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CASE REPORT

Distal vaginal atresia misdiagnosed as imperforate hymen: A case managed by transperineal vaginal pull through (distal colpoplasty)

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Abstract Three different types of vaginal outlet obstruction are described in the literature: imperforate, transversal vaginal septum and vaginal atresia. Diagnoses were made in different ages of life, from neonatal to the teenage years. Clinical and ultrasound examinations and MRI dominated in establishing the diagnosis. 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 distal vaginal agenesis in an 13 year old girl which was managed by dissecting the lower half of vagina and pull-through vaginoplasty.

Rarity and variable presentation of congenital genital tract anomalies can lead to delayed diagnosis and erroneous management. A high index of suspicion and cross-sectional imaging can help in early diagnosis. A comprehensive management is imperative to preserve the reproductive potentials, as significant proportion of patients may experience sexual difficulties, menstrual irregularity, and infertility.

1. Introduction

The vaginal outlet mechanism consists of undisturbed fluid passage from vagina through hymenal opening. The genital tract outflow is important for secretion and menstrual effluxion and as a pathway in reproductive function. Congenital outflow obstruction may occur at different levels and with different clinical presentations (1).

Embryological development of vagina results from lower portion paramesonephric ducts fusion and regression forming

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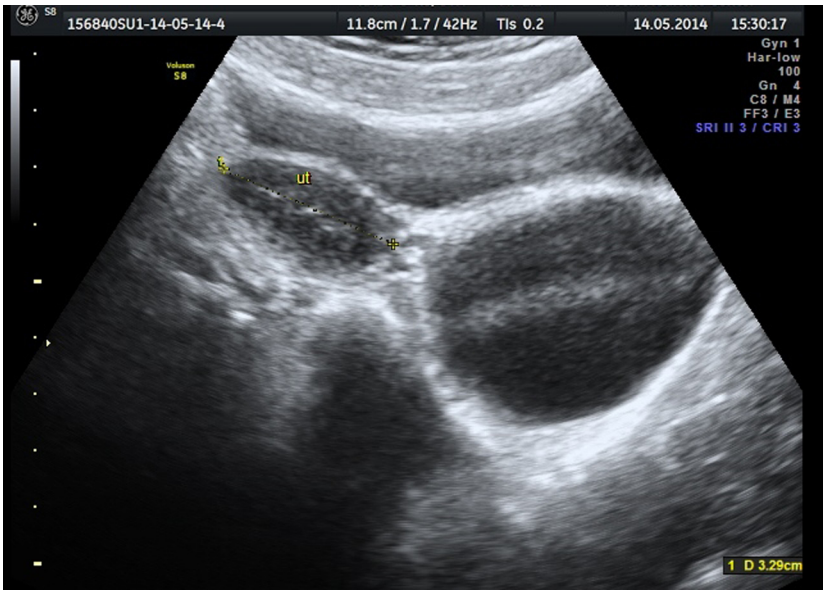

Fig. 1 Transabdominal ultrasound revealing haematocolpus.

the uterovaginal primordium (gives rise to the uterus and superior part of vagina). Contact of the uterovaginal primordium with urogenital sinus induces formation of paired outgrowths named sinovaginal bulbs. The sinovaginal bulbs fuse to form the vaginal plate. The cells of the fused bulbs undergo apoptosis to form the lumen of the vagina. Until late foetal life the lumen of the vagina is separated from the cavity of the urogenital sinus by a membrane – the hymen (2,3).

Abnormal development can result in any of the following three: imperforate hymen (failure of epithelial degeneration), low, mid, or high transverse septum of the vagina (incomplete unit), or atresia of the vagina resulting in persisting of a portion of solid cells cord. Recently interest has focused on expression and function of the mammalian HOX genes as a possible aetiology of these genital developmental abnormalities (4).

Despite different origins of some parts of the vagina and hymen, their obstructed forms are clinically manifested as hydro (metro) colpos in the neonatal period and haemato (metro) colpos at the beginning of puberty.

Vagina originates from two embryonic structures: the upper part from Mullerian duct system and the lower third from the urogenital sinus. Canalization of the vaginal canal is complete by the 20th week. Misdevelopment presented as failure of fusion or canalization of these two systems in vertical plane may be clinically present with a spectrum of Mullerian duct anomalies. Vaginal atresia (VA) is one of them. Missing portion of the vagina is replaced with fibrous tissue (5). According to the American Society for Reproductive Medicine 1998 Classification, vaginal atresia is categorized as Type I (6). Some authors believe that transverse vaginal septum is partial vaginal aplasia (5).

Clinical findings vary depending on the anatomy of the vaginal outlet and the changes in the upper vagina and uterus. The upper vagina becomes enormously distended when the girl starts to menstruate, usually producing a palpable abdominal mass arising from the pelvis. The Fallopian tubes can be normal, although they may be distended allowing escape of the fluid into the peritoneum. Other anomalies are occasionally seen with hydrometrocolpos. Some other combinations of structural anomalies may be present, such as Mayer–Rokitansky–Küster–Hauser syndrome, Bardet–Biedl syndrome, Fraser syndrome, and Winter syndrome (7–9).

The diagnosis is usually made when symptoms of obstruction are obvious. Ultrasound, abdominal and endorectal (10), and other imaging studies confirm the physical examination. Urogram would demonstrate anterior and superior displacement of bladder and, possibly, hydronephrosis and hydroureter. As Mullerian agenesis can be associated with other anomalies, particularly those of kidneys and skeleton (11,12), further investigations are indicated.

Therapy is directed to relieve the obstruction of the vaginal outlet and provide normal sexual life and reproductive function. Regarding the pathological problem, cultural and religious considerations, it is necessary to inform the parents about the risk of defloration. Vaginal reconstruction is required, sometimes by an abdominoperineal approach, but stenosis and fistula formation could complicate postoperative period.

2. Case report

An 13 year old girl presented with primary amenorrhoea and pain in the abdomen. She was not married. On examination, she was 160 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 bulge was seen but not at the introitus. On rectal examination which is highly essential and important, a tense cystic mass suggestive of haematocolpos was felt anteriorly about 4 cm above the anus opening which with no bulging membrane was suspicious of segmental vaginal atresia. Pelvic ultrasound (Fig. 1) illustrated a haematocolpos of 10 cm size with haematometra but it was difficult to assess the level of obstruction whether imperforate