

Pseudoacute abdomen in female caused by haematometrocolpos and haematosalpinx because hymen imperforatus: diagnosis and therapy of imperforate hymen

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Received 12 December 2006; accepted 22 April 2007

Abstract: We present a case of atresia hymenalis in a 12.5 year old female who was admitted to gynecological emergency with a history of severe lower abdominal pains over the course of a few months. Provided are clinical pictures of the “Pseudoacute abdomen”. Upon clinical examination, we found blue, shiny, bulging and imperforated hymen. An abdominal mass was palpated in the vicinity of the navel. Transabdominal ultrasound demonstrated mass filled with thick liquid. “Mercedes hymenal incision” was performed in general anaesthesia. About 1.2 litre of dark, thick blood was removed. Postoperative course was uneventful. The two years of follow-up were characterized with regular psychomotoric development for the age with regular menstrual cycles.

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Keywords: Imperforate hymen, haematocolpos, haematometra, haematosalpinges; pseudoacute abdomen, hymenal incision

1 Introduction

The genital tract develops during embriogenesis, from 3 weeks gestation to the second trimester. The hymen is derived from the epithelium of the urogenital sinus, and during

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the fetal stage it represents the distal part of vagina. By the fifth month of gestation, the canalization of the vagina is complete. An imperforate hymen is usually the result of the urogenital sinus not canalizing. Histologically, the hymen is defined as a connective tissue membrane covered by stratified, squamous epithelium. The normal hymen is described as fairly thin, elastic and translucent. In pathologic conditions, it becomes thickened, fibrotic and nonelastic. A highly placed transversal septum is often found, and it is important to distinguish imperforate hymen from other anomalies. Variants in hymenal configuration are often seen, varying from hymen semilunaris, annularis, cribriformis, septus, fimbriatus, navicularis to atresia hymenalis (hymen imperforatus) [1].

Hymen imperforatus or atresia hymenalis is at the extreme of a spectrum of variations in hymenal configuration. Anomalies of the vagina, hymen, and uterus are not accompanied by abnormalities of ovarian development. Endocrinologic function is without abnormality, leading to expected pubertal changes [1, 2].

We present a case of atresia hymenalis in a 12.5 year old female with clinical pictures of the “pseudoacute abdomen” caused by haematocolpos, haematometra and haematosalpinges.

2 Case report

The 12.5 year old female patient was admitted in gynecological emergency presenting with severe abdominal pains and enlargement of the abdomen over a course of a few months. The recurrent pains were extremely strong on the day of admission and moderate for a few days prior to admission. She had defecated on the previous day. Patient experienced frequent urination with pain. The patient was from her mother's third pregnancy. She was thin. The signs of puberty after the Tanners classification were telarche 2, pubarche 2, no menarche; weight 46 kg, height 157 cm.

Upon examination, we found a mass mainly located in the lower part of the abdomen. The mass was elastic, tense and very painful. Pediatrics general examination: normal psychophysical condition, normal pubertal hormonal status (FSH, LH, PRL, E2) without urological anomaly (detected with ultrasound and intravenous urography).

Gynecologic examination showed a blue, shiny, bulging and imperforate hymen. A transabdominal pelvic ultrasound demonstrated a mass between navel and pelvis filled with thick liquid. It suggested that the diagnosis was haematocolpometra and haematosalpinges.

There was no fluid in the abdominal cavity. After the routine laboratory examination (haemogram, urinogram, CRP is normal) a “mercedes” incision was performed. About 1.2 litre of dark, thick blood was removed.

The vaginal epithelium was sutured to the hymenal ring. The tumor mass was getting smaller. Antibiotics were given prophylactically (cefuroximaxetil 2×250 mg for 7 day period). The recovery was uneventful. Control ultrasound examination was performed five days later and showed some residual liquid in left tube. The uterus was found normal without sactosalpinges.

Two years later the patient had regular psychomotoric development for her age, regular menstrual cycles and bleeding with normal ultrasound findings.

3 Discussion

Imperforate hymen is one of the simplest and most common anomalies in the female genital organs, occurring in 0,1% of girls born at term [1]. Families with more than one individual affected by imperforate hymen have been described. No common mode of inheritance has been identified. It is not known whether it is an autosomal or an x-linked transmission.

Almost 350 years have passed since Pare first described haematocolpos as a complication of imperforate hymen. About 300 references over the past hundred years described a case similar to ours [1–3]. The literature includes examples of incorrect and late diagnosis. One, a 13,5-year old girl, was subjected to two operations, the first a bilateral tubectomy followed by a hysterectomy, before the correct diagnosis was made [1].

In spite of the recommendations for early inspection of the external genitalia in newborns, variations in hymenal anatomy can escape diagnosis until the time of menarche. Occasionally, a presence of mucocolpos, abdominal mass with urinary retention has been described in infancy and childhood. A surgical technique to correct such a defect has been described. However, in the asymptomatic patient, waiting until puberty is suggested before deciding whether such procedure is necessary [1, 2]. In prepuberty, the imperforate hymen may result in hydrocolpos, mucocolpos and pyocolpos with other complications: urinary retention, hydroureter and hydronephrosis with or without clinical pictures of “pseudoacute abdomen” [6, 7]. Large amounts of retained liquid have been reported. If the condition remains undetected, menarche can trigger the problem.

The reported quantities are very variable, and the maximum quantity reported in the literature is 3000 ml. The typical reason for consulting a doctor is for low abdominal pain, often by monthly attacks. The recurrent pain represents a female’s menstrual periods, which are the origin of the development of haematocolpos. Frequently, vaginal and rectal pressure is present. Severe constipation, low-back pain and urinary retention are described as presenting symptoms. The lower back pain may cause painful defecation. The haematocolpos irritates the sacral plexus, radiating to the lower extremities. Acute urinary retention is another symptom that can be the first sign of complications leading to a diagnosis of imperforate hymen. Frequently, acute urine retention and dilation of the genital organs create a “tumor” that can be palpated in the vicinity of the navel and clinical pictures of the “pseudoacute abdomen” syndrome, without need for the laparotomy [1–8]. Imperforate hymen complicated with ruptured haematosalpinx with acute abdomen is a rare occurrence [7, 8].

Diagnosis includes history of illness, inspection, clinical examination and pelvic and abdominal ultrasound. MRI is reserved for evaluation of questionable anatomy or the possibility of abnormalities [1–4]. Inspection of the external genitalia shows a thin, bluish, translucent membrane that bulges with Valsalva maneuver just inferior to the urethral

meatus. A pelvic or abdominal mass in the vicinity of the navel can be palpable on abdominal or rectal examination. The findings for diagnosis include a large collection of blood within the uterus (haematometra) and an even larger collection of blood within the vagina (haematocolpos). Additional findings may include blood-filled fallopian tubes (haematosalpinges) and signs of retrograde menses, occasionally to the point of the development of intraabdominal endometriosis and severe adhesions. Pelvic ultrasound via the transabdominal, transperineal, or transrectal route is indicated as the initial diagnostic test. MRI follows if any question about the anatomy exists [1].

The differential diagnosis includes many conditions, some rare and others relatively common like labial adhesions. A cribriform hymen or a hymenal band can lead to incomplete hymenal obstruction. An alternative to surgical correction involves digital dilation using a topical anesthetic cream. Obstructing longitudinal or transverse septa require careful preoperative evaluation to define the anatomy. Medical therapy has no place in the management of an imperforate hymen. The timing of surgical therapy is based on the presence of symptoms. A symptomatic mucocele presenting in the neonatal period should be managed expediently. If an asymptomatic imperforate hymen without mucocele is diagnosed during childhood, it can be managed during puberty and prior to the development of a haematometra or haematocolpos. The presence of estrogen stimulation in puberty facilitates the surgical repair and healing.

The surgical treatment has not differed much during the past 50 years. It is usually described as hymenotomy or hymenal incision. The objective of the hymenotomy is to open the hymenal membrane to leave a normally patent vaginal orifice that does not scar. The hymenal orifice is enlarged using a circular incision following the lines of the normal annular hymenal configuration. An alternative is a cruciate incision along the diagonal diameters of the hymen, rather than anterior to posterior, that avoids injury to the urethra. An excess hymenal tissue is removed. The vaginal epithelium is sutured to the hymenal ring using interrupted stitches with fine absorbable suture. If a large hematocolpos is present, the surgeon has to evacuate blood using a suction tube. Small variations in technique are described, such as the use of the laser or electrosurgery rather than the scalpel. Concurrent diagnostic laparoscopy may be performed to allow lysis of adhesions and excision or cautery of endometriosis, as well as lavage of retrograde blood in the pelvic cavity. Infectious complications are rare, and prophylactic antibiotics are not required. Nonsteroidal anti-inflammatory drugs may be prescribed for the cramping. Topical lidocaine jelly is recommended for the vaginal orifice [1–3].

Clinical outcome and prognosis are good. Injury to the urethra, rectum or bladder is possible if the anatomic defect is not defined clearly and if the actual condition is vaginal agenesis or complicated muellerian abnormality, rather than a simple imperforate hymen.

Excessive bleeding may be seen after incision without suture. Sterility is certainly the greatest problem with both the tubes involved [1–5].

The retrograde menses and endometriosis associated with an obstructed outflow tract behave in a more benign manner than spontaneously occurring endometriosis without obstruction, although this assertion is based on clinical experience rather than outcome

reports or evidence [1, 3]. Accurate diagnosis together with adequate treatment may reduce the need for the re-operations and infertility (long-term fecundity) in cases with operate obstructing vaginal malformations [5].

The main causes of the development of the “pseudoacute abdomen” syndrome were distinguished: fracture or contusion of the bones, contusion of the abdominal wall and organs of the abdominal cavity and organomegaly, influenza, segmental myocardial infarction, exacerbation of chronic disease of the abdominal cavity, injury to the organs adjacent to the diaphragm and other.

Imperforate hymen usually escapes diagnosis until the time of menarche, when the young female presents with excessive symptoms and changes on external and internal genitalia. This morbidity can be avoided if clinicians (obstetricians, pediatricians, gynecologists, family physicians) are trained to examine the genitalia of newborns and young children. Timing of the surgical correction should be planned appropriately. The future should hold a better prognosis for young girls if their gynecologic health and fertility is followed from the beginning of their lives.

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