CASE REPORT

Female hypospadias presenting with urinary retention and renal failure in an adolescent: uncommon and late presentation with significant hidden morbidity

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SUMMARY

An 11-year-old girl presented with poor urinary stream with vaginal voiding since childhood. She had palpable bladder and raised serum creatinine ($671 \mu\text{mol/L}$). Suprapubic catheterisation was performed after a failed urethral attempt. Following catheterisation she improved symptomatically. She underwent cystoscopy through the suprapubic portal as a guide wire could be negotiated through bladder neck; external urethral meatus was found as a dimple higher up in anterior vaginal wall. Female hypospadias is usually an incidental diagnosis during catheterisation for various reasons. Diagnosis could be established only after antegrade cystoscopy. Anecdotal cases have been reported in the literature with similar presentation. Early diagnosis is essential as renal failure could be a late presentation if patients continue to void with stenosed meatus. Long-term follow-up is needed as these patients may need definitive management later on.

TREATMENT

She underwent antegrade cystoscopy through the suprapubic portal. On cystoscopy multiple trabeculations were found all over the bladder, and a dimple was appreciated at the bladder neck (figure 3) through which a guide wire was negotiated. Meatus was found higher up in the anterior wall of the vagina (figure 4). She was catheterised with an 18 Fr Foley catheter over a guide wire which remained in situ for 2 weeks.

OUTCOME AND FOLLOW-UP

At present the patient is voiding well after a successful voiding trial with hypospadiac meatus. She has been advised regular follow-up and a possible need of hypospadias repair later on.

DISCUSSION

Female hypospadias is a less reported congenital anomaly. Embryological defect appears to be failure

CASE PRESENTATION

An 11-year-old girl had history of poor urinary stream with vaginal voiding since childhood. She developed loss of appetite with nausea and vomiting since 1 month and went into acute urinary retention 3 weeks later. On evaluation, the bladder was palpable and urethral meatus could not be appreciated. A dimple was found at the anterior vaginal wall, but a guide wire could not be negotiated. She underwent suprapubic catheterisation after a failed urethral attempt.

INVESTIGATIONS

Her renal function was deranged (serum creatinine $671 \mu\text{mol/L}$). On ultrasound evaluation of the kidney, ureter and bladder, she was found to have a thickened urinary bladder with bilateral hydronephrosis. On micturating cystourethrogram grade 5 vesicoureteral reflux (figure 1) was found. MRI of the abdomen and pelvis showed similar findings, and no communication was found between the bladder and vagina (figure 2). Following suprapubic catheterisation, she improved symptomatically and her serum creatinine decreased from 671 to $61 \mu\text{mol/L}$ within 2 weeks. The renal DTPA scan showed left side split renal function of around 21% with a total glomerular filtration rate of 58 mL/min .

Figure 1 Micturating cystourethrogram showing a trabeculated bladder with grade 5 vesicoureteral reflux over the left side.

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Figure 2 MRI of the abdomen and pelvis showing no communication between the bladder and the vagina.

of fusion of the urethral folds bringing the meatal opening higher up in the vagina. Patients may be normal with no other associated disorders. But sometimes it may present with bicornuate uterus, vaginal septum, vaginal atresia and urogenital septum.¹ Diagnosis is mostly established during catheterisation.² Female hypospadias has been classified by various authors. Blum classified it into three categories. Type 1 is a longitudinal communication between the posterior wall of the urethra and the anterior wall of the vagina, type 2 is persistent urogenital sinus, where the vagina enters into the urethra and the hymen lies deep in the vagina, and type 3 urethra opens into the vagina proximal to the hymen.³ Our case was type 3 hypospadias with

Figure 3 Antegrade cystoscopy showing bladder neck through which a ureteric catheter was negotiated.

Figure 4 Meatus was found higher up in the anterior wall of the vagina.

stenosed meatus. Various ages of presentations have been found in the literature, from childhood to old age. Although recurrent urinary tract infection is more common in the sexually active group, older patients are usually diagnosed only during catheterisation. Here we have reported an unusual case of an adolescent who presented with a feature of renal failure and acute urinary retention. Diagnosis could be established only after antegrade cystoscopy. Anecdotal case reports are available for this uncommon entity.^{4 5} The cause appears to be voiding with stenosed meatus causing bladder outlet obstruction leading to obstructive uropathy. There are various methods of management once diagnosis has been established. As this patient presented to us for renal failure, she was treated accordingly and once the renal function normalised, she was kept for regular follow-up. These patients once sexually active are more prone for recurrent urinary tract infection and will require definitive management later on. Vaginal flap urethroplasty when needed can be performed with encouraging results.⁶

Learning points

- ▶ Female hypospadias may go unnoticed until there are complications, and catheterisation is needed for another reason.
- ▶ Early diagnosis is desirable as patients may develop renal failure due to obstructive uropathy.
- ▶ Clinical examination may reveal the diagnosis, but occasionally antegrade cystoscopy is needed.
- ▶ Long-term follow-up is needed to avoid complications including renal failure.
- ▶ Female hypospadias can be an occult congenital anomaly and neonatal examination along with various checks in infancy should be carried out to actively look for the three perineal openings clearly seen and documented, for early diagnosis and to avoid late presentation and the subsequent significant obstructive uropathy.

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