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Management issues in a patient of posterior urethral valve with Renal rickets

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Abstract

Posterior urethral valves (PUV) are the most common cause of lower urinary tract obstruction in male neonates. The course and treatment of the disease can further be complicated if it coexists with Renal rickets. We present a rare case of PUV complicated with Renal rickets, which was managed successfully at our institution.

Keywords

Posterior urethral valve; renal rickets.

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Introduction

Posterior urethral valves (PUV) are the most common cause of mechanical infravesical obstruction in boys; the reported incidence is 1 per 8,000 to 1 per 25,000 live births [1]. Despite its rarity, PUV is the cause of renal insufficiency in approximately 10-15% of children undergoing renal transplant, and about one third of patients born with PUV progress to end-stage renal disease (ESRD) [2]. As the child grows, renal metabolic demand increases proportionately. However, renal failure cause altered calcium and phosphate metabolism which results in deficient mineralization. In children, demineralization at growth plate results in various musculoskeletal deformities, known as Renal rickets. Renal rickets associated with PUV are very rare, and it was first described by Derow et al in 1939 [3]. Various musculoskeletal deformities along with renal failure pose multitude of

challenges both for the anesthetist and the surgeon. We report a case of PUV complicated with Renal rickets who were treated at our institute.

Case Report

A 12 year old boy, a follow up case of PUV, admitted to our hospital for valve fulguration and vesicostomy closure. There was delayed milestone of development. At 3 years of age, he developed dribbling of urine and investigation revealed PUV and, vesicostomy by a pediatric surgeon was performed. Additionally, he was operated for right undescended testis. When he came to our clinic stunted growth, weight 18kg, large head circumference with frontal and parietal bossing, non eruption of teeth, hypoplasia of dental enamel, wide wrist joint, enlargement of costochondral junction (Ricket rosary) and increased anteroposterior diameter of thoracic cage was obtained at physical examination [Fig. 1]. Biochemical parameters such as serum calcium (8.6 mg/dl) and phosphorous (4.1 mg/dl) were within the normal range and serum creatinine (2.7 mg/dl) was increased. Serum alkaline phosphatase and serum PTH levels were high. Cystourethrogram revealed dilated prostatic urethra together with grade IV right VUR.

At pre-anesthetic evaluation, Malampatti Grade 4 airway, retrognathia, high-arch Palate, decreased muscle tone and sluggish deep tendon reflexes were noted. A difficult intubation was anticipated.

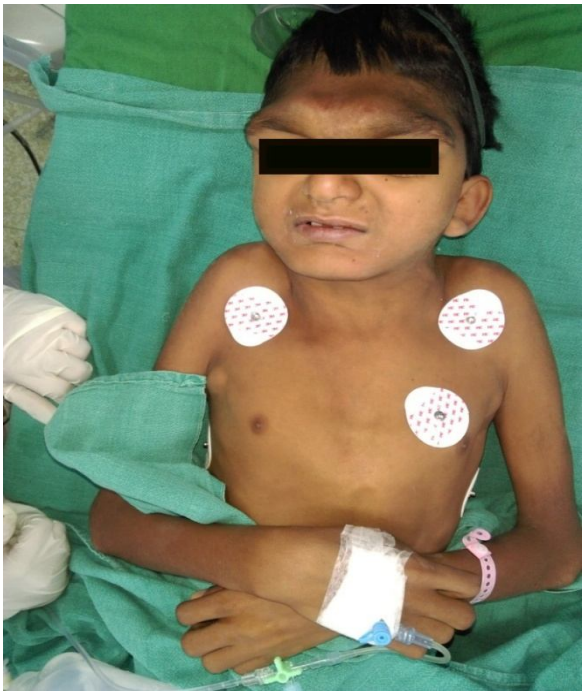

Fig. 1. Appearance case with posterior urethral valve and Renal rickets.

After taking informed consent, the patient was shifted to operating room. Continuous ECG, NIBP (non invasive blood pressure) and SpO2 (pulse oximetry) were monitored. Intravenous access was established by 20G veinflon over dorsum of right hand. Preoxygenation with 100% Oxygen was given for 2-3 minutes. Sevoflurane was used for induction. Flexible fiberscope (PENTAX®, Europe GmbH) passed into

glottis under direct vision through oral route. Intubation was performed by mounting a 4.5 mm ETT over the fiberscope. Muscle relaxation was achieved by Atracurium Besylate (0.6 mg/kg). Anesthesia was maintained by Sevoflurane and Nitrous Oxide. Cystopanendoscopy revealed Type I PUV. Fulguration was done by pediatric resectoscope at 5,7 & and 12 O clock position. Vesicostomy was closed by 2-0 vicryl. Duration of surgery was 45 minutes. On conclusion of surgery and upon initiation of spontaneous breathing, reversal was done by Neostigmine and Glycopyrrolate. Patient then shifted to recovery room for 3 hrs and oxygen given by Venturi-Mask. Post operative course was uneventful and child was discharged 5 days after the operation.

Discussion

PUV is the most common cause of 16.5% of all pediatric renal transplants in USA [4]. The severity of this disease is correlated by the age that symptoms are first seen. These patients may die at born or in early infancy. Deficiency in mineralization at the growth plate of these children causes a clinical condition known as Renal rickets.

Renal rickets was first described by Barber et al in 1921[5]. He defined as a condition of children marked by stunted development that often associated with bone deformity and symptoms of uremia, due to chronic renal insufficiency. It is classically seen in patients with distal renal tubular acidosis.

Development of Renal rickets in patients with PUV was first described by Derow et al in 1921[3]. Since then, various literatures have supported this rare association. Gangopadhyaya et al, followed 548 patients with PUV over 5-20 years and reported incidence of Renal rickets of 6% [6].

Because of various skeletal deformities and associated renal insufficiency, these children present various challenges to anesthetist and surgeon for elective surgery. These difficulties include difficult intubation, fluid and electrolyte disturbances, modification of drug dosages and immune suppression that increases the risk of postoperative sepsis.

Practical approach to these children should include correction of fluid and electrolytes, i.e serum sodium, potassium, calcium and phosphorous levels to lie within normal range. When difficult intubation is anticipated (like in our case), flexible fiberscope can be kept ready before hand. Sevoflurane Nitrous Oxide combination is preferred in renal failure patients. Prognosis is variable depending upon the age of the child, duration of renal failure and severity of the defect. By resolution of

hyperparathyroidism, renal osteodystrophy and metastatic calcification are often noted following successful kidney transplantation [7]. Limb deformities are usually irreversible and require corrective osteotomy and limb detorsion.

Conclusion

Diagnosis of Renal rickets should be suspected in any patient of PUV presenting with multiple skeletal deformities and chronic renal failure. Successful management of the condition requires careful evaluation, monitoring and multidisciplinary approach involving anesthetists, nephrologists, pediatric surgeons and orthopedists.

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