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Urinary bladder neck diverticular stone in patient with multiple congenital anomaly (*Jarcho-Levin syndrome*)

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ABSTRACT

Urinary bladder stone usually is a manifestation of other pathological process, including foreign object or micturition dysfunction. Urinary bladder diverticula can cause urinary stasis that leads to urinary bladder stone. We present a very rare case which is bladder neck diverticula stone in 36-year-old male with multiple congenital anomaly (Jarcho-Levin syndrome) with stone in urinary bladder neck diverticula and the management in our treatment facility.

Introduction

Urinary tract stone is the third most common disease in urology after urinary tract infection and prostate gland hyperplasia. Urinary tract stone is a prevalent disease worldwide, but this disease is more commonly found in area also known as stone belt, in which Indonesia is included. Of all the cases of urinary tract stone, about 5% of them is urinary bladder stone.[1,2]

Urinary bladder stone is usually a manifestation of other pathological process, including micturition dysfunction or foreign object. Micturition dysfunction leads to urinary stasis. Foreign objects such as urinary catheter and ureteral stent can also cause the formation of urinary bladder stone.[2]

One of the causes of urinary stasis that leads to urinary bladder stone is the presence of a urinary bladder diverticulum. Urinary bladder diverticulum occurs when bladder's urothelium herniated into the muscular layer. Outer diverticulum wall consist of irregular and non-functional smooth muscle fibers, which leads to incomplete voiding and urinary retention that can be seen through radiologic imaging.[1,3–5]

Bladder diverticulum is classified into 2 types, congenital and acquired bladder diverticulum. Congenital urinary bladder diverticulum is the type that causes urinary stasis and bladder diverticula stone. In this case, we present a very rare case about a bladder neck diverticula stone in 36-year-old male with multiple congenital anomaly (Jarcho-Levin syndrome), and how the management was taken in our facility.[1,3]

Jarcho-levin syndrome is defined as collection of symptoms or congenital disorder that consist of many criterias, including chest wall abnormality or chest asymetry, costal bone abnormalities, scoliosis and talipes, neural tube spinal defect (meningomielocele), spina bifida, and could also be accompanied by few urinary tract abnormalities.

Case presentation

36-year-old male, Indonesian, came into our clinic with a history of unintentional passing of urine from the urinary bladder since 2 years before hospitalization. Since 4 years ago, patient felt an involuntary leakage of urine and distention in his lower abdomen. Patient also stated that change of position can cause the urine flow back to normal. Boil-like lump appeared in hemiscrotal area around 2 years before hospitalization. Later, the lump burst spontaneously releasing pus mixed with urine.

Since birth, patient has several congenital anomaly including costal, vertebra, and both leg-bone abnormalities (Fig. 1). Patient also could not feel any sensation of voiding or defecating, which was thought to originate from meningomyelocele he had since his birth. The meningomyelocele was removed surgically when he was 3-years-old.

Physical examination showed fistulas in both sides of his hemiscrotum (bilateral). There were no signs of edema, fluctuation, or any hyperemic areas; and there is a hard mass palpated at his hemiscrotum bilaterally (Fig. 1). From digital rectal examination, we found his sphincter anal tone is adequate, a smooth mucosal lining, non-collapse ampulla, with prostate volume less than 20 gr and a soft consistency. Radiology examination showed a large bladder stone in the pelvic region. Urethrography and cystography showed there was a diverticulum, and signs of contrast extravasation near the bladder neck area (Fig. 2). Fistulography also showed a contrast extravasation from the bladder

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neck area into the scrotal area (Fig. 3).

Patient has undergone diagnostic urethroscopy, open vesicolithotomy, bladder mucosal biopsy and an open cystostomy procedure. During urethroscopy, the urethra and urethral external meatus was found to be within normal limit. Stone was found in the urinary bladder neck diverticulum. Open vesicolithotomy procedure showed a 5 × 4 cm solid yellowish stone. After the stone was removed, permanent cystostomy catheter was placed, and surgical wounds were closed. Patient was hospitalized for five days post-operative, and was sent home with good condition and an effective permanent cystostomy.

Discussion

A urinary bladder neck diverticular stone in patient with Jarcho-levin syndrome is a rare case and still hard to find, and this is the first case in our hospital. Patient was reported to have several other congenital disorder, including costal bone asymetry and shape abnormality, leg bone and leg joints abnormality, meningomielocele, and other abnormalities which included in Jarcho-Levin syndrome criteria. Patient with Jarcho-Levin syndrome often accompanied with a number of concurring defect such as urogenital and anal anomalies, hernias, heart malformations, neural tube defects, and lower limbs defects. In this patient, he developed neurogenic symptoms due to neural tube defects (history of meningomyelocele). The symptoms include numbness and paresthesias in his lower extremities, and neurogenic bladder which causing chronic retention and urinary stasis that leads to the formation of bladder stone and urinary diverticulum.

Urethroscopy showed normal urethral orifice, there was no fistula opening along the urethral lumen, and a yellowish stone with a size of 5 × 4cm was present in the urinary bladder neck diverticulum. During open vesicolithotomy procedure, diverticulum was seen at the urinary bladder neck. Cystostomy was done afterwards, and surgical wounds were closed.

Post-operative wound care was perfomed for 5 days, and pelvic drain was removed 2 days post-operative. Cystostomy catheter tube was functioning effectively, with daily urinary production between 1500 and 2000 cc per day, yellowish in color. Patient was sent home with good condition and no complaint.

The cause of this patient's urinary bladder diverticula is most likely anatomical congenital anomaly. The patient is 36-year-old, which correlates to many studies that most urinary tract stones commonly appear among patients from 30 to 60 years of age. This patient also have a urinary bladder diverticulum, which has been explained before in literatures, that urinary bladder diverticula could cause urinary stasis that leads to stone formation.[1,3]

Conclusion

In this patient, the manifestation of urinary bladder neck diverticula is urinary stasis that leads to urinary bladder stone formation. The principle management to prevent the recurrence of urinary bladder stone is to manage the obstruction, and to control both the intrinsic and

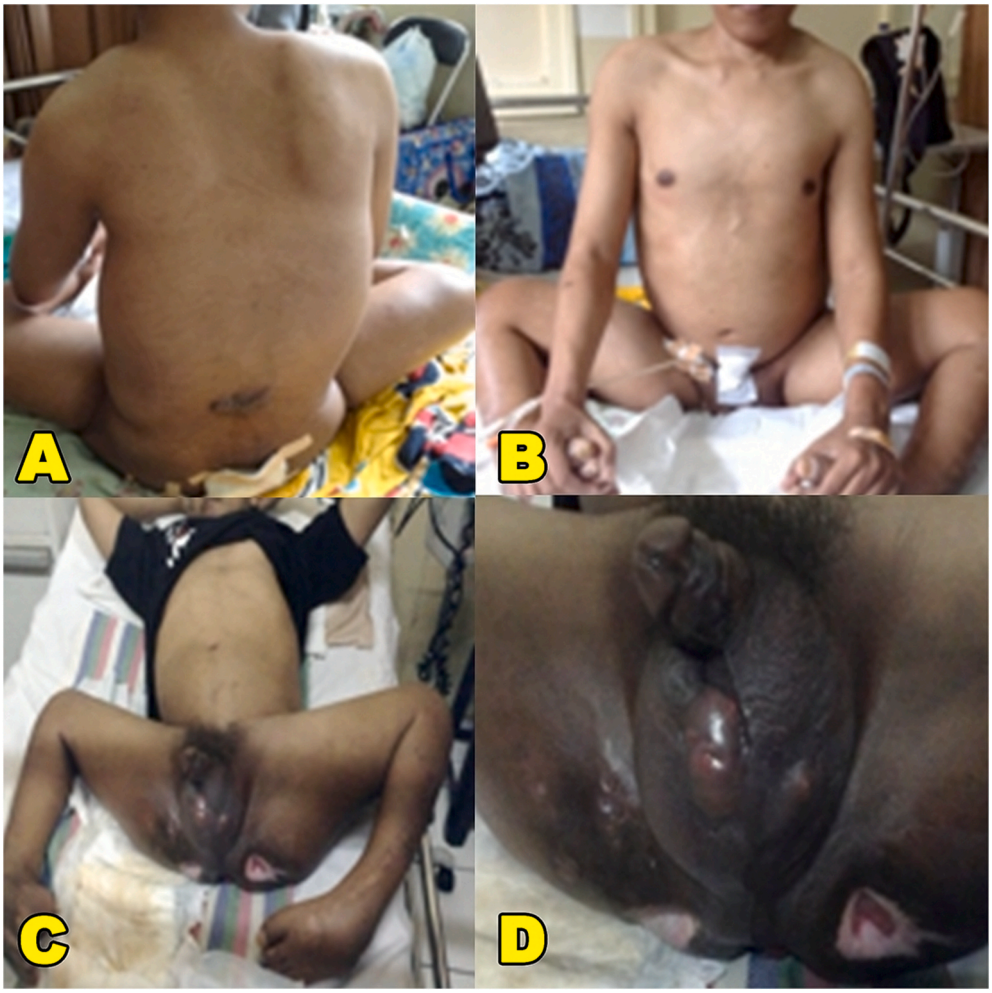

Fig. 1. A and B. Clinical pictures of patients showed several congenital anomaly including costal, vertebra and both leg bone abnormalities; C and D. Fistula in bilateral hemiscrotum.

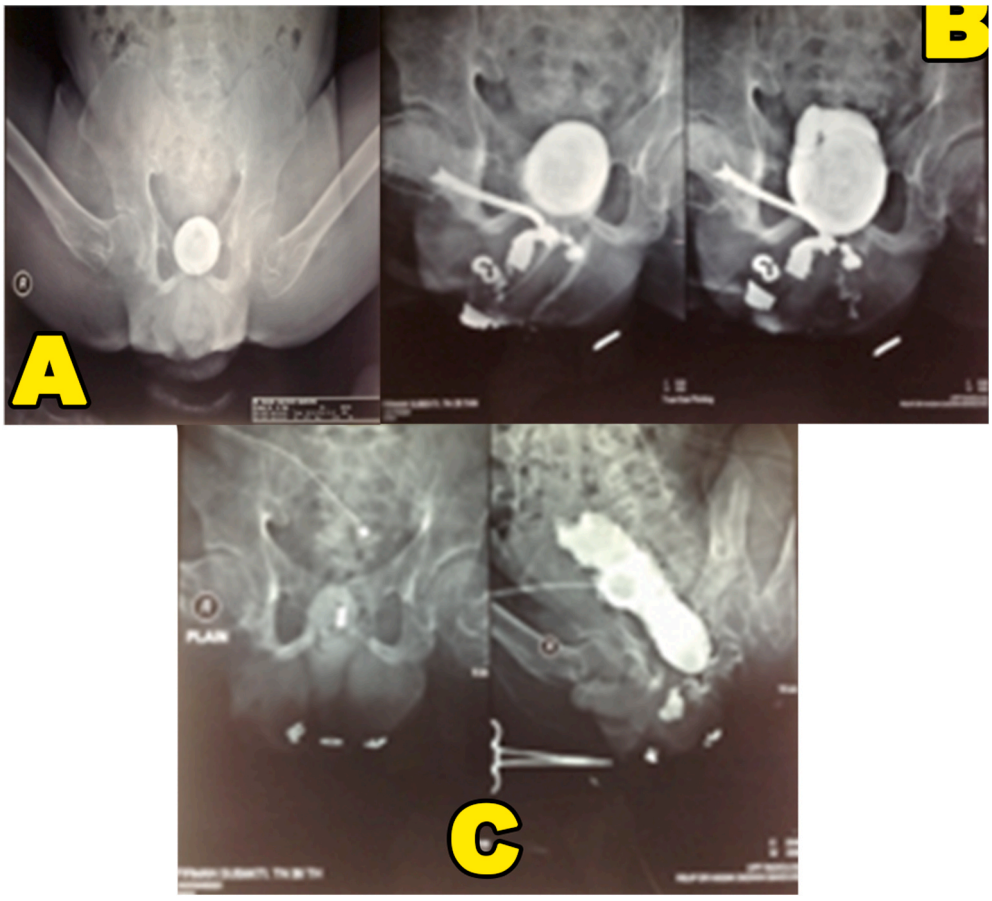

Fig. 2. A. Large bladder stone was found in the pelvis region. B and C. Urethrography and cystography showed there was contrast extravasation in near bladder neck area.

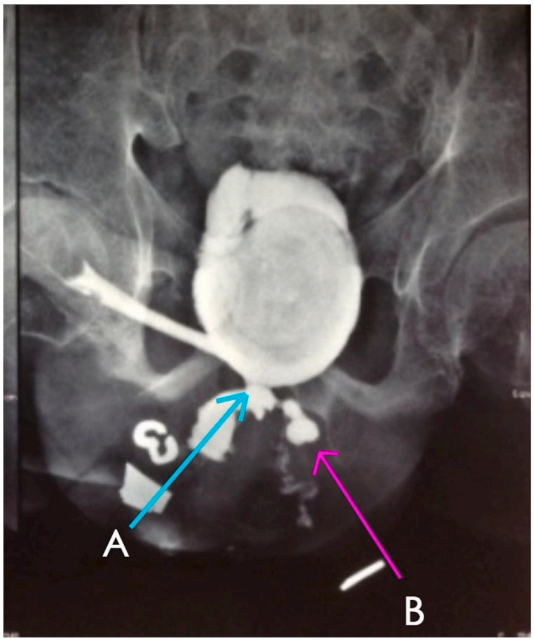

Fig. 3. A. Diverticulum which emanating from bladder neck area; B. Fistula coming from bladder neck area into scrotal area.

extrinsic risk factors. Management of choice including close observation, endoscopic management and open surgical therapy.

Source of support

None.

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