INTRODUCTION

Complex congenital anomalies of female reproductive tract are uncommon events that are hypothesized to occur most commonly as a result of Müllerian dysregulation during embryogenesis. These anomalies present with varying degrees of Müllerian fusion defects and agenesis leading to a broad spectrum of phenotypes as classified by American Society for Reproductive Medicine. Female reproductive tract abnormalities are generally encountered in 2 – 3% of women. Various combinations have been described in literature. Imperforate hymen is the most common defect. Incidence of isolated vaginal agenesis is 1:5000 women and that of uterus didelphys in fertile women is 0.16%, but the combination of the two is not known. However, incidence of Müllerian anomalies is higher in population with impaired fertility, with 8% having uterus didelphys. The present case depicts an unusual anomaly with a common presentation and yet another different mode of treatment. It can be a technical challenge to restore the function and anatomy of a complex congenital anomaly, and imaging modalities like ultrasonography and MRI can be of great help in making a surgical blueprint.

CASE REPORT

An 18 years old girl presented in emergency with primary amenorrhoea and pain in the abdomen. She was married 5 months back and complained of coital difficulty. On examination, she was 156 cm in height with well developed secondary sexual characters, abdomen was soft and no lump was palpable. Vagina was replaced by a small dimple and no bulge was seen or palpable at the introitus. On rectal examination, a tense cystic mass suggestive of haematocolpos was felt anteriorly about 4 cm above the anus opening.

Pelvic ultrasound illustrated a haematocolpos of 12 x 7.5 x 10 size with a normal uterus, however, MRI pelvis suggested a bicornuate uterus with longitudinal septum in the upper 5 cm of vagina with haematocolpos, haematometra and bilateral haematosalpinx (Figures 1 and 2). No renal tract abnormality was found on intra venous pyelography. Patient was planned for diagnostic laparoscopy and vaginal reconstruction. Laparoscopy revealed a double uterus with bilateral haematosalpinx; both ovaries were normal. A transverse incision was given on the vaginal dimple and dissection was done carefully for approximately 5 cm to reach the tense bulge of the haematocolpos. Incision was given on the bulge and approximately one and a half liters of chocolate colored blood was drained. On inspecting the upper vagina, a longitudinal septum was seen in upper third of vagina with two cervices. Dilators were placed in both

ABSTRACT

Complex malformations of female genital tract are not so common. Their correct identification is of paramount importance for appropriate management. A thorough knowledge of embryology, pre-operative imaging with MRI and examination under anaesthesia is essential to identify accurately the constellation of anomalies and to plan appropriate management. This case reports the coexistence of Müllerian abnormality with vaginal agenesis in an 18 years old girl which was managed by dissecting the lower half of vagina and pull-through vaginoplasty.

Key words: Uterus didelphys. Partial longitudinal vaginal septum. Vaginal agenesis. Haematocolpos. MRI. Mullerian duct abnormality.

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the cervix and continuity with the uteri was confirmed laparoscopically by seeing the transmitted movements with dilator. Mucosal margins of the upper vagina were pulled down till the introitus and sutured there and a vaginal mould covered with amnion was inserted in the vagina to keep the newly formed lower half of vagina patent. The mould was replaced after 4 days and kept for another 4 days. Patient was trained to change the mould herself. At the time of discharge from hospital, the vagina and the introitus were wide enough for intercourse. The patient was told to insert the mould at night for 3 months. On follow-up after 2 months, she had an adequately wide vagina.

**DISCUSSION**

Uterus and vagina are formed embryologically by the dynamic process of differentiation, migration, fusion and canalization. A wide variety of abnormalities of uterus and vagina are caused by the disruption of one or more of the above processes. Complete failure of fusion of the two Müllerian ducts can result in a uterus didelphys with longitudinal vaginal septum. Selective agenesis of lower vagina, segmental agenesis and vaginal atresia is a separate entity and is usually associated with normal Müllerian development.

The combination of uterus didelphys with partial longitudinal vaginal septum and agenesis of bilateral lower vagina is not reported as of now, although closely resembling combinations have been reported by Whitfield *et al.* and Moawad *et al.*6,7 Whitfield *et al.* reported 3 cases where instead of partial longitudinal vaginal septum, it was a complete duplicated hemivagina. Moawad *et al.* reported a case with a transverse vaginal septum instead of distal vaginal agenesis. Uterus didelphys with unilateral hemivaginal obstruction instead of bilateral has been reported many times earlier but only 2 case reports of bilateral obstruction could be found.8-10 A review of literature revealed that combinations of defects are potentially noteworthy as they require individualized therapeutic approach.

Moawad *et al.* used a different approach in the treatment of his patient. His patient, a 15 years old girl who presented with primary amenorrhea and cyclical pelvic cramps was made to perform vaginal dilatation over a period of 10 weeks to a depth of 4 cm.4 This method thinned out the transverse vaginal septum and required only little dissection for excision of the septum, but the relief of pain and obstruction should be carried out as soon as possible as was done in the present case.

Whitfield *et al.* performed a staged pull through hemivaginoplasty instead of single stage operation in this case, because of the asymmetric uterine didelphys with bicornis.7

Although incidence of Müllerian abnormalities is not low and cases of isolated defects of various types have been reported previously but the combination of anomalies as in present patient is unique. This suggests the need of careful pre-operative evaluation of the anomalies and individualization of the therapeutic approach to restore functional anatomy and reproductive potential.

The complex combination of various Müllerian anomalies can be a challenge to the gynaecologist and there may be a difficulty in diagnosing the type of malformations. A thorough knowledge of embryology, pre-operative imaging with MRI and examination under anaesthesia is essential to identify accurately the constellation of anomalies and to plan appropriate management.

**REFERENCES**

