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Case Report

A Rare Case of Neglected Transverse Vaginal Septum with Poor Consequences: A Case Report

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ABSTRACT

Background: Transverse vaginal septum is a rare mullerian anomaly. Patients with a complete transverse vaginal septum generally complain of primary amenorrhea. Here, we presented a rare case of high transverse vaginal septum with abdominal mass during infancy in addition to chronic pelvic pain and cyclic hematuria during adulthood.

Case presentation: The patient was a 26-year-old female with complain of severe abdominal and pelvic pain with significant weight loss. She had a history of surgery for correction of hydrometrocolpos (fluid accumulation in the uterus and vagina) because of transverse vaginal septum at infancy period. She had no gynecologic follow-up until the age of 26 years. At the last surgical evaluation, diffuse pelvic endometriosis with advanced metastatic colon cancer was noted.

Conclusion: After diagnosis of transverse vaginal septum during infancy period, detailed counseling should be performed about the other treatment and follow-up which is needed to prevent severe complications that could affect the quality of life.

Keywords: Hydrometrocolpos, Vaginal septum, Vesicovaginal fistula, Endometriosis

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Introduction

Transverse vaginal septum is a rare mullerian anomaly that occurs when there is a failure of vertical fusion in the urogenital sinus and paramesonephric ducts. The incidence of a transverse vaginal septum appears to be between 1 in 21,000 and 1 in 72,000 (1). In neonates and young infants, an imperforate transverse vaginal septum can lead to serious and life-threatening problems caused by the vaginal obstruction and compression of surrounding organs by fluid that has collected above the septum. The fluid comes from endocervical glands and müllerian glandular epithelium in the upper vagina that has been stimulated by the maternal estrogen (2). Patients with high transverse vaginal septa are most likely to experience pelvic pain earlier than those with septa located lower in the vagina; due to smaller space for the hematocolpos (blood accumulation in the vagina) that occurs after initiation of menses (1).

The aim of this report was to introduce a rare case of high transverse vaginal septum which presented with abdominal mass during infancy in addition to chronic pelvic pain and cyclic hematuria during adulthood.

Case report

A 26-year-old single woman with severe abdominal and pelvic pain referred to Imam Khomeini hospital, an academic hospital of Tehran University of Medical Sciences, in September 2018. She also reported a significant weight loss (about 20 kg) during the last 6 months because of severe anorexia. She also complained of abnormal defecation. She had a history of huge abdominal mass during infancy (2 months old) and abdominal surgery was performed at that time. The diagnosis was hydrometrocolpos because of transverse vaginal septum. At the same time, the surgeon tried to open the vaginal septum, but the rectum was perforated and the surgery was ended. She had amenorrhea (never experienced normal menstruation); but did not consult with any gynecologist since the recent year. She also complained of cyclic hematuria, so she underwent cystoscopy by an urologist and a vesicovaginal fistula was detected 2 months before recent gynecologic visit. At gynecological visit, abdominal examination revealed a 12 × 6 cm firm and tender mass in the midline. The vaginal examination revealed the obstructed vagina, also a large tender pelvic mass was palpated via rectal examination. Magnetic resonance imaging (MRI) showed low transverse vaginal septum and large hematocolpos with a huge pelvic mass which was about 12 × 35 cm in size between rectum and cervix; moreover, suspected vesicovaginal fistula was also reported (Figure 1). Spiral computed tomography (CT scan) of the abdomen and pelvis with contrast media revealed a mass (82 × 66 × 134 mm) in pelvic cavity with irregular wall thickness enhancing (Figure 2). The uterus and ovaries could not be seen. Sigmoid was passing behind the mass and narrowed, which was in favor of sigmoid involvement. Bladder wall involvement was also seen. Mild hydronephrosis were noted in both kidneys. Colonoscopy was performed and revealed a big infiltrative and vegetative lesion from rectum to sigmoid junction which made the lumen too stenotic and scope could not pass through, so multiple biopsy was taken. Complete blood count (CBC) and liver and kidney function tests were normal. The tumor markers including CA125 was 126.4 U/mL (normal range is up to 35 U/mL), CA 19.9 was more than 12,000 U/mL (normal is ≤37 U/mL), and CEA was 55.19 U/mL (normal range is up to 5 U/mL). HE4 and ROMA index which are the specific ovarian tumor markers were normal. Surprisingly, the pathological assessment of rectal biopsies revealed chronic inflammation. After consultation with the patient and her family, the exploratory laparotomy was planned. The abdomen was opened with midline incision. After entrance to the peritoneal cavity, a frozen pelvic was noted, the small intestine was inflamed and diffuse superficial tumoral seeding was observed over the intestinal, vesical, and uterine surface. External surface of uterine and bladder wall were completely inflamed. Uterine size was about 12 cm. There was a 7 × 8 cm complex mass in the right ovary. Tumoral seeding was noted over the small intestine and rectosigmoid colon, so several biopsies were taken. The frozen section report was colon adenocarcinoma. Colestomy was performed and abdomen was closed. Permanent pathological evaluation also confirmed the diagnosis of metastatic colon adenocarcinoma.

Informed consent was obtained from the patient's family to publish the information and accompanying figures.

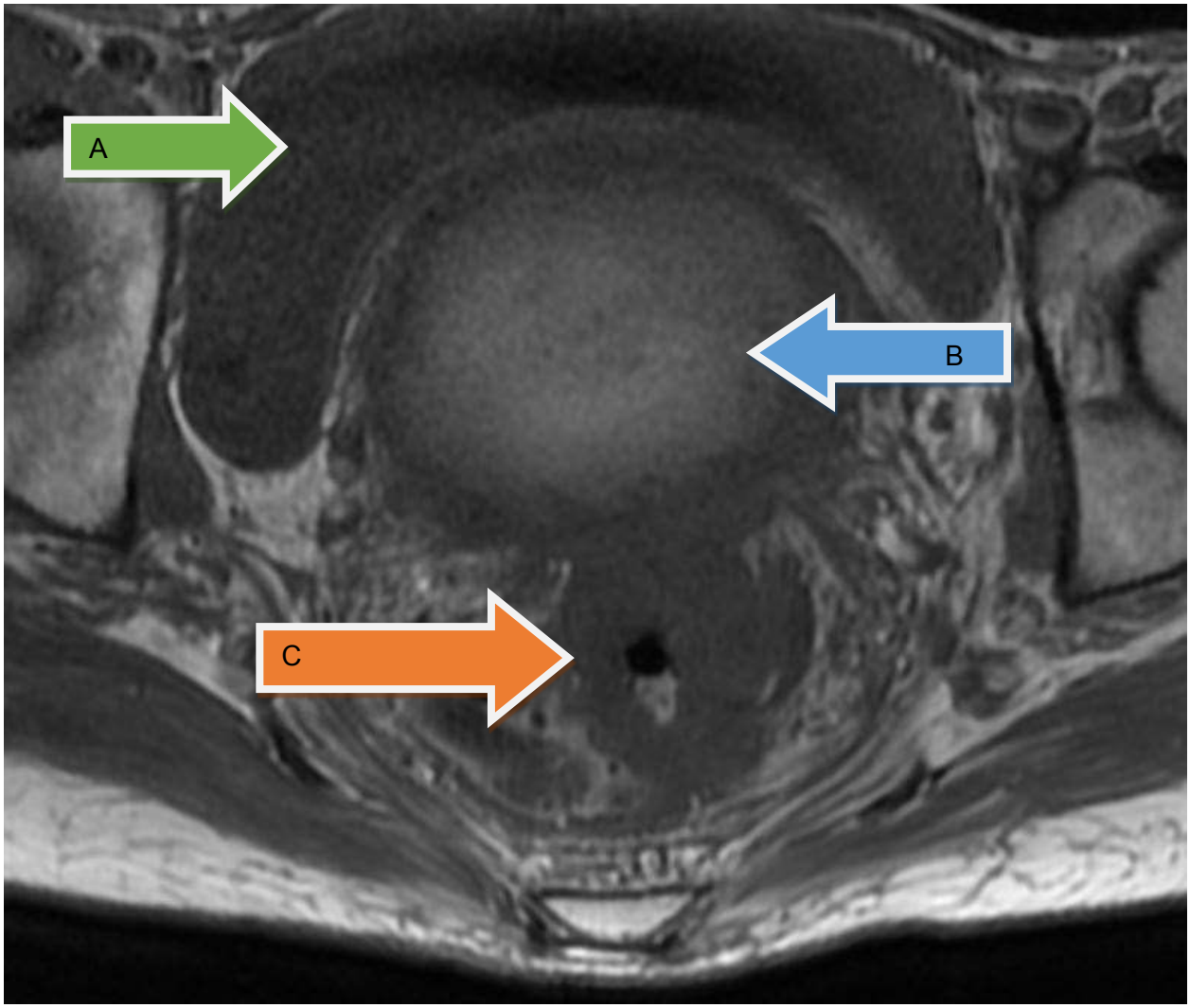

Figure 1. Magnetic resonance imaging of: A) Bladder; B) Uterine; C) Pelvic mass.

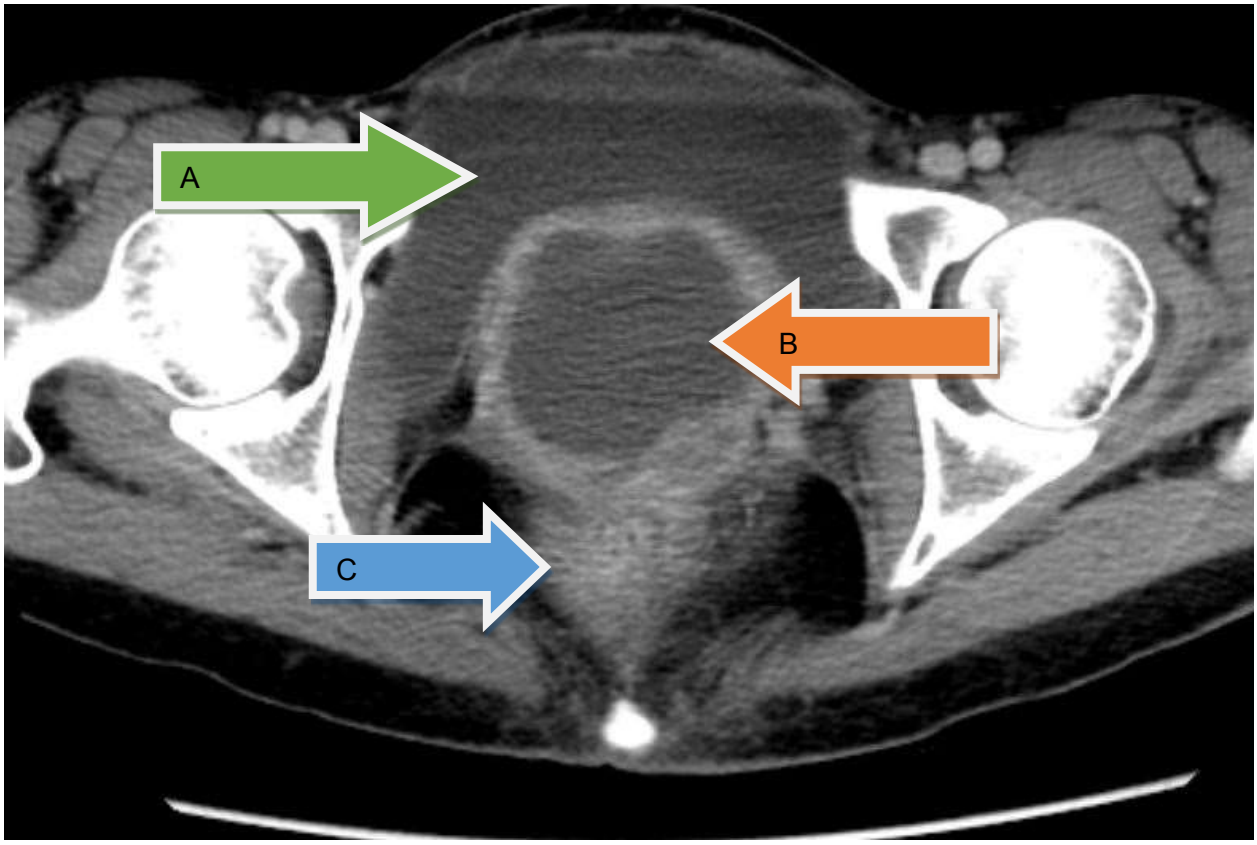

Figure 2. Computed tomography of: A) Air bubbles in bladder; B) Uterine; C) Pelvic mass.

Discussion

Transverse vaginal septum may be rarely detected in an infant or young child which may be presented as an abdominal mass (1). Also, in the present case, the first presentation of the anomaly was an abdominal mass at the infancy period (at 2 months old). This early presentation is also reported in some other studies (3-5). One of the important differential diagnosis of transverse vaginal septum is imperforated hymen, which also could mimic the same presentation at neonatal or adolescent period (6). In the present case, the correct diagnosis was not established before the first surgery, so laparotomy with large midline incision was performed at infancy period. After the diagnosis of hydrometrocolpos, they tried to perform vaginoplasty which led to rectal perforation. However, the correct management especially during infancy or childhood is an important issue; some investigators recommended that initial decompression and short-time vaginal catheterization can be an appropriate treatment for hydrocolpos during infancy period (7). A successful transabdominal drain for decompression of abdominal mass with abdominal vaginoplasty drain without transvaginal septal resection was reported as an effective and safe treatment method in young children (3). Although hydrometrocolpus was not drained, complications like pyocolpos, hydronephrosis, sepsis, and failure to thrive or ruptured hydrocolpos may be occurred (4). After puberty period, the diagnosis of transverse vaginal septum could be managed transvaginally with successful outcomes through septum resection in addition to using vaginal mold (a device for keeping the vagina open) to prevent vaginal restenosis (8, 9). Unfortunately, the present patient did not refer for correction of vaginal septum until the age of 26 years.

Untreated transvaginal septum even after menarche may lead to advanced endometriosis and severe pelvic pain in addition to infertility and low quality of life (10). Just like the present patient, in whom inadequate treatment and poor follow-up led to vesicovaginal fistula and severe pelvic pain due to deep infiltrating endometriosis. On the other hand, optimal management of this anomaly usually has desirable outcomes (10).

Conclusion

After diagnosis of transverse vaginal septum during infancy period, detailed counseling should be performed about the other treatment and follow-up, which is needed to prevent severe complications that could affect the quality of life.

Conflict of interests

The authors declare that there is no conflict of interests.

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