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Case Report

Obstructive mullerian anomalies: a case series

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ABSTRACT

AIM of the study was to review the heterogeneous clinical presentations and management options for some of the obstructive mullerian anomalies through a case series. Background: Müllerian duct anomalies (MDAs) are a miscellaneous group of entities that result from the non-development, defective vertical or lateral fusion, or resorption failure of the müllerian ducts due to genetic mutation. 5 cases of obstructive mullerian anomaly are reviewed. Cases of OHVIRA (obstructed Hemivagina with Renal Agenesis) syndrome, transverse vaginal septum, imperforate hymen and obstructed rudimentary horn of unicornuate uterus are included. Results: We found that cyclical abdominal pain was the most common presenting complaint. There is a high incidence of associated renal anomalies. Psychosocial counselling before treatment is necessary to address the functional and emotional aspects of the patient. Surgical management was done in all patients with good postoperative outcome. Conclusion: Obstructive mullerian anomalies need to be evaluated by a meticulous examination and imaging studies to reach the diagnosis with precision. The treatment has to be tailored to the specific anomaly.

Keywords: Imperforate hymen, Obstructive Mullerian anomalies, OHVIRA Syndrome. Transverse vaginal septum, Unicornuate uterus

INTRODUCTION

Development of female genital tract begins by the fusion of the müllerian ducts, at approximately the 10th week of gestation. Müllerian duct anomalies (MDAs) are a miscellaneous group of entities that result from the nondevelopment, defective vertical or lateral fusion, or resorption failure of the müllerian (paramesonephric) ducts due to genetic mutation, developmental arrest or acquired defects. Though incidence rates of müllerian duct anomalies (MDAs) vary widely most authors report incidences of 0.1-3.5%.^{1,2}

Most müllerian duct anomalies (MDAs) are associated with functioning ovaries and age appropriate external genitalia. Some of the anomalies may be asymptomatic

and are diagnosed incidentally. Obstructive anomalies however prevent normal menstruation and may lead to hematocolpos, hematometra, retrograde menstruation, endometriosis and pelvic adhesions.³

These abnormalities often present with primary amenorrhea, obstructed menstruation, severe dysmenorrhea or dyspareunia.

A high index of suspicion is necessary to diagnose these disorders and an adequate workup is quintessential before planning the treatment. The present case series attempts to highlight the varied clinical presentations, associated congenital anomalies and the optimum mode of management of some of the obstructive müllerian anomalies.

CASE REPORT

A 14-year-old unmarried girl reported to gynecology OPD with complaint of irregular bleeding per vaginum for 2 years. She gave history of severe abdominal pain during menstruation. She attained menarche at the age of 12 years. Breast and pubic hair showed tanner stage 4 development. Menstrual cycle was for 8-9 days and recurred every 28-30 days. Her general physical examination was normal.

Case 1

On local examination, abdomen was soft and there was no organomegaly. A mass could be felt in vagina on per rectal examination. Ultrasound revealed bicornuate uterus with a mass of 5.7*2.9*2.8 cm in vaginal region with internal echoes suggestive of haematocolpos, normal bilateral ovaries and clear pouch of douglas with normal kidneys. MRI revealed uterine didelphys and, hemicervix was terminating into hematocolpos of size 41*36*26 mm. The dilated right hemivagina was drained and the septum was excised followed by reanastomosis of the vaginal edges (Figure 1). The right cervix was seen above the level of the obstruction. Postoperatively a diagnosis of OHVIRA (obstructed Hemivagina with Renal Agenesis) syndrome was made. The patient is under regular follow up.



Figure 1: Obstructed hemivagina after drainage.

Case 2

A 12-year-old girl was admitted in gynecology ward with c/o cyclical pain for 4-5 days for 3 months. She did not have any history of galactorrhea, headaches, visual disturbances, hirsutism or acne. On examination external genitalia appeared to be normal. The patient had blind vaginal pouch. On per rectal examination, a cystic mass was felt in the vagina. Ultrasound revealed a heteroechoic collection of size 5.4*4.4 cm in the endometrial cavity. MRI of pelvis revealed a normal sized uterus with a normal myometrium. A well-defined collection of 6*4.4 cm was noted in vagina. Both the ovaries and kidneys were normal. Patient was operated for excision of the transverse vaginal septum followed by reanastomosis of

the vaginal edges. Postoperatively, patient was motivated to do regular vaginal dilatation. For this a soft mould was made by using condoms over autoclaved foam and secured with silk suture and later a five-cc syringe with cut edges was used at the centre of the foam to make a hard mould. The patient needed repeat dilation under anesthesia one-month post-surgery due to inappropriate self-dilatation. However, with reassurance and counselling, compliance improved, and the patient had an uneventful course thereafter till her last visit nine months after the procedure.

Case 3

Similar to the previous case, a 13-year-old girl was admitted in gynaecology ward with c/o cyclical pain for 4-5 days for 5 months. Her Examination and Investigations led to a provisional diagnosis of transverse vaginal septum. Patient was operated for excision of vaginal septum. However due to thickness of the septum, reanastomosis of the vaginal edges could not be done and a split thickened skin graft was used for vaginoplasty. The patient had an uneventful postoperative course and is under regular follow up.

Case 4

A 13-year-old girl presented with c/o cyclical lower abdominal pain for 2-3 months. Secondary sex characters were well developed. On examination a firm, non-tender pelvic mass of size 18 weeks size with restricted mobility was felt per abdomen. Imperforate hymen with a bulge was seen on local examination of vagina (Figure 2) Ultrasound revealed a collection of 11*5.7 cm in endometrial cavity. Both ovaries and kidneys were normal.

With the provisional diagnosis of imperforate hymen, patient was treated surgically. A cruciate incision was given on the centre of bulge. Drainage of hematometra was done followed by eversion of margins. Postoperatively the patient is having regular menstrual cycles.



Figure 2: Imperforate hymen.

Case 5

A 28-year-old nulligravid woman presented to the outpatient department with severe abdominal pain. Her last menstrual cycle was 10 days back. She attained menarche at the age of 14 years and had severe dysmenorrhea since then. Her cycles lasted for 4 to 5 days and recurred at regular intervals of 28 to 30 days. Her pulse rate was 104 per minute and blood pressure was 100 by 70 mm of Hg. Rest of the general physical examination was normal. On per speculum examination, cervix and vagina was normal. A per vaginum examination revealed an anteverted normal sized uterus and a 5*6.5 cm hard tender mobile mass in left fornix for pregnancy test was negative. Ultrasonography showed a homogeneous, solid 5*5.5 cm mass in left adnexa. In view of severe pain, a decision for laparotomy was taken.

Per operatively, uterus was normal in size with right uterine deviation. The right fallopian tube was normal. A 6*7 cm distended non communicating rudimentary horn was attached to the uterus by a fibrous band (Figure 3). Bilateral kidneys were normally placed and there were no signs of endometriosis. The pain due to obstructed rudimentary horn probably caused the tachycardia. The rudimentary horn was excised and histopathological examination showed normal endometrium and myometrium.



Figure 3: Obstructed rudimentary horn.

DISCUSSION

Normal development of the female reproductive tract involves a series of complex interactions that direct differentiation of the müllerian ducts and urogenital sinus (UGS) form the internal female reproductive tract. When an interruption or dysregulation occurs in any of the dynamic processes of differentiation, migration, fusion and canalization, a wide spectrum of müllerian duct anomalies can result.⁴ Of importance is the fact that other defects frequently accompany müllerian abnormalities. The developing kidney and urinary system are closely related to the reproductive tract and their abnormalities are often associated with defects in the reproductive system.

Although many women with müllerian anomalies are asymptomatic, several gynecologic signs and symptoms are associated with specific anomalies. Endometriosis may be cause of infertility. Of all müllerian anomalies, those involving the uterus are most commonly implicated in poor obstetric out-comes like increased rate of recurrent pregnancy loss, pre-term delivery, malpresentations, intra- uterine growth retardation and increased caesarean delivery rates. Imaging has a crucial role in the diagnosis and management of these patients. MRI is considered the gold standard technique for diagnosing müllerian anomalies and is both sensitive and specific⁵. The detailed discussion of all the anomalies is beyond the scope of the present article.

However, the individual case studies are discussed briefly.

Case 1

Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) is a rare congenital urogenital anomaly. The incidence of OHVIRA syndrome is very small and only isolated case reports have been published. Diagnosis of this syndrome is challenging due to presence of nonspecific symptoms. Patients with this anomaly usually present after menarche with cyclical dysmenorrhea, as in the present case later evolves into persistent pelvic pain. Treatment invariably requires surgical intervention in the form of excision of vaginal septum to relieve obstruction. In addition to relief of pain due to obstruction, surgery also reduces chances of pelvic endometriosis due to retrograde menstrual bleeding. Patients are able to have normal sexual life. Some are even able to conceive and carry pregnancy to term. The surgeon must, therefore, make every effort to preserve the obstructed uterus. The right side is affected nearly twice as frequently as the left side.⁷ The present case showed the right-sided involvement as well.

Case 2 and 3

Transverse vaginal septum.

Transverse vaginal septum is a wall of tissue created when the uro -genital sinus and müllerian ducts inside the vagina do not develop correctly. The external genitalia appears normal, but the vagina is shortened and blocked. Symptoms may include amenorrhea, periods that last beyond normal 4 to 7 days cycle and abdominal pain, caused by blood collecting in the upper vagina. The incidence of this anomaly is unknown but may be between 1:2100 and 1:72000.⁸ They are classified according to their location in the vagina (low<3 cm from the introitus mid 3-6 cm and high >6cm), thickness and presence or absence of perforation . Treatment involves surgical resection of the septum with approximation of proximal and distal vagina. If the septum is thin, the edges of vaginal mucosa can be approximated easily as in case 2 but a graft may be required in case of thick septum

as in case 3. As the final decision of using a graft can be conclusively taken intraoperatively only, it is advisable to keep the logistics and personnel (e.g. plastic surgeons) ready to avoid any unnecessary delay.

All such procedures should be followed by regular vaginal dilatation as it helps to maintain vaginal capacity and prevents vaginal stenosis. Vaginal stenosis is a common problem postoperative phase (as was seen in the present case) and can be addressed by regular dilatation as has been shown in an observational study of 46 cases of transverse vaginal septum. Reproductive outcomes are varied and further long-term studies are needed to completely assess the same.⁹ Ideally a mould made of dental material or some of the commercially available moulds may be used. However, the moulds with autoclaved foam are economical and very well accepted by the patients like in the present case where the patient returned to her native village with moulds handed over by us. This ensured compliance and also didn't put any financial burden on the patient.



Figure 4: Vaginal Mould of autoclaved foam.

Case 4

Imperforate Hymen

This condition has an estimated prevalence of 0.1% and usually appears as an isolated finding.¹⁰ Although not an MDA, an imperforate hymen can have clinical and imaging characteristics similar to those of a low transverse vaginal septum. Classical presentation is that of increasing cyclical pain in the absence of menstruation in a visible bulging membrane on gently parting the labia. Treatment is with complete resection the hymen, but resection should not be too close to the vaginal mucosa which may lead to scarring and stenosis.

Case 5

Unicornuate uterus

During embryogenesis, the failure of one mullerian duct to develop and elongate results in a unicornuate uterus. The unicornuate uterus may occur alone, but it is

frequently associated with a rudimentary horn. The American Fertility Society classification divides this group into 4 categories based on the presence or absence of a rudimentary horn.¹¹ Associated urologic anomalies are frequent. Women with noncommunicating, functioning rudimentary horns may present with pelvic pain usually secondary to hematometra or endometriosis. MRI reliably helps in diagnosing such patients. The diagnosis in the present case could be confirmed only intraoperatively as MRI could not be done in the preoperative period due to logistic issues. It is pertinent to mention here that this diagnosis should be kept as a rare possibility in patients undergoing emergency laparotomy due to a provisional diagnosis of torsion or hemorrhage in the adnexal mass as in the present case. Such patients should not be considered for reconstruction metroplasty and excision of the horn is the treatment of choice.¹²

Role of counselling in post-vaginoplasty patients

Besides correction of anatomic defect, management of patients with MDAs includes psychosexual counselling to address the functional and emotional effects of genital anomalies. As patient motivation is an important factor in predicting successful outcome especially when regular vaginal dilation is required, this counselling along with peer group meetings (of similar patients) helps them to deal with it in a better way. This strategy was adopted in one of our patients where postoperative vaginal stenosis was noticed. It also allows adolescent and young women to understand their reproductive potential e.g. in cases of mullerian agenesis.¹³

CONCLUSION

An early correct diagnosis of the Mullerian anomalies is the goal to relieve the symptoms and prevent complications, caused by retrograde menstruation which may result in endometriosis and also, preserve sexual and conception abilities. Effective Management of patients of MDA include, correct diagnosis of the anomaly, evaluation for assessment of concomitant anomalies and psychosocial counselling before treatment to address the functional and emotional concepts.

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