Frequency of Mullerian Duct Abnormalities

Humaira Rani*, Sobia Nawaz**, Nadia Saif*, Asma Tanveer Usmani**

*Department of Obstetrics and Gynaecology, Wah Medical College, Wah cantt,
**Department of Obstetrics and Gynaecology, Benazir Bhutto Hospital, Rawalpindi

Abstract

Background: To determine frequency of most common mullerian duct abnormalities.

Methods: In this interventional study, twenty seven patients were studied in which mullerian duct anomalies were detected. They were selected by convenience sampling. All relevant information was written on a designed proforma. All patients with primary amenorrhoea, primary infertility, dysparunia, aparunia, severe dysmenorrhoea, recurrent pregnancy loss and preterm labour were included, while patients with secondary amenorrhoea and secondary infertility were excluded.

Results: Out of twenty seven, 25 were 13—30 years of age, two patients were 35 and 52 years of age. 51.9% were married, 40.7% unmarried and 7.4% were divorced. Patients having amenorrhoea were 55.6% while 44.4% had regular menstrual cycle. The most common Mullerian duct anomaly detected was blind end vagina with absent uterus (22.2%). Three patients had transverse vaginal septum (11.1%), 3(11.1%) had longitudinal vaginal septum and 3(11.1%) had vaginal agenesis only. Two (7.4%) patients each had double uterus and subseptate uteri respectively.

Conclusion: Mullerian duct anomalies are the commonest cause of primary amenorrhoea but frequency of presentation to hospital is very low. It may be due to poor knowledge and shyness of parents as well as patients. We have to educate them that it is treatable and their daughters can become active members of society.

Key Words: Primary amenorrhoea, Mullerian defects.

Introduction

Abnormalities of upper genital tract of female, which are present at or before birth, are called “Congenital Mullerian Duct Anomalies”. Developmental anomalies of Mullerian ducts are some of quite fascinating disorders encountered by gynaecologists. Mullerian ducts are primordial anlage of female reproductive tract. During embryogenesis, two Mullerian ducts appear as buds in outer part of each intermediate cell mass during 5-6 weeks of intrauterine life. The ducts from each side fuse together, and then canalized to form two uterine tubes, the uterus and upper one third of vagina. Varying degree of mullerian duct anomalies can occur due to agenesis, hypoplasia, noncanalization, malfusion and duplication. The involvement may be unilateral or bilateral. These anomalies are frequently associated with renal and axial skeletal anomalies. The entire process is completed by the 22nd week of development.

They are usually diagnosed at puberty or shortly after puberty. In the prepubertal period, presence of normal external genitalia and appropriate developmental milestones often mask abnormalities of the internal reproductive organs. They may be asymptomatic and in most of the cases remain undiagnosed. The patients present with primary amenorrhoea, menstrual disorders, difficulty in coitus, fertility deprivation and acute abdomen. Some of the conditions may be life threatening, for example ectopic pregnancy in rudimentary horn of uterus. The incidence reported varies widely. The range is 1 in 200—600 fertile women. There is 40% incidence of co-existent renal tract abnormalities in these patients. Thorough history and examination is mandatory. Chromosomal analysis is required in some cases. Ultrasonography, hysterosalpingography and laparoscopy help in making final diagnosis. In recent years many cases were found due to availability of better diagnostic procedures and awareness of people. This awareness may further be increased by educating and counseling our people that correction of abnormalities, well in time can provide symptomatic relief and improve reproductive performance of affected individual. This will give courage to community to bring the affected girls to us and not to consider them heterosexual and segregate from society.

Patients and Methods

This study was conducted in Department of Gynae/ Obstetrics of Benazir Bhutto Hospital Rawalpindi, from February 2003 to Feb 2004. & May 2006 to August 2006. Total 27 patients were selected.
Selection was done by considering inclusion criteria, that is all patients with primary amenorrhoea, girls at the age of menarche with acute abdomen or lower abdominal cyclical pain, primary infertility, dysparunia, apariania, severe dysmenorrhoea, recurrent pregnancy loss and preterm labour. Patients with secondary amenorrhoea and secondary infertility were excluded from study. Data collection was through convenience sampling.

The patients were picked up from Gynaecology outpatient department and Gynaecology ward. They were managed and investigated on outdoor basis except girls with hematocolpos and hematometra, who were admitted due to abdominal pain through emergency. Detailed history from patients and mother was taken regarding menstrual and family history. Complete physical examination including secondary sexual characteristics was performed. Abdominal examination was carried out for any palpable swelling. Inspection and examination of vulva and introitus for abnormalities of external genitalia was done. In married women, pelvic examination was carried out for vaginal patency, presence of uterus or any pelvic mass. In unmarried girls, per rectal examination was performed for same purpose. All relevant investigations like karyotyping, ultrasound, IVU, diagnostic laparoscopy and hormonal profile were performed according to patient’s requirement, and keeping in mind the presence or absence of secondary sexual characteristics. All information was written on a designed proforma. Variables selected were age, marital status, way of presentation, menstrual history, obstetric history, family history, socioeconomic history, secondary sexual characteristics, intravenous urography, karyotyping and type of anomaly detected.

Descriptive statistics were used to calculate frequencies, means and standard deviation. Computer programme statistical package for social sciences version 10 was used for data processing and analysis of results.

Results

Twenty-seven cases of Mullerian duct anomalies were detected during the study period. Mean age of patients was 23.07 years. Fifty one percent were married, 40.7% were unmarried and 7.4% were divorced. Majority belonged to low socioeconomic class. Nulliparous patients were 59.3% while 40.7% were parous women.

Primary amenorrhoea is an important clue in detection of Mullerian duct anomalies. In our study, 55.6% patients came with this complaint while rest of the patients had regular menstrual cycle. Regarding family history, only one patient had family history of mullerian duct anomalies.

<table>
<thead>
<tr>
<th>Type of anomaly</th>
<th>Number of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blind End Vagina /Absent Uterus</td>
<td>6</td>
<td>22.2</td>
</tr>
<tr>
<td>Vaginal Agenesis</td>
<td>3</td>
<td>11.1</td>
</tr>
<tr>
<td>Longitudinal Vaginal Septum</td>
<td>3</td>
<td>11.1</td>
</tr>
<tr>
<td>Transverse Vaginal Septum</td>
<td>3</td>
<td>11.1</td>
</tr>
<tr>
<td>Absent Uterus</td>
<td>2</td>
<td>7.4</td>
</tr>
<tr>
<td>Double Uterus</td>
<td>2</td>
<td>7.4</td>
</tr>
<tr>
<td>Subseptate Uterus</td>
<td>2</td>
<td>7.4</td>
</tr>
<tr>
<td>Bicornuate Uterus</td>
<td>1</td>
<td>3.7</td>
</tr>
<tr>
<td>Arcuate Uterus</td>
<td>1</td>
<td>3.7</td>
</tr>
<tr>
<td>Left Tubal Agenesis</td>
<td>1</td>
<td>3.7</td>
</tr>
<tr>
<td>Rudimentary Uterus</td>
<td>1</td>
<td>3.7</td>
</tr>
<tr>
<td>Double Cervix</td>
<td>1</td>
<td>3.7</td>
</tr>
<tr>
<td>Transverse Cervical Septum</td>
<td>1</td>
<td>3.7</td>
</tr>
<tr>
<td>Total</td>
<td>27</td>
<td>100</td>
</tr>
</tbody>
</table>

Some patients specifically presented with primary amenorrhoea, coital difficulty and intermittent lower abdominal pain. Other patients came with malpresentations and on lower segment cesarean section, they were found to have subseptate uterus, bicornuate uterus and arcuate uterus. Patient with bicornuate uterus had previous history of repeated malpresentation. Regarding secondary sexual characteristics, 25 had well developed characteristics while two patients had absent secondary sexual characteristics.

The most common type of Mullerian duct anomaly was found to be blind end vagina with absent
uterus in girls with primary amenorrhoea, followed by longitudinal and transverse vaginal septate seen in parous and nulliparous women respectively (Table I). Beside this general classification, a widely accepted method of categorization of Mullerian Duct Anomalies is provided by American Fertility Society (AFS) 5,7. Major uterine anatomic defects are the basis of AFS categorization. Table 2 tabulates the results according to AFS classification. Most of patients belonged to class I, followed by class II and class III.

**Discussion**

The review of international literature shows there are many studies on mullerian duct anomalies showing its incidence, prevalence, diagnostic tools and management according to specific type6. When some variables of present study were compared with other studies, the mean age group is 23.07 year, similar to that seen in other studies. Because this is the problem of young age, early diagnosis and management saves life and further complications like endometriosis in case of massive hematocolpos and hematometra, ectopic pregnancy in rudimentary horn of uterus 9. Obstetric history, in case of married women is also an important diagnostic factor. Repeated malpresentations and recurrent mid-trimester loss or preterm labour may also be associated with subseptate, arcuate or bicornuate uterus10. This was an incidental finding during caesarean section in our study. Although Diethylstilbestrol related uterine anomalies are quite common, no case was found in this study. These include T-shaped endometrial cavity, widened lower uterine segment, midfundal constrictions and hypoplastic uterus, and cervical and vaginal structural abnormalities 11. Diethylstilbestrol use was banned in 1971, because of its association with vaginal clear cell carcinoma 12.

Another study, which was done in July 1986, 96 women with recurrent first trimester abortions, underwent hysterosalpingography to rule out mullerian duct anomalies. Abnormalities seen in these patients were septate, arcuate and bicornuate uterus 13. Menstrual history is also very important in case of mullerian duct anomalies. In this study group, patients with septate and bicornuate uteri had regular menstrual cycle with dysmenorrhoea only. Patients with absent uterus and vaginal agenesis presented with primary amenorrhoea. On reviewing literature regarding vaginal agenesis, Wise and Bates conducted a study in August 1984 14. It was a retrospective study on 92 women with vaginal agenesis over the last 15 years. As a result of this experience, several modifications of vaginoplasty were developed 15,16.

Socioeconomic history is also considered regarding primary amenorrhoea. People belonging to lower socioeconomic class have little knowledge about mullerian duct anomalies. They hide facts from others by considering their daughters heterosexual. They have fear of segregation from the society so they do not report to the hospital. In this study, one patient was 52 years of age. She came with dysparunia. According to her, she was menopausal. On examination and investigation, she was found to be a case of blind vagina with absent uterus. She was hiding the fact from her family for the last 25 years. She did not take any treatment for primary amenorrhoea and infertility as well. As compared to low class, high socioeconomic class has better knowledge due to education and internet services. Most of patients in this study belonged to lower socioeconomic class. If we review international literature, we see large study group, while only 27 patients were notified during this study period and most of studies were done internationally with very little local data available. One study already described was done in Karachi 9.

The developmental abnormalities of mullerian duct system are quite fascinating disorders encountered by obstetricians and gynaecologists. These abnormalities are one of the most important causes of primary amenorrhoea, which has both personal and social implications. Our community is not familiar with these abnormalities. Many patients get depressed considering themselves as freak. Many of them are segregated from their families being considered as heterosexual. They feel that they have no place in community. They cannot marry and produce children. Parents become very much disturbed about future of their daughters. Many of these issues can be managed surgically and patients can lead a normal sexual life. For example vaginal creation by vaginoplasty is successfully done in our unit for seven patients. Awareness of community is very important as many patients do not report to the hospital due to lack of knowledge and shyness. In the extended period of study, it was noted that two girls came themselves for primary amenorrhoea, so probably the knowledge is increasing gradually. We can educate the people by television programmes, teachers in high schools and social workers. We can take the parents in confidence and try to relieve their fears and thus, we can make their daughters, successful members of the society.

In conclusion, Mullerian duct abnormalities are morphologically a diverse group of congenital disorders involving female genital tract. Although Mullerian duct abnormalities are most common cause of primary amenorrhoea but frequency of presentation
is very low. It may be because of shyness of parents and lack of knowledge that some of these disorders can be managed surgically and patient can lead a normal sexual life. The problem of their daughters can be solved and they can become successful members of society.

References