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Superior vesical fissure in a case of prune belly syndrome with infravesical obstruction: Beneficial pop-off with a fatal congenital anomaly

Umesh Bahadur Singh, Shrikesh Singh, Tanvir Roshan Khan
Department of Pediatric Surgery, Dr Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India

ABSTRACT

A wide spectrum of renal, ureteral, and urethral abnormalities has been reported in a patient born with Prune belly syndrome (PBS). The obstruction in the urinary tract can be found anywhere from uppermost (pelviureteral junction) to lowermost (prostatic membranous urethra) urinary tract. The mesenchymal developmental arrest is the main embryological factor responsible for the major features of the syndrome like urethral abnormalities or gastrointestinal abnormalities. PBS patients with urethral obstruction commonly had a patent urachus, because of which such patients survived, early death usually occurred in those patients who did not had a patent urachus. We are reporting an interesting case of prune belly syndrome with superior vesical fissure (an exstrophy variant). To the best of our knowledge this was first reported case of association of two congenital anomalies having common embryological defect process. Presence of superior vesical fissure gives a new insight about the benefits gained by the child for surviving with such fatal congenital anomalies.

Key Words: Prune belly syndrome; exstrophy variant; superior vesical fissure.

Corresponding Author: Dr. Tanvir Roshan Khan
Department of Pediatric Surgery, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India,
E mail: doctrk@gmail.com
ORCID ID: 0000-0001-9155-0339
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Introduction

Children with Prune Belly Syndrome (PBS) can present with various urogenital abnormalities, ranging from severe urogenital and pulmonary problems incompatible with life to mild abnormalities, if any, that require no treatment other than orchiopexy. PBS patients with urethral obstruction commonly have a patent urachus, early deaths usually occur in those, who do not have a patent urachus. The vesicostomy is a known treatment modality for the patients of PBS with urethral obstruction. We are reporting an interesting case of PBS with Superior Vesical fissure. To the best of our knowledge this was the first reported case of association of two congenital anomalies, both arising from a defective development of the lateral plate mesoderm.

Case report

The index case was a four months male baby, antenatal unsupervised, born full term with

uneventful perinatal course. Since birth, the urine was continuously leaking from an abnormal opening about 2 cm below the umbilicus and the abdominal wall was flabby [Fig. 1].

Fig. 1. Clinical picture of patient.

External genitalia and anal opening were normal. The patient attended Out Patient department at the age of 4 months, weighing 6.3 kg and growth parameters were appropriate for the age of child. The meatal opening was normal size but 5 f catheter was not passing beyond the perineum into the bladder. Ultrasound of the urinary tract was the first investigation done that showed bilateral hydroureteronephrosis, megaureters and undescended testis. Micturating cystourethrogram was done by filling bladder from superior vesical fissure opening that showed hypoplastic urethra with suspected posterior urethral valve (PUV), but the ureteric reflux could not be assessed as bladder filling was not adequate [Fig. 2]. For further detailed evaluation of urinary bladder and abdominal musculature, magnetic resonance imaging (MRI) was done that showed bilateral megaureters with cystic dilatation of both kidneys, small capacity bladder communicating with skin surface below umbilicus, thin abdominal wall and bilateral undescended testis, features consistent with PBS and superior vesical fissure [Fig. 3].

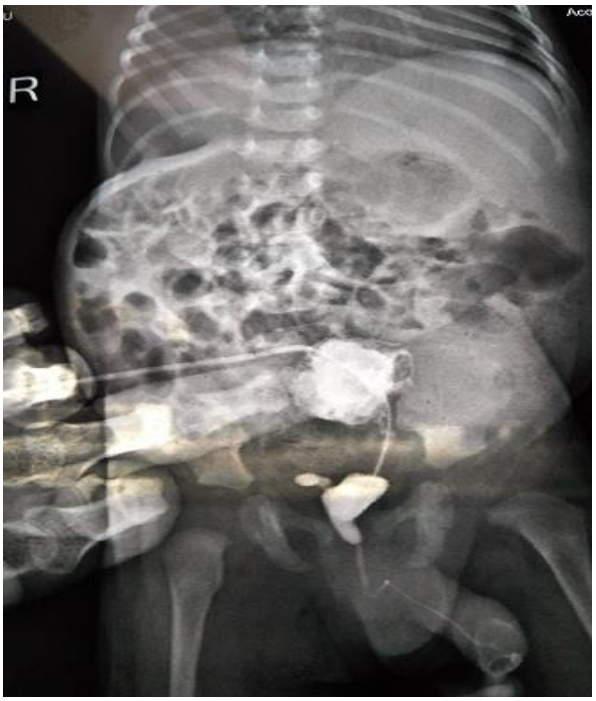

Fig. 2. Micturating cystourethrogram done by filling bladder from superior vesical fissure, showing hypoplastic urethra with posterior urethral valve.

Renal functions were preserved as value of serum urea and creatinine was within normal limits and dimercaptosuccinic acid (DMSA) scan was not showing any scarring in kidneys. The urine output was adequate and Radioisotope renography (ethylenedicysteine scan) showing unobstructed but slow urine drainage into the bladder with evidence of unobstructed upper urinary tract at the pelvi-ureteric junction and vesico-ureteric junction. Cystoscopy showed thick type I PUV with roomy posterior urethra and stenotic or hypoplastic anterior urethra from verumontanum to bulbar urethra distally, two

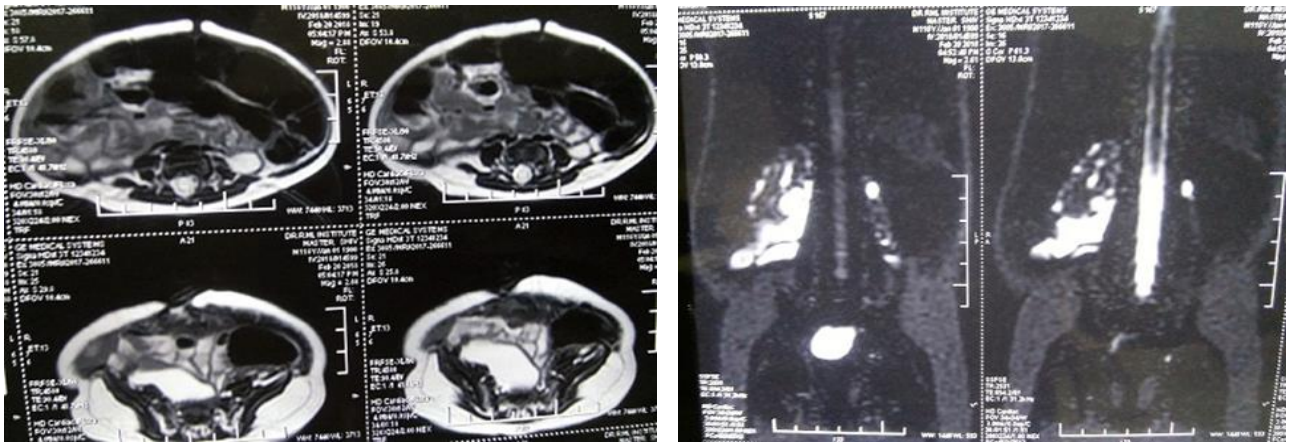

Fig. 3. Magnetic resonance imaging (MRI) showing bilateral megaureters with cystic dilatation of kidneys, thin abdominal wall and superior vesical fissure.

normally looking ureteric orifice and small capacity smooth outlined bladder. PUV fulguration was done successfully and progressive urethral dilatation (four times at 2 weeks interval) was done to improve urethral calibre. Subsequently, when we were satisfied with unobstructed bladder outlet and urethra, orchiopexy and vesical fissure closure was done at the age of 6 months. Now, patient is passing urine in good stream now with frequency at about 2 hourly interval, and is on uroprophylaxis and monthly follow up (6 month at present) for any urinary tract infection (UTI) or renal scarring. Recently done cystoscopy showed some improvement in bladder capacity and in next two months we have planned micturating cystourethrogram to look for further increase in bladder capacity or any ureteric reflux. Otherwise ureteric implantation can be done if megaureters are posing risk of backpressure to the kidneys or obstructing the urine drainage into bladder or causing recurrent UTI with renal scarring. Abdominoplasty can be done later on when child has grown and survive with correction of urinary system anomalies.

Discussion

The reported incidence of PBS is 1 per 30,000 to 50,000 live births. Of all PBS cases, less than 5% occur in females. Children with incomplete forms of this condition may present with typical abdominal wall changes but no urologic or testicular manifestations [1]. Some previous studies suggested that genetic mechanism was involved in the phenotypic presentation of prune-belly syndrome (X-linked inheritance), but some recent reports do not support X-linked inheritance [1]. Sporadic cases have also been associated with other chromosomal abnormalities, including Turner's syndrome and trisomies 13 and 18 [2]. Obstruction at the junction of the glanular and penile urethra is suggested by some recent studies and these urethral obstruction or microurethra were commonly associated with patent urachus [2]. Urethral obstruction is a surgical emergency to be addressed. Passerini-Glazel et al. [3] suggested progressive urethral dilation for urethral hypoplasia associated with PBS. Vesicostomy can also be a temporary urine diversion method for these patients. Eighty one percent of PBS patients may also have elongated, dilated and tortuous megaureters [2]. Different school of thought exists regarding reconstruction of the urinary tract as proponent of conservative treatment argues that nonobstructive system can be managed conservatively with present suppressive therapy. Proponent of aggressive

therapy points to Waldbaum and Marshall's review of 56 patients, in which 70% died and 16% were gravely ill [4]. Regardless, ureteral reimplantation in patients with PBS, the results may not be encouraging as many of these reimplantation patients developed ureteral strictures of the distal reimplanted ureters [5].

The mesenchymal developmental arrest, which accounts for the major features of the PBS, may also explain the urethral abnormalities. Gastrointestinal abnormalities (affects 30% of PBS patients) include malrotation, atresia, stenosis and rarely reported anorectal agenesis and omphalocele, however superior vesical fistula and extrophy has not been reported till date. Marshall and Muecke [6] have suggested embryological basis of exstrophy– epispadias complex as abnormal cloacal membrane prevents the lateral mesoderm from migrating and closing towards the midline and disrupts the development to the lower abdominal wall [7]. Superior vesical fissure is the rarest form of exstrophy variants having abnormally low placed umbilicus and infra-umblical opening into the bladder. In some cases, the musculature of the abdominal wall is totally absent, but most commonly there is uneven involvement. The medial and inferior muscles typically are most deficient [8].

Urethral obstruction in severe cases of PBS proves fatal to a neonate and the babies born alive mostly have a patent urachus, which preserves the upper urinary tract from early damage in intrauterine life. The decision of aggressive surgical correction of urinary tract in these children should be based on the clinical presentation and not solely on radiographic appearance. Before the surgical correction of these multiple urinary anomalies, detailed anatomical and functional evaluation of genitourinary system helps to priorities the sequence of correction of anomalies. The management of hypoplastic urethra, megaureters and small capacity bladder poses the great challenge to the urologist and warrants close follow up of long duration. In this neglected and late presented patient, in spite of severe lower urinary tract anomaly, the patient was able to survive, mainly due to preserved renal functions and unobstructed upper urinary tract. In classical PBS, the bladder is thin walled, of large capacity without trabeculations, however in the index case, due to superior vesical fissure, the domain of bladder had not grown.

Conclusions

Superior vesical fissure can be rarely seen in a neonate born with PBS, which may be a good sign for the immediate survival of patient born with severe congenital anomalies of PBS. The preserved renal functions in these patients may give time window to the clinician for further evaluation and planning of staged management.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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