Pseudotail with closed neural tube defect and neurogenic bladder masquerading as posterior urethral valve: a case report

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Abstract

Background: Pseudotail with occult spinal dysraphism with neurological deficit is a rare phenomenon. Ignoring pseudotail as sign of occult spinal dysraphism may lead to catastrophe in diagnosis and further management.

Case presentation: We are reporting a case of a 30-month-old male child who presented with a tail-like structure at lower back and dribbling of urine since birth. On initial evaluation at some other hospital, he was misdiagnosed as posterior urethral valve and underwent fulguration of valve. However, dribbling of urine persisted after initial procedure. On evaluation at our center, he was found to have pseudotail with occult spinal dysraphism and neurogenic bladder.

Conclusion: Pseudotail with occult spinal dysraphism with neurological deficit is a rare phenomenon. Neurogenic bladder in such case can be misdiagnosed as posterior urethral valve. So, neurogenic bladder with pseudotail should be evaluated cautiously to avoid misdiagnosis and mismanagement.

Keywords: Human tail, Pseudotail, Posterior urethral valve, Occult spinal dysraphism, True tail

Background

A human tail is a rare congenital anomaly. The most common site of origin of human tail is the lumbosacrococcygeal region. Till date, around 60 cases have been reported in the literature [1]. Presence of human tail is a tell-tale sign of underlying spinal dysraphism and tethered cord [2] which may be associated with neurological deficit in the form of bowel/bladder incontinence and lower limb weakness. Neurogenic bladder is the most common urinary tract disorder in patients of spina bifida occulta [3]. We are going to present a case of pseudotail with lipomyelocele with neurogenic bladder masquerading as posterior urethral valve (PUV).

Case presentation

A 30-month-old male child was presented to our outpatient department with history of a tail-like structure at lower back since birth (Fig. 1). He also had history of urinary dribbling since birth and intermittent constipation after 6 months of age.

He had history of high-grade fever with dysuria at 1 year of age for which he was investigated in other government hospital and was diagnosed as posterior urethral valve (Fig. 2). After diagnosis of posterior urethral valve, transurethral fulguration was performed. Post fulguration, the urinary stream improved slightly but dribbling persisted. For urinary dribbling and tail-like structure at lower back, he consulted to our outpatient department. During visit at our institute, we advised and taught parents for clean intermittent catheterization (CIC). According to the parents, the tail was small at birth and gradually increased in size. On physical examination, the appendage was human tail-like, soft, well circumscribed, 4-cm long and 1-cm thick, and was attached to the back of the sacrum. It was non-tender and covered with normal skin. There was no voluntary movement in the tail. Per abdominal examination,
bladder was palpable and expressible. Anal opening was patulous and normal in position. Bilateral lower limb weakness was present with limping gait. Other general and systemic examinations were normal. No obvious other congenital defect was noted. There was no family history of other developmental or congenital anomaly. The possibility of a human tail with underlying occult spinal dysraphism associated with neurogenic bladder and bowel was considered. Magnetic resonance imaging (MRI) of the spine was done which showed a sacral spinal dysraphism with lipomyelocele formation with extradural extension of fat and low-lying (L4) tethered spinal cord and pseudotail (Fig. 3).

After routine investigation, diagnostic cystoscopy for bladder pathology and excision and repair of lipomyelocele with laminectomy was planned. On diagnostic cystoscopy, bladder was trabeculated with mild dilatation of posterior urethra, but no obvious posterior urethral valve was seen. After cystoscopy, patient was positioned to prone for excision of pseudotail. With longitudinal elliptical skin incision around pseudotail with laminectomy of L4 vertebra, pseudotail along with extradural fat was removed. Spinal cord detethering was also done. Postoperative period was uneventful. Per urethral catheter was removed on postoperative day 5, and patient was put on CIC. Patient was discharged on anticholinergic drug (oxybutynin) with advice of regular CIC. He was also advised high-fiber diet and laxative at night as a part of bowel management program. At 1 month follow-up, patient is doing well with no complaints of constipation and urinary incontinence. He is being planned for a follow-up with ultrasound of urinary tract to look for post void residue and isotope scan for the evaluation of renal scarring.

Discussion
Human tail is a midline protrusion in caudal region having combination of muscle, adipose tissue, and covered with skin [4]. During the 5th and 6th gestational week, the human embryo has a tail with 10–12 caudal vertebrae [1]. This tail usually regresses by reduction in the number of vertebrae and fusion, leaving behind the vestigial coccyx. It usually disappears by 8th week of gestation. On the basis of embryological origins, human tails
are classified into two categories: true tails and pseudotail [5]. True tails are formed by embryological remnants which include muscle, adipose, and connective tissues. However, no vertebrae are seen in true tails. Pseudotails are lesions that have an underlying pathology such as spinal column and coccygeal malformations (lipoma or teratoma) [6]. Lu et al. [7] suggested that surgical excision is sufficient for true tail. He also described pseudotails as tail-like structure which is associated with spinal dysraphism. The human tail is considered to be a marker of underlying occult spinal dysraphism [8]. Pang et al. suggested that human tail is associated with high rates of spinal dysraphism (50%) and tethered cord (81%) [9].

Bladder dysfunction is common in spina bifida. Vesicoureteral reflux may occur in up to 40% of children with spina bifida by age 5 year. Sixty-one percent of young adults with spina bifida experience urinary incontinence [10]. The lower urinary tract symptoms depend on patient age during intervention and the level of spina bifida. Suprasacral lesions generally result in neurogenic detrusor overactivity with detrusor-sphincter dyssynergia [11]. In case of bladder sphincter dyssynergia, oblique voiding cystourethrogram demonstrate an unusual posterior urethral dilatation due to bladder contraction against the incompletely relaxed external sphincter giving false impression of posterior urethral valve [12]. The radiological signs of posterior urethral valve in voiding cystourethrography are dilated and elongated posterior urethra with a ratio of more than 5:1 [13]. Other features are vesicoureteric reflux and trabeculated bladder with hypertrophied bladder neck. In index case, there was dilated posterior urethra due to neurogenic bladder which was wrongly interpreted and managed as posterior urethral valve.

Anticholinergic-like oxybutynin act as a bladder smooth muscle relaxant. They improve bladder dynamics by suppressing detrusor hypertonicity and hyperreflexia [14].
Institution of bowel management program with high fiber diet, laxatives, and enema improves the symptoms of fecal and urinary incontinence in patients with neurogenic bladder due to spinal dysraphism [15].

**Conclusion**

Pseudotail with occult spinal dysraphism and neurological deficit is a rare phenomenon. In index case, due to its rarity and ignoring pseudotail as a marker of underlying occult spinal dysraphism, it was misdiagnosed and treated as posterior urethral valve. So, neurogenic bladder with pseudotail should be evaluated cautiously to avoid misdiagnosis. It is recommended that a careful physical examination, along with proper radiographic imaging of lumbosacral vertebrae help in diagnosing true or false tail and associated spinal dysraphism.

**Abbreviations**

MRI: Magnetic resonance imaging; CIC: Clean intermittent catheterization; PUV: Posterior urethral valve

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**Authors’ contributions**

RJS—manuscript writing and organizing pictures. MMA—editing and labeling pictures. AK and RR—literature searching. AKS—proof reading of manuscript. BK—conceptualization of manuscript. All authors have read and approved the manuscript.

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Ethics approval and consent to participate
Not applicable

Consent for publication
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Competing interests
The authors declare that they have no competing interests.

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