an ultrasound can be performed for confirmation. Ultrasound examination of the abdomen showing a large, fluid-filled cystic mass, and vaginal and uterine dilatation due to fluid accumulation, are findings that confirm the clinical diagnosis of a hematocolpometra.^{15,17,18}

Management of imperforate hymen is by incision of the hymen, drainage of accumulated blood behind the hymen, and excision of hymenal tissue to form a patent outlet.^{1,2,7,19} Hematocolpos puncture without definitive corrective action should be avoided because thick fluid may not be removed through a small hole, and there is an increased risk of ascending infection, which can cause pelvic inflammatory disease or tubo-ovarian abscess.^{17,21} In this case, hymenal excision was performed for drainage of hematocolpos and hematometra. Early diagnosis and immediate corrective action to remove accumulated blood can prevent endometriosis and maintain fertility.¹⁶ Complications of hematocolpos include endometriosis, bilateral hematosalping,

impaired ciliary activity in fallopian tube cells, and ruptured hematosalpinx, which puts the patient at risk for fertility problems and an increased risk of ectopic pregnancy in future.^{1,2,16}

The prognosis of patients with imperforate hymen after corrective action is generally good.^{2,7} Several studies have shown an improvement in the quality of life after corrective action. The study reported good sexual function after corrective action; even some research subjects became pregnant and gave birth to healthy children.^{11,14} The condition of these patients after the action was good. The prognosis of *quo ad vitam*, *quo ad functionam*, and *quo ad sanctionam* for this patient is *bonam*.^{2,20}

Imperforate hymen is important to be diagnosed as early as possible to avoid complications that can occur due to late diagnosis. Clinicians can prevent delays in diagnosis, misdiagnosis, and the resulting morbidity by examining the external genitalia as part of their daily clinical practice.^{21,22} Evaluation of the patient's family members is important because some cases of the imperforate hymen are familial.^{2,20}

The American Academy of Pediatrics (AAP) recommends examining the external genitalia as part of a comprehensive physical examination of all infants and adolescents at routine health visits.^{16,21,23} Examination of the external genitalia is performed to ensure that there is normal genitalia anatomy, to assess pubertal status, and an ultrasound examination of the abdomen to look for abnormal lesions, infection or trauma. In addition, the hymen was assessed for patency and configuration, including the presence of an imperforate hymen. Adolescents with imperforate hymen should be immediately referred to an obstetrics and gynaecology specialist to prevent blood accumulation in the vagina and upper genital tract.^{22,24}

CONCLUSION

This case demonstration aims to remind clinicians of the importance of examining the external genitalia as part of the routine physical examination in adolescent girls. By carrying out a thorough physical examination, including examination of the genitalia, delays in diagnosis and complications can be prevented. In addition, Hematocolpos and hematometra due to imperforate hymen should be considered a differential diagnosis for female adolescents who present with complaints of lower abdominal pain and experience primary amenorrhoea.

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CONFLICT OF INTEREST

There is no internal conflict from the author either financially or institutionally.

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