AN UNUSUAL PRESENTATION OF IMPERFORATE HYMEN

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ABSTRACT

Imperforate hymen is a congenital anomaly that is rarely seen in female genital system. Imperforate hymen is reported in only 0.1% of all female live births. It is thought to be caused by a congenital defect in the degeneration of the central hymen cells. Although it is generally an isolated anomaly, sometimes other associated genitourinary anomalies may accompany this defect. Patients with this anomaly are generally asymptomatic until puberty and in the period of adolescence, where the patients present with primary amenorrhoea, cyclic pelvic pain and or suprapubic mass. History and pelvic examination are the main steps in the diagnosis of the condition, while imaging modalities such as ultrasonography are useful in the determination of other possible associated congenital anomalies. The treatment for this anomaly consists of vaginal reconstructive surgery called hymenectomy. A case of imperforate hymen in an adult is hereby presented, with a review of the literature.

Key words: Adult, imperforate, hymen.

INTRODUCTION

At menarche every female is expected to menstruate monthly. However the presence of an imperforate hymen prevents the discharge of menstrual fluid and result in progressive distension of the vagina with altered blood (haematocolpos) and later of the uterine cavity, known as haematometocolpos or haematometria.^{1, 2} The accumulated blood remains fluid but concentrates and darkens in colour over the time, owing to the absorption of water.³ This condition is seen among teenage females with the peak incidence occurring between the ages of 14 and 15 years.^{4, 5} The vagina and the uterus, which are distensible, can accommodate a large volume of fluid, so that symptoms may be delayed into adulthood.⁶

Ultra-sonography is a valuable modality in the diagnosis and

the assessment of the extent of involvement of the vagina, the cervical canal, the uterine cavity and sometimes, the fallopian tubes.⁷ Other associated congenital anomalies such as renal atresia or ectopia and polycystic kidneys have also been evaluated by the use of ultrasound.⁵

CASE REPORT:

A.A is a 22 year old unmarried female who presented to the hospital with a complaint of abdominal mass which was first noted three years earlier, but has progressively increased in size. There was absence of menstrual discharge, though, she admitted to having monthly cyclical lower abdominal pains in the last 8 years. There was no history of lower back pain, sciatica, constipation or vomiting, but there was increased frequency of micturition. Physical examination revealed a young

woman with normal female features. She had a normal external female carrying angle at the elbow joint (an angle formed by the long axis of the arm and that of the forearm). There was a palpable, mildly tender suprapubic mass arising from the pelvic cavity, with its fundus at the level of the umbilicus. Perineal examination revealed normal external genitalia and anal opening. A smooth, diaphragm-like mass protruded within the introitus which was more when the patient strained. A provisional diagnosis of imperforate hymen was made.

Transabdominal ultrasound revealed a well-defined centrally located mass containing fairly homogenous internal low echoes. It was estimated to have dimensions of 15x17x9cm in its A-P diameter, length and transverse diameters. No septations or

central vascularity was seen (figure 1). Both ovaries were clearly identified separately and did not show any abnormalities. The kidneys were also within normal limits. Appearances were consistent with a dilated fluid-filled uterus. Diagnosis of an imperforate hymen was confirmed. The excision of a transverse vaginal septum was performed and about 1.5 litres of dark coloured blood was drained. The patient was discharged home on the fourth post-operative day. She has subsequently been experiencing monthly menstrual flow.

Figure 1: Sonograph of the pelvis showing a dilated uterine body (right mass). The dilated uterus is seen on the left. DIL. RT= dilated right uterine tube.

Figure 2: Sonograph of the pelvis showing a dilated uteruscavity in longitudinal (right) and transverse (left) scans. UB=Uninary Bladder.

DISCUSSION

Imperforate hymen is a rare

diagnosis of blood collection in the vaginal and uterine cavities due to vaginal out flow obstruction, has been known since antiquity.⁸ About 400 years ago, Ambrose Pare described a case in which, because of primary amenorrhoea, pain and abdominal swelling, the condition was diagnosed as pregnancy.⁹ The first documented report of this condition in the English literature was in 1856.⁹ This condition is found to be uncommon, with a frequency of 1 in 70,000 female births.¹⁰ A transverse vaginal septum is the main feature that is seen, which results from overgrowth of tissues at the opening of the vaginal outgrowth into the urogenital sinus.^{11, 12} It has also been suggested that imperforate hymen is inherited as a simple autosomal recessive disorder but other workers have not been able to confirm this due to the rarity of the condition.^{4, 13}

It is stated that imperforation result from overgrowth of tissues at the opening of the vaginal up growth into the urogenital sinus.¹⁰ Ekenze¹² suggested that imperforate hymen is inherited as a simple autosomal recessive disorder; however, other workers have not been able to confirm this claim due to the rarity of the condition.^{4, 13}

Imperforate hymen occurs in approximately 0.1% of female newborns,¹³ and will either lead to hydrocolpos and hydrometrocolpos in the young or, as haematocolpos and haematometocolpos in teenagers and young adults.^{14, 15} Many cases of imperforate

hymen escape detection as hydrocolpos, to be detected later as haematocolpos, the latter being much more frequently encountered.¹⁴

The clinical history and findings of an imperforate hymen are characteristic in the adolescent. These include primary amenorrhoea despite normal physical female development, history of monthly cyclical lower abdominal pains and progressively enlarging suprapubic mass. Other features include low back pain, frequency of urination and/or acute urinary retention.⁵ This patient's presenting complaint was the suprapubic mass while ultrasound confirmed the diagnosis. It was also easy to assess the extent of involvement of the uterus.

Ultrasound and CT/MRI are diagnostic modalities of choice and can reveal other associated orifices. A distended vagina is identified ultrasonographically by its midline position, which is posterior to the urinary bladder and also by its elongated shape. Haematocolpos has a transonic appearance with posterior echo enhancement. A fine diffuse echopattern may be noted due to the altered blood content. The echogenicity of the uterine collection is not affected by the age at presentation, provided the history of the cyclical pain has been established.⁵ Hymenal diaphragms may be identified by ultrasound.⁷

Imperforate hymen may be seen in isolation, but may be accompanied by complete

duplication of the genital tract namely, the vagina, uterus and a bifid clitoris.¹ Other abnormalities are urethral stenosis, double ureter, renal atresia, renal ectopia, polycystic kidneys, imperforate anus and tracheo-oesophageal fistula.⁴ Chondrodystrophy with polydactyly (Ellis van Creveld Syndrome) has also been associated with imperforate hymen.¹⁶ Our patient's case is an isolated occurrence.

Lack of adequate awareness among physicians and the relative rarity of the condition has resulted in error of diagnosis. Late onset of sexual development, ability of vagina and uterus to expand, normal looking vaginal orifice in mid and high vaginal septum and limited access to expert medical facilities in the developing world can also delay the presentation as seen in this case.

CONCLUSION

In conclusion therefore, imperforate hymen must be suspected in the adolescent and adult females who have never menstruated and who complain of cyclical lower abdominal and low back pain or acute urinary retention. Since ultrasound as an imaging modality is now readily available, it should be employed in this category of patients, in addition to the findings on physical examination.

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