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Newer Approach in Management of OHVIRA Syndrome (Variant of Mullerian Duct Anomaly)

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Abstract: ***This case report details the surgical management of a 14-year-old girl diagnosed with Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome - a rare congenital anomaly often misdiagnosed due to its overlapping symptoms with common abdominal conditions like appendicitis. In my view, what stands out most is the successful integration of endoscopic trans-illumination during exploratory laparotomy, allowing for clear delineation and precise resection of the transverse vaginal septum. The patient initially presented with acute urinary retention and lower abdominal pain, symptoms that intensified due to hematocolpos secondary to the obstructed hemivagina. Despite the rarity of OHVIRA, it is evident that early detection using a combination of ultrasound and MRI remains vital, particularly in adolescent females with renal anomalies and menstrual irregularities. This case emphasizes the utility of a single-stage surgical approach, combining effective drainage and anatomical restoration, which not only alleviated symptoms but also preserved future reproductive potential. Taking this further, the report reinforces the growing importance of multidisciplinary coordination and imaging-led decision-making in managing Müllerian duct anomalies. It also reflects the nuanced evolution of surgical interventions—from historically invasive methods to more refined, targeted techniques aimed at minimizing long-term complications such as infertility and endometriosis.***

Keywords: OHVIRA syndrome, hematocolpos, transverse vaginal septum, Müllerian anomaly, adolescent gynecology

1. Introduction

The Mullerian ducts develop to form the fallopian tubes, uterus, cervix, and upper one-third of the vagina at 6 weeks' gestation. In addition, The Wolffian ducts have crucial roles in developing the Müllerian ducts and kidneys. Thus, abnormal development of the Wolffian ducts leads to uterine and vaginal malformations complicated by urologic abnormalities.[1] Müllerian anomalies are congenital developmental anomalies of the female reproductive tract with an overall incidence, including both major and minor müllerian anomalies, of 7–10% while after excluding the minor ones the incidence is 2–3%.[2]

OHVIRA syndrome, also known as Herlyn-Werner Wunderlich syndrome (HWW syndrome), is a Mullerian duct anomaly which is associated with uterus didelphys, unilateral obstructed hemivagina, and ipsilateral renal agenesis.OHVIRA syndrome belongs to the group of ORTAs (Obstructive reproductive tract abnormalities) with incidence varying between 0.1% and 3.8% in the general female population and 7% in all mullerian anomalies.[3,4] In present case report, we reported a case of OHVIRA syndrome managed with help of endoscopic trans-illumination at our hospital.

2. Case Report

A 14 year old presented with pain in lower abdomen and acute retention of urine for 4 to 5 days.Pain in abdomen was sudden in onset, gradually progressive, non radiating, intermittent and relieved on medication. The patient attained menarche at 12 yrs of age.

Menstrual history: 4-5 days bleeding, in a 26-30 days cycle, regular normal flow with presence of dysmenorrhea, soakage of 3-4 pads per day.

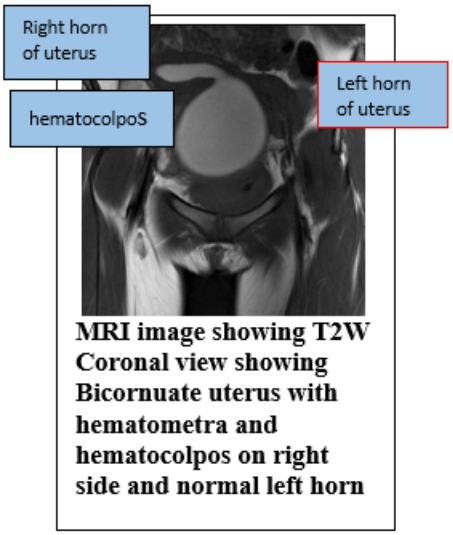
On General Examination, patient average built, weight and height. Her secondary sexual characters and breast development was normal as per her age, per abdominal examination-abdomen was soft, tenderness present over right lower abdomen.

Laboratory examination was unremarkable and normal liver and renal function. Urinalysis was unremarkable.

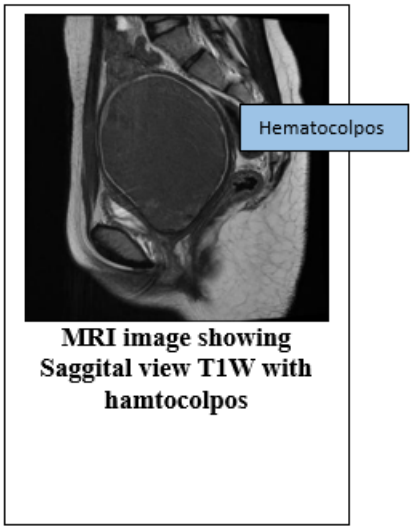
USG abdomen+pelvis showed -Bicornuate unicollis uterus with non communicating right horn showing collection with fine internal echos, right hydrosalpinx. Evidence of 2 uterine horns with 2 uterine cavities noted measuring left horn 1.8 X 3.5 X 5.6 cms and right horn measuring 9*9*13.5cms. There is right hydrosalpinx measuring 9.9 X 3.2 X 7 cms with a wall thickness of 9 mm. Both ovaries are visualised normally.

MRI pelvis was done revealed Bicornuate bicollis configuration of uterus. Right horn- gross hematocolpos (max diameter 82.2mm) noted on right which is extending superiorly in the right endometrial canal and right fallopian tube .It appears T1 hyperintense and T2 hypointense and showing foci of blooming on GRE. Left horn of uterus shows normal endometrial canal, endocervical canal is effaced due to adjacent hematocolpos in the right horn. Bilateral ovaries appears normal. Right kidney not visualised. Bicornuate Bicollis configuration of uterus with obstructed right vagina (likely secondary to transverse vaginal septum) and upstream hematocolpos with hematometra and hematosalpinx. In view of absent right kidney features

suggestive of OHVIRA syndrome (Obstructed Hemivagina with Ipsilateral Renal Agenesis Syndrome)



MRI image showing T2W Coronal view showing Bicornuate uterus with hematometra and hematocolpos on right side and normal left horn



MRI image showing Saggital view T1W with hamtocolpos

Under general anesthesia Per speculum and per vaginal examination was done. Evidence of normal left side cervix with no any cervical opening over right side with bulging over right side due to hematocolpos with transverse vaginal septum. The patient underwent exploratory laparotomy with drainage of hematocolpos with right sided transverse vaginal septum resection.

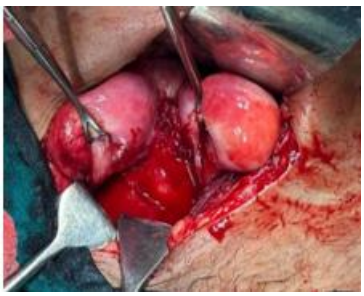


Image showing bicornuate uterus with hematocolpos

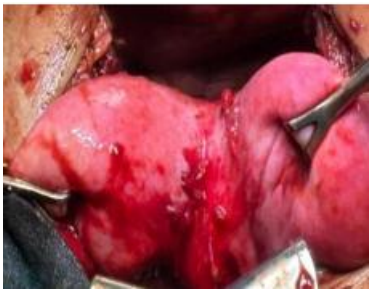


Image showing bicornuate uterus after draining blood inside the cervix

Around 300 ml of chocolate colored thick gelatineous fluid was drained

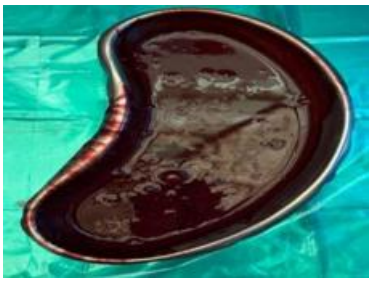


Image showing 250-300 ml of choclate like gelatineos fluid

Vaginoscopy performed revealing transverse vaginal septum on right side.

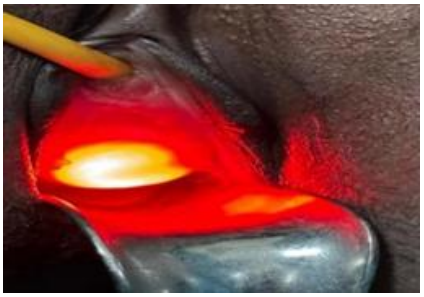


Image showing vaginoscopy-Telescope with light source was inserted through the incision and septum delightened

Incision taken on septum, remaining blood allowed to flow and edges of vaginal septum undermined by vicryl2 .0.A Foley's catheter was then inserted into the vaginal cavity, and the balloon was inflated with 10 ml of normal saline to maintain patency of the newly created passage. An intraperitoneal drain was placed, and the abdomen was closed in anatomical layers. On day 5 foleys inserted into the cavity was removed. On day 8 all sutures removed and patient has been discharged on day 9

3. Discussion

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is usually diagnosed at the time of menarche when slow accumulation of blood from menses remains obstructed in one of the vaginal vaults. The

resulting hematocolpos causes distention of the uterus and fallopian tubes subsequently. The local pressure as a result of this distention on surrounding structures causes the pain in the abdomen. Clinically, it is often misdiagnosed as appendicitis but the presence of unilateral kidney should prompt an inspection of OHVIRA syndrome. Delay in diagnosis have been attributed to lack of understanding of this condition by radiologists, gynecologists, urologists, nephrologists, pediatricians, and pediatric surgeons.[5]

OHVIRA is a type III anomaly according to the AFS and ESHRE / ESGE classifications. According to the new clinical and embryological classification of female genital tract malformations modified from Acién,1992, OHVIRA belongs to class II.[6] Renal agenesis including the more common right side and Müllerian ducts anomaly often coexists. So, in all patients with Müllerian ducts anomaly, it is important to investigate the urinary tract anomaly.

The possible complications associated with this syndrome are a consequence of retrograde menstruation, such as endometriosis, pelvic adhesions, infectious collections and pelvic inflammatory diseases. Delayed diagnosis can lead to complications such as abscess formation, pelvic inflammatory disease, endometriosis, pyometra and infertility.[7]

Diagnosis of OHVIRA syndrome requires a multimodal approach, which includes a detailed history, meticulous examination, appropriate imaging studies and a high index of suspicion. In our case, the diagnosis was made on clinical examination. Ultrasound and MRI are the recommended imaging modalities for identifying this condition, with ultrasound being the first choice. 3D ultrasound is an excellent substitute for MRI and gold standard for identifying Mullerian duct anomalies. It has 93% sensitivity and 100% specificity in this regard and associated extragenital components (**8**)

Primary management of the OHVIRA syndrome is requires surgical intervention like of excision of vaginal septum to relieve obstruction. Historically, surgeons advocated a two-stage procedure, with the initial surgery to reduce the hematocolpos and the second to re-sect the excess septum after a period of wound-healing and vaginal remodelling. However new trend is to do both in the single stage vaginoplasty with complete resection of the septum which not only relieves symptoms but also prevents long term complications like endometriosis and even improves sexual functions and conception chances**. (9)**

The management of OHVIRA syndrome begins with initial incision of the vaginal bulge to relieve the obstructed hematocolpos. Often, a vaginoplasty is required to excise the vaginal septum if it was the cause of the obstruction. However, hemihysterectomy of the obstructed uterus is not recommended anymore as studies have shown that pregnancy in a previously obstructed uterus is still viable.[9,10]

In our case an exploratory laparotomy was planned with the goals of evaluating pelvic anatomy and relieving the obstruction caused by hematocolpos. A key step in this surgery was the use of a vaginoscope equipped with a light source and camera. This facilitated enhanced visualization of the vaginal canal and enabled precise identification of the transverse vaginal septum. The septum was incised under direct vision, allowing for a complete resection while minimizing the risk of injury to surrounding structures. Suturing of the septal edges was done meticulously using Vicryl 2-0 to ensure proper healing and to prevent re-stenosis. A Foley's catheter was then inserted into the vaginal cavity, and the balloon was inflated with 10 ml of normal saline to maintain patency of the newly created passage.

Arakaki et al.,[1] reported a 13-year-old with right obstructed hemivagina and renal agenesis, diagnosed via MRI.The patient was managed with septal resection with an electrocautery scalpel under trans abdominal ultrasonography guidance followed by laparoscopic examination was done.The advantage in this case was Early diagnosis and use of minimally invasive approach.In our case the patient had similar age advantage in detailed intraoperative Vaginoscopy; however, we used laparotomy due to large hematocolpos.

Begoña Navarro Díaz et al.,[7] presented a case of 14-year-old with right renal agenesis and obstructed hemivagina, diagnosed via MRI, treated with septal resection. The main advantage in their case was early diagnosis and imaging, early MRI use. In our case there was a added advantage of dealing with urinary retention and performing Vaginoscopy-guided surgery

Pillai et al.,[5] described a classic OHVIRA case managed trans-vaginally, approach was minimally invasive, allowing for faster recovery. In contrast our patient required a broader surgical exposure due to complex anatomy and large hematocolpos and there was a slight disadvantage that the procedure was more invasive.

The management of OHVIRA syndrome requires a multidisciplinary approach, and the current literature emphasizes the diversity and challenges in the diagnosis and treatment of this condition

4. Conclusion

OHVIRA syndrome is an uncommon congenital anomaly with clinical significance. MRI plays an important role in detection as majority of cases are misdiagnosed. OHVIRA syndrome management required surgical intervention, which should be minimally invasive, anatomically sound. In present case, introduction of endoscope to delineate transverse vaginal septum which is neosurgical approach in management of OHVIRA syndrome with a transverse septum.

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