UNILATERAL PARTIAL VAGINAL ATRESIA WITH BIZZARE CLINICAL SYMPTOMS

by

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Congenital absence of vagina, imperforate hymen, septate vagina with single or double uterus are common varieties of genital tract malformations and do not present any problems in diagnosis. When there is partial atresia of vagina with a functioning uterus, diagnosis is also not difficult with classical history of primary amenorrhoea and cyclic pain. Usually in these cases upper part of the vagina is developed to a varying degree. When the septum between the upper and the lower vagina is not too thick, it is possible to do the dissection vaginally and drain the haematocolpos and advance the upper vagina to meet the lower vagina (Jeffcoate 1969, Hingorani 1971). It is desirable in these cases to leave a vaginal mould for a few months to avoid stricture formation at the junction of the upper and the lower vagina. If major part of the vagina or part of the cervix is not developed then dissection with combined abdomino-vaginal approach is easier and safer. This anomaly is not very common as only 8 such cases were encountered in this series of 65 cases of genital tract malformation. Table shows the types of anomalies seen.

Distribution of cases in relation to type of anomaly.

Type of genital tract malformation.	No. of cases
Congenital absence of vagina with no uterus	20
Imperforate hymen	8
Rigid hymen	2
Double uterus with septate vagina	7
Single uterus with septate vagina	5
Normal vagina with bicornuate uterus	8
Partial vaginal atresia with functioning uterus	3
Normal vagina with no uterus	8
Unilateral partial vaginal atresia with double uterus	4

Unilateral partial vaginal atresia with double uterus is rarely encountered. Only three such cases have been encountered at the All India Institute of Medical Sciences Hospital in ten years from 1961-71. There are difficulties in diagnosis in these cases, where Mullerian duct on one side is fully developed with uterus, cervix and patent vagina and through this the patient menstruates regularly, but on the other side, the uterus cervix and upper vagina are developed and there is atresia of lower vagina resulting in cryptomenorrhoea on that side. As these patients menstruate regularly and the cervix on the side where there is partial atresia of vagina, is neither felt nor seen, creates difficulties in diagnosis. Four such cases who had unusual clinical manifestations and presented difficulties in diagnosis are

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reported below. Case No. 1 has been reported earlier (Hingorani *et al* 1965).

Case 1

Miss A. K. 18 years, had menarche at the age of 14 years and was having regular menstrual periods with slight dysmenorrhoea. She had recurrent haematuria for 2 years for which she had been extensively investigated and also had undergone two operations with no relief. Her previous records showed that at one time, she had been diagnosed as a case of renal tuberculosis with a non-functioning left kidney. The diagnosis had been based on history of haematuria, with intravenous pyelography showing no secretion of dye on the left side, with appearance of 'golf hole' ureteric opening on the left as seen on cystoscopy, and with positive culture of atypical tubercle bacilli from urine. She had received antitubercular treatment with only some relief in dysuria. Attempts at the removal of 'Non functioning tubercular left kidney' had revealed congenital absence of left kidney. A pyosalpinx had been detected at that time.

A second operation had been done for removal of pyosalpinx as it was thought that the pyosalpinx was a possible cause of recurrent cystitis and haematuria. At this operation presence of double uterus was observed. Following this a detailed gynaecological history revealed that haematuria was cyclical, though it used to continue after the menstruation was over. Hysterogram revealed right uterus which was communicating with the vagina through which patient was menstruating regularly. A cystoscopy during menstruation revealed, menstrual blood coming through an opening (which was communicating with the left genital tract) which earlier had been mistaken as 'golf hole' ureteric opening typical of tuberculosis. At the final operation left uterus and upper part of left vagina were removed and patient was cured of recurrent haematuria. It was now possible to explain all her symptoms. Patient had failure of fusion of Mullerian ducts and had two uterii and partially atretic left vagina. In this she developed haematocolpos after menarche, and after two years, pressure in this was sufficient to cause a fistula in the bladder which resulted in recurrent haematuria. The fistulous opening was narrow so that menstrual blood drained only partially and continued to drain even after menstruation was over. The opening helped the ascending infection and to infection in the haematosalpinx and caused recurrent cystitis which was somewhat relieved with streptomycin.

Case 2

Miss M. M., aged 17 years, had investigations and treatment for pyrexia of unknown origin for 1 year without any relief. She had menarche at the age of 14 years. Periods were regular but she had dysmenorrhoea. On examination under anasthesia a cystic mass was felt in the left and anterior fornix. On needling, a chocolate coloured fluid was aspirated. With experience of first case, partial atresia of left vagina was considered and with due care, avoiding any injury to the bladder, an incision in the cystic mass was made. After draining the haematocolpos the left cervix could be felt and seen and patient was relieved of fever and dysmenorrhoea after this procedure.

Case 3

Mrs. G. B., aged 28 years, PO + O was married for 12 years. Her main complaint was infertility. She had no dysmenorrhoea and no dyspareunia. She had a large cystic mass in the left and anterior fornix. An obstetric resident had admitted her with the diagnosis of vaginal cyst. However, with the experience of the above two cases, and the finding of shift of the cervix to the right side, correct preoperative diagnosis of partial atresia of left lower vagina with double uterus was made.

Case 4

Mrs. R., aged 19 years, came with main complaint of vaginal discharge for 2 years since her marriage. Her menarche was at 17 years, with regular menstruation and no dysmenorrhoea. Diagnosis, by the registrar was ? infected vaginal cyst. Patient had a cystic mass which was occupying the right upper vagina and was pushing the cervix to the left side. In view of the findings being identical to the above 3 cases,

diagnosis of congenital unilateral partial vaginal atresia of vagina with bicornuate uterus was made. Intravenous pyelography revealed absence of right kidney. On needling the cystic mass vaginally old blood was obtained and the diagnosis was confirmed. Septum was then excised and right cervix could then be felt and seen.

Discussion

Desces (1854) described a case of incomplete fusion of Mullerian ducts with unilateral obstruction where patient ultimately died of this condition and at autopsy the exact nature of anomaly was detected.

Menouria, with the upper vagina communicating with the bladder has been described with the single uterus (Jones and Wheeless 1969). However, in the cases of that type, with single uterus, with congenital absence of vagina and in the absence of normal menstruation there is no difficulty in diagnosis unlike case No. 1 with the double uterus.

Masani (1967) reported thirteen cases of haematometra, and of these two had haematometra in the rudimentary horn of the bicornuate uterus. Main presenting symptom, in both these cases, was severe dysmenorrhoea. He emphasized that in these young patients, as they are regularly menstruating, the diagnosis may be missed and often a mistaken diagnosis of a fibroid or an ovarian cyst with torsion is made. However, presence of severe dysmenorrhoea and a mass in young girls should make one think of this type of genital tract malformation. In both those cases the horn of the uterus with haematometra was excised. Apparently they did not have unilateral partial vaginal atresia, with upper haematocolpos. However, if such a possibility is entertained the management of the case becomes simple.

In three cases the unilateral obstruction of the vagina was on the left side and urinary tract was also not developed on the left side. Klaften (1931) refering to this condition as "Haematocolpos Lateralis" also noted ipsilateral absence of the kidneys. Same was found by Woolf and Allen (1953). Previous analysis of concomitant malformation of the genital and urinary tract systems in the female at this Institute (Chawla *et al* 1963) had shown gross malformation of urinary tract in 55% of cases of genital tract anomaly. Where there is congenital absence of a kidney it should make one look for genital tract anomaly, even though the woman may be regularly menstruating. Examination under anaesthesia, as was done in case 2, aids the diagnosis. This practice particularly in virgins has also been recommended by Miller (1922).

Summary

Sixty-five cases of genital tract anomalies are analysed. Of these, four cases had incomplete fusion of Mullerian ducts with unilateral obstruction and presented with recurrent haematuria, pyrexia of unknown origin and as vaginal cyst and with vaginal discharge. These four cases are presented and discussed.

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