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Case Report

A rare case of persistent urogenital sinus in an adult woman associated with unilateral rudimentary horn with ipsilateral renal agenesis and contralateral dermoid cyst

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ABSTRACT

Persistent urogenital sinus (PUGS) is an uncommon developmental cloacal anomaly, with Incidence of 0.6 in 10000 female births. Herein we depict the case of a 22-year of age lady who presented with Infertility for 16 months with dyspareunia and was found to have Persistent urogenital sinus associated with other urogenital anomalies as unilateral rudimentary horn with ipsilateral renal agenesis and contralateral dermoid cyst. The patient was successfully treated with the excision of the sinus, the rudimentary horn and the dermoid cyst.

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Un caso raro de seno urogenital persistente en una mujer adulta asociado a cuerno rudimentario unilateral con agenesia renal ipsilateral y quiste dermoide contralateral

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RESUMEN

El seno urogenital persistente (PUGS) es una anomalía cloacal del desarrollo poco común, con una incidencia de 0,6 en 10000 nacimientos de mujeres. Aquí representamos el caso de una señora de 22 años que presentó Infertilidad durante 16 meses con dispareunia y se encontró que tenía seno urogenital persistente asociado con otras anomalías urogenitales como cuerno rudimentario unilateral con agenesia renal ipsilateral y quiste dermoide contralateral. El paciente fue tratado con éxito con la escisión del seno, el cuerno rudimentario y el quiste dermoide.

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1. INTRODUCTION

Urogenital sinus happens because of the arrested migration of the Mullerian ducts from the Muller tubercle to the vestibule [1]. Urogenital sinus anomalies are characterized by the intersection of the urethra and vagina to frame a common channel of differing length with a solitary perineal opening. Thus, Persistent Urogenital Sinus (PUGS) can be grouped into high and low peculiarities as per the length of the common channel (>3 cm or <3 cm, separately). All in all, short urogenital sinuses are more common [2].

The presence of a urogenital sinus addresses a transient period of the normal development of the lower genital tract in the female fetus (3). It is one of the congenital disorders that are assessed to be 6 in each 100,000 female births (1). While PUGS can present as an isolated anomaly, it has additionally been associated with different diseases, including congenital adrenal hyperplasia or McKusick-Kaufman syndrome [4, 5].

A diagnosis of PUGS is challenging because of the complexity and uncommonness of this condition. As exhibited by the current case, the presence of a urogenital tract anomaly can be barely noticeable by clueless experts while investigating basically normally looking external genitalia with typical labial folds and a normally situated anus. Consequently, an intensive clinical assessment of the perineum is crucial as the finding of a solitary opening in the introitus is pathognomonic of a PUGS diagnosis. Herein I describe the first case of persistent urogenital sinus in a grown-up lady with a new level.

2. CASE REPORT

Nulligravida, 22 years of age lady presented to the outpatient clinic at Ain Shams University Maternity hospital for being infertile for 16 months with trouble in finishing sex (superficial dyspareunia). She had menarche at 12 years old; since that time, she had regular monthly cycle, on average changing three pads each day for 3 days with no history of circumcision. On general examination, she had typical female features with well-developed breasts (Tanner stage 5) and normal female hair distribution. Blood pressure was normal, and there were no palpable abdominal masses.

Local vulval assessment uncovered typical looking external genitalia with a solitary opening in the introitus (Figure 1). The patient was approached to return when she bleeds to distinguish the site of seepage of the menstrual blood. After fourteen days she bled and the menstrual blood was discovered to be coming out through the introital opening.

Outpatient Endoscopy was done through the opening in the introitus which uncovered a restricted passage that drives posteriorly to vaginal hole that prompts a typical vagina, cervix and one uterine horn. External urethral meatus was seen just over the opening in the introitus and front to the vaginal hole, where urethra-cystoscopy was done in the same setting uncovering typical urethra and bladder mucosa with no proof of fistula, endometriosis or any tumor.

Investigations as abdomeno-pelvic ultrasound and MRI was requested which uncovered unicornuate uterus at left side with rudimentary horn at right side along with missing right kidney which was affirmed later by IVP. The

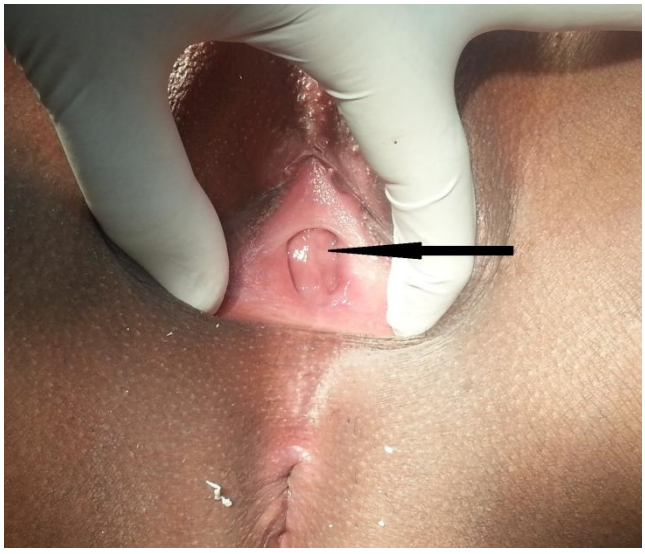

***Figure 1**: Introitus with a single opening (arrow).*

ultrasound and MRI found accidently Left ovarian well defined echogenic cyst with well delineated out-line occupying the uterovesical pouch measuring 7×7.5 cm with an impression of Dermoid cyst. The patient was checked on as booked and had hemoglobin 13 g/dL. Renal and liver function tests along with serum level of the hormones (FSH, LH, TSH, testosterone, estradiol) and ovarian tumor markers everything was normal. Urine analysis showed no evidence of urinary tract infection. A diagnosis of persistent urogenital sinus and a decision was taken to excise the septum of the sinus along with laparotomy at the same setting for excision of the ovarian cyst and rudimentary horn.

After taking Informed consent from the patient, repair was carried out with the patient in the standard lithotomy

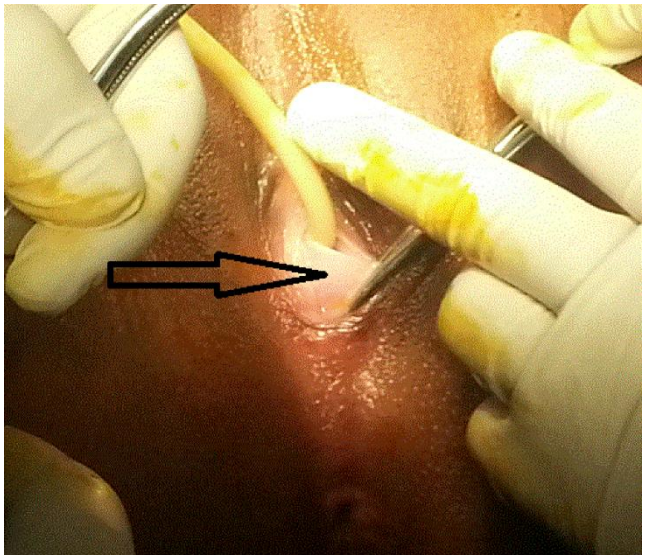

***Figure 2**: Arrow shows that the sinus extends below external urethral meatus and covering the vaginal orifice.*

position under general anesthesia, and 18 Fr foley catheter introduced to the urinary bladder and inflated with 10 mL normal saline. Holding the sinus wall was done using artery forceps (Figure 2) with incision and later excision of 3 mm thickness septum using monopolar diathermy knife and the edges of the excised sinus was sutured using interrupted absorbable sutures in a circumferential manner with Vicryl 2–0 sutures. The vaginal opening and external urethral meatus was uncovered separate in the vestibular region (Figure 3). The vagina was patent, admitting two fingers freely to a depth of 10 cm.

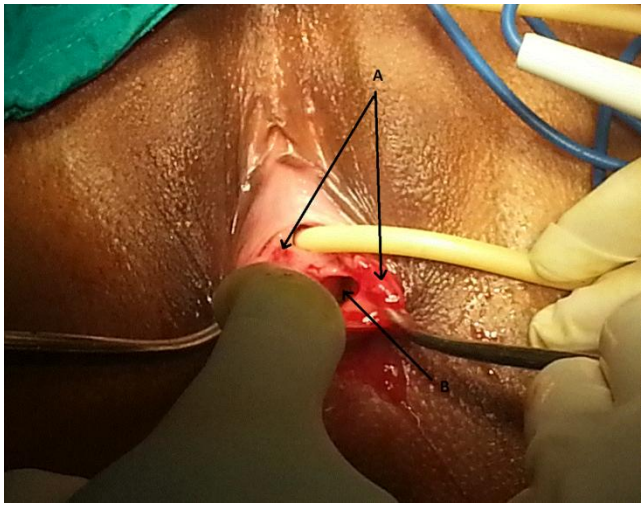

***Figure 3**: A: edges of the excised sinus; B: the vagina after sinus excision.*

Pfannenstiel incision was done at the same setting revealed left uterine horn of normal size with left ovarian cyst of diameters 8 × 10 cm that was excised and later proved to be dermoid cyst by histopathology. Right sided rudimentary horn was excised lifting both ovaries in place.

The patient was discharged on the second postoperative day. After one month the patient bled through the ordinary vaginal opening and there was no dyspareunia. After four months the patient got pregnant.

3. DISCUSSION

Anomalies of the female genital tract can present and be recognized at birth through adulthood. Advances in the field of developmental biology are giving clinicians a more prominent understanding of the origins of congenital anomalies like persistent cloaca and urogenital sinus [3].

PUGS is a developmental cloacal anomaly, which presents as a single common passage for urethra and vagina in female neonates. Neonates with urogenital sinus present with abdominal expansion and frequently have ambiguous genitalia, seldom the vulva might be normal. Examination of external genitalia, voiding cystouretherogram,

genitoscopy and genitography affirm the presence of persistent urogenital sinus with urinary retention in uterus and vagina [6].

This anomaly is uncommon, with Incidence of 0.6 in 10000 female births and is associated with a wide variety of syndromes. It is a common communication of vagina and urinary tract anywhere from urethral meatus to bladder, and they exit in the perineum as a solitary opening. Urogenital sinus can happen because of different reasons like iatrogenic causes, obstructed labor, trauma, and infections [7]. Molecular factors have been implicated as a possible cause by various workers [6].

An arrest in development of the Mullerian ducts at 9 weeks' gestation, after fusion with the urogenital sinus, manifests as a common urogenital sinus. A long urogenital sinus with a short vagina and high urethral opening will result if the defect happens at an early stage. On the other hand, a short urogenital sinus with an almost normal vaginal vestibule and low urethral opening will happen if the arrest happens late in development, similar to the current case. The genital portion of the urogenital sinus gives rise to the inferior third of the vagina and the vestibule, into which the vagina proper and the urethra open [8]. In western world, however, most instances of urogenital sinus happen within the context of disordered sexual differentiation (DSD) [9].

Mullerian tubercle induces formation of paired caudal endodermal outgrowths (Sino-vaginal bulbs) from the urogenital sinus. The cells inside the Sino-vaginal bulbs multiply to form a cord of tissue called the vaginal plate later canalized in a caudal-to-cranial direction to form distal vagina. The portion of urogenital sinus distal to the Mullerian tubercle undergoes exstrophy and everts to turn into the vestibule. As a result of this process, the urethra and vagina acquire separate openings in the vulva [10]. Embryological Explanation- Probably, in the current case, the arrest being developed was in the later stages, where the vagina and urethra gained a different opening yet both drain in a urogenital sinus with one opening on the vulva.

Here in the current case, I describe a very rare congenital anomaly in an adult female which initially made a great difficulty in its identification whether it is PUGS or low transverse vaginal septum with low urethrovaginal fistula [11]. In the later, the septum ought to be restricted distinctly to the vaginal hole, however here the septum stretches out anteriorly to underneath the level of the external urethral meatus covering all the vestibular area making a free communication between the urinary and genital system over the septum that structure the floor of the sinus, with a solitary vulval opening channels both menstrual blood and urine. This makes the case to be a variant of PUGS with a new level of communication between urinary and genital system.

In conclusion, I have herein described the first case of persistent urogenital sinus in a grown-up lady with a new level. It was associated with other urogenital anomalies in the form of unilateral rudimentary uterine horn with ipsilateral renal agenesis together with contralateral Dermoid cyst. The patient was successfully treated with excision of the sinus and associated rudimentary horn and dermoid cyst.

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