



Association of Cowper syringocele with posterior urethral valve: First case report with review of literature

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Abstract We are reporting a case of 12 year old boy presenting to us as with obstructive voiding accompanying urinary tract infection and diagnosed as a case of posterior urethral valve (PUV) and Cowper syringocele. Up to our knowledge and search results from internet (pubmed, medline), there was no previous report in the literature regarding their association.

Key Words Posterior urethral valve; syringocele; bulbourethral gland “duct ectasia”.

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INTRODUCTION

Posterior urethral valves (PUV) make up the vast majority of congenital urethral obstructions. They occur in 1 in 8000 to 25,000 live births and make up 10% of urinary obstructions diagnosed in utero. PUV classically occurs only in males [1, 2]. Baskin and colleagues [3] in their recent review of literature concluded that the precise

origins regarding the anatomy and embryology of posterior urethral valves remain undefined. Cowper’s glands are two bulbourethral glands that lie superior to the perineal membrane and are invested in the broad base of the external urethral sphincter muscles [4]. They secrete clear mucous into the bulbous urethra during sexual arousal. A complete description of the gland was published in 1699 by Cowper [4]. Cowper’s syringocele or Bulbourethral gland “duct ectasia” is a cystic dilatation of the main duct of Cowper’s bulbourethral gland. The syringocele was first described by Fenwick [5].

CASE REPORT

A 12-year-old boy presented with symptoms of obstructed voiding weak stream and dysuria without any history of trauma or urethral instrumentation. Physical examination was essentially normal. Urinalysis showed 60 to 80 white blood cells per high power field and cultures were negative. Uroflowmetry was obstructive with a maximum flow rate of 8.5 ml. per second and an average flow rate of 5 ml. per second. The volume voided was 180 ml. Voiding cystourethrogram (VCUG) demonstrates a type I posterior urethral valve with a dilated posterior urethra, obstruction of the flow of contrast material from the bladder and a well delineated filling defect ventrally over proximal bulbar urethra (Fig. 1). Urethrocystoscopy revealed a thin membrane in the proximal urethra leaflets of which encroach into the urethral lumen along with a cystic swelling at the proximal part of the bulbar urethra near the external urethral sphincter, consistent with Cowper's syringocele (Fig. 2). We performed endoscopic fulguration of the

PUV with open excision of the syringocele. Patient followed up for cystourethroscopy and urinalyses a year later were normal. Retrograde urethrogram (RGU) showed normal urethral outline (Fig. 3).

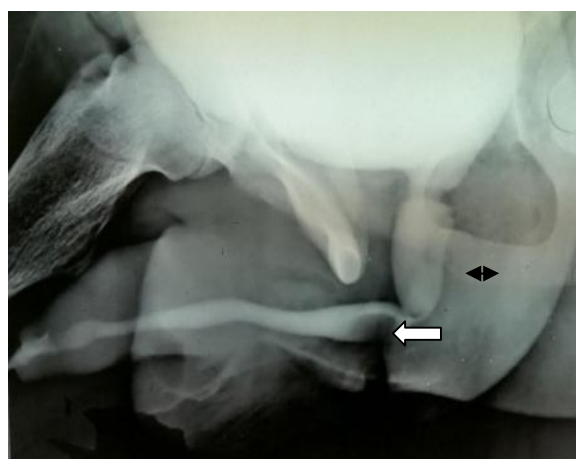


Fig. 1. Dilated posterior urethra (double arrow head) and obstruction of the flow of contrast material from the bladder and a well delineated filling defect ventrally over proximal bulbar urethra (block arrow).

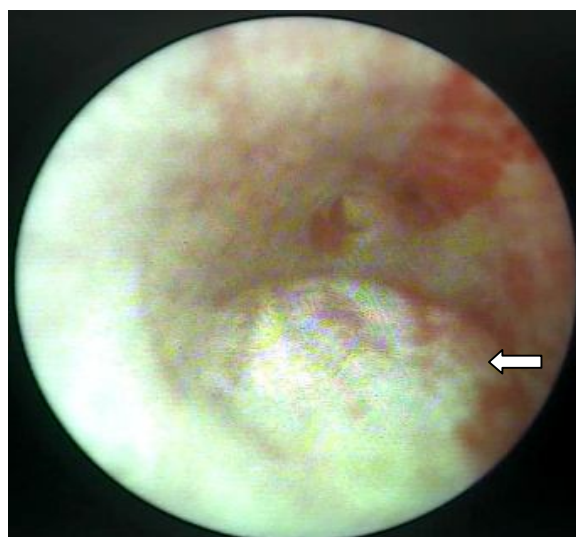


Fig. 2. Cystic swelling at the proximal part of the bulbar urethra (block arrow) near the external urethral sphincter.



Fig. 3. Post operative retrograde urethrogram after 9 months showed normal urethral outline.

DISCUSSION

The dilated Cowper's gland ducts are known as syringocele. Based on radiological and cystoscopic appearance, Maizels et al [6] differentiated 4 types of Cowper's syringocele: 1) simple syringocele, a minimally dilated duct; 2) perforated syringocele, a bulbous duct that drains into the urethra via a patulous ostium and appears as a diverticulum; 3) imperforate syringocele, a bulbous duct that resembles a submucosal cyst and appears as a radiolucent mass; and 4) ruptured syringocele, the fragile membrane that remains in the urethra after a dilated duct

ruptures. According to Bevers et al [7] the difference among the 3 communicating forms of Cowper's syringocele bears no clinical significance as they all present with the same symptoms hence they classified syringocele as either open or closed type. Open syringocele may present post-void incontinence, whereas closed syringocele may cause infravesical obstruction [4]. Our patient had a closed type of syringocele.

There are no data on the incidence of Cowper's syringocele. An incidence of 1.5% on cystograms has been reported in boys. Bevers et al [7] were of the view that due to unfamiliarity with the diagnosis Cowper's syringocele may be more common than currently realized.

Embryology of PUV though not clear is related to the persistence of the path of migration of the mesonephric duct distal to the Muller's tubercle and abnormal insertion of the mesonephric ducts into the fetal cloaca. Cowper's gland arise as a pair of buds from the endodermal epithelium of the urogenital sinus distal to the muller's

tubercle and extend backward paralleling the urogenital sinus and then penetrate the mesenchyme of the corpora cavernosa to become glandular, they open into the bulbar urethra. The genesis of Cowper's syringocele is not completely clear. It has been considered to be a congenital retention cyst of the intraurethral portion of the main duct of Cowper's gland. However, it can be acquired as well due to obliteration of the glandular duct in adults, perhaps caused by previous infection or trauma. Association of PUV with Cowper's syringocele in our case is neither explained clearly as a developmental anomaly nor can be ruled out.

Syringoceles may be entirely asymptomatic and found only incidentally, or they may obstruct urinary flow and present with symptoms of irritation or obstruction or both. The patient may also present with hematuria, urethral discharge, perineal pain or painful perineal mass. Patients with simple syringocele usually are asymptomatic or have a urinary tract Infection (UTI) and

require no surgical treatment, while in the perforate type urine may reflux into the cyst and lead to frequent UTIs. The imperforate type may lead to obstruction of the urethra, and a ruptured syringocele. The ruptured syringocele may act as a diverticulum where urine can stay, causing recurrent UTIs and post-void dribbling and may present as urethral diverticulum [1,8].

Obstruction due to PUV affects the entire urinary tract above the level of obstruction. Clinical presentations are age dependent with the more severely affected boys presenting earlier in life [9]. The majorities of boys who present later in life do so with urinary tract infection and/or voiding dysfunction and generally have more normal urinary tracts.

Our patient aged 12 yr presented with voiding difficulty and recurrent UTI with no history of trauma or instrumentation. The cause of the symptomatology is very difficult to differentiate between PUV or Cowper's syringocele.

Diagnosis of Cowper's syringocele can be confirmed with urethroscopy or retrograde urethrogram. Ultrasonography with 7.5 MH linear probes can be considered a useful imaging technique and a valid alternative to conventional

radiology [10]. PUV can be readily diagnosed with VCUG and urethral imaging. We performed VCUG and USG in our patient and successfully diagnosed PUV with Cowper's syringocele.

Classically syringocele require endoscopic deroofting except the asymptomatic simple syringocele. This procedure was successful in most of the cases reported in the literature. Surgical excision was reported in few cases. Catheter drainage followed by Primary valve ablation is the preferred initial surgical treatment for PUV.

In our case the combination of endoscopic valve fulguration and open syringocele excision was done in order to prevent the accidental damage to sphincter by endoscopic means. Had the

distance of syringocele well away from sphincter the endoscopic option would be the alternate choice. Follow up of the patient after 9 months showed no voiding difficulty, no episode of UTI and the RGU showed normal urethral outline (Fig. 3).

Cowper syringocele is a rare entity to be described and we are reporting the first case of association of both Cowper syringocele and PUV. The outcome of treatment depends upon the clinical status during presentation. As the presentations of both the entities are overlapping, the treatment decision should be taken carefully.

CONFLICT OF INTEREST

None declared.

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