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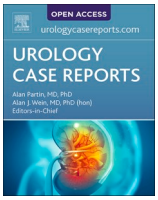

Endourology

Simultaneous tubeless supine percutaneous nephrolithotomy and cystolitholapaxy in a patient with spina bifida

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ABSTRACT

Spina bifida is a congenital condition that often leads to significant urological complications, including an increased risk of kidney and bladder stones. Performing percutaneous nephrolithotomy (PCNL) on patients with spina bifida presents unique challenges due to the anatomical deformities. We present a case of a spina bifida patient with right staghorn stone and bladder stones in a previously augmented bladder. Simultaneous tubeless supine PCNL and cystolitholapaxy was successfully performed. This case highlights the versatility of the supine position in managing complex stones in patients with spina bifida.

1. Introduction

Spina bifida is a birth defect affecting 1 to 10 per 1000 live births worldwide.[1] It often leads to numerous urological complications, including neurogenic bladder, recurrent urinary tract infections (UTIs), vesicoureteral reflux and kidney failure. Patients with spina bifida are also at a significantly higher risk of developing kidney and bladder stones. The lifetime incidence of urolithiasis is 2.7–3.6 times higher compared to the general population, and the risk of bladder stones is even more pronounced, ranging from 22.5 to 33.3 times higher.[2]

Contributing risk factors include UTIs, impaired urinary drainage, metabolic anomalies, and the incorporation of intestinal mucosa into the urinary tract.

Percutaneous nephrolithotomy (PCNL) is the gold-standard treatment for kidney stones larger than 20 mm. PCNL can be performed safely in either the prone or supine position. The supine position offers several advantages for both anesthesiologists and urologists, including the convenience of a less time-consuming single patient positioning and the ability to access the urinary tract from both anterograde and retrograde directions simultaneously.[3]

Performing PCNL on patients with spina bifida presents unique challenges due to the anatomical deformities. Additional complicating factors include a large stone burden, scoliosis, short stature, restrictive lung disease, and the presence of spinal hardware. Despite these difficulties, PCNL remains a feasible option. However, complication rates are higher (20–64 %) and stone-free rates (SFR) are lower (9–60 %) in spina bifida patients compared to the general population.[4]

In this report, we present a case of a spina bifida patient with both kidney and bladder stones, where single-tract tubeless supine PCNL and transurethral cystolitholapaxy (TUCL) were performed simultaneously by two surgeons.

1.1. Case presentation

The patient is a 34-year-old female with a history of spina bifida. After birth, a ventriculoperitoneal shunt was placed due to hydrocephalus. At age 6, bladder augmentation was performed using stomach tissue, along with a Mitrofanoff appendicovesicostomy. The patient performs self-catheterization five times a day. Additionally, corrective spinal surgery and PCNL on the right side was performed previously.

The patient was referred to our department after a routine follow-up ultrasound revealed a staghorn stone in the right kidney and two bladder stones. Physical examination showed a somatically retarded, immobile, low-stature patient with a BMI of 17,9. A low-dose CT scan (Figs. 1–3) confirmed a staghorn stone in the right kidney (stone volume: 2000 mm^3, density: 1000 HU) and two stones in the augmented bladder, measuring 33 × 34 mm and 7 × 8 mm (stone volume: 1500 mm^3, density: 700 HU). The stone volume was calculated using the GE AW Server 3.2 Ext. 3.0. According to the Guy's stone score nephrometry system, the patient was classified as score 4.

The anesthesiology evaluation assigned an ASA (American Society of Anesthesiologists) score of 3 and a Mallampati score of 3, indicating a potentially difficult intratracheal intubation. Laboratory tests, spirometry, chest X-ray, and urine culture were also performed. Preoperative

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Fig. 1. CT scout view: the right staghorn stone, bladder stone, spinal hardware and ventriculoperitoneal shunt are shown.

Fig. 2. Coronal plane of the CT scan: right staghorn stone and bladder stone in the augmented bladder are shown.

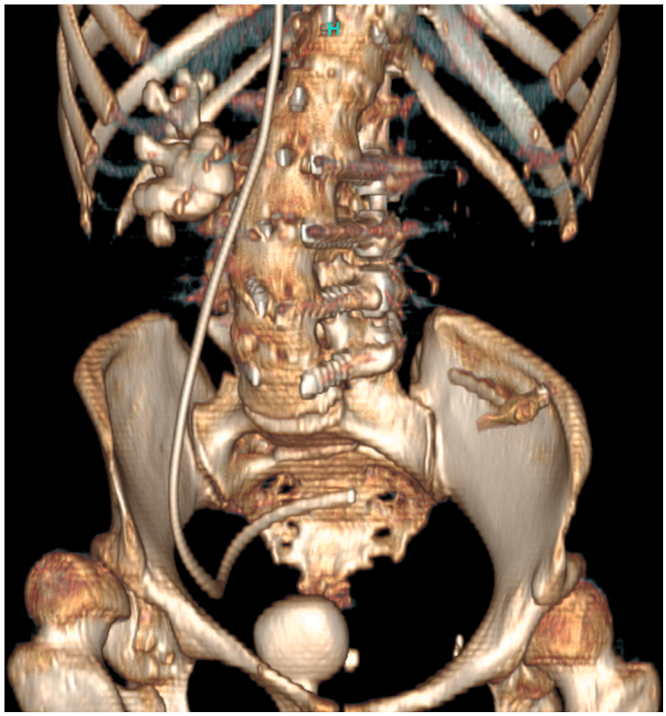

Fig. 3. 3D reconstruction of the CT scan.

laboratory tests revealed anemia (Hgb: 109 g/L), but kidney function was normal (creatinine: 43 μmol/L). Due to the presence of *Escherichia coli* in the urine culture, prophylactic broad-spectrum antibiotic was started the day before surgery.

After setting up the operating room (Fig. 4), under general anesthesia, the patient was positioned in the Barts flank-free supine position with a 15-degree tilt, with special attention to protecting pressure points. The posterior axillary line, the rib line, and the iliac crest were marked on the patient. A 19 Ch rigid cystoscope was inserted and the right ureteral orifice was identified. A 5 Ch open-ended ureteral catheter was placed on the right side. An ultrasound-guided, fluoroscopically adjusted puncture of the lower calyx was performed with an 18-gauge needle under the posterior axillary line, and a 0.035 mm hydrophilic guidewire was advanced to the ureter. Teflon dilators were used for dilation, followed by the placement of a 28 Ch Amplatz sheath and a 25 Ch nephroscope (Olympus™). Lithotripsy was performed using the ShockPulse (Olympus™) device. From the single access through the lower calyx, all of the calyces could be reached, eliminating the need for additional tracts.

Simultaneously, a 25 Ch nephroscope and ShockPulse lithotripter

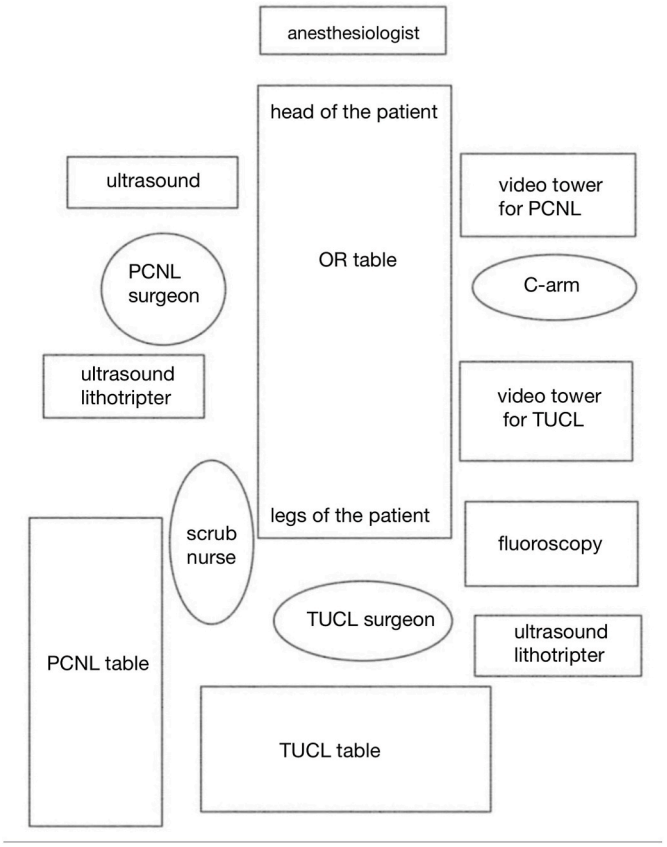

Fig. 4. Setup of the operating room.

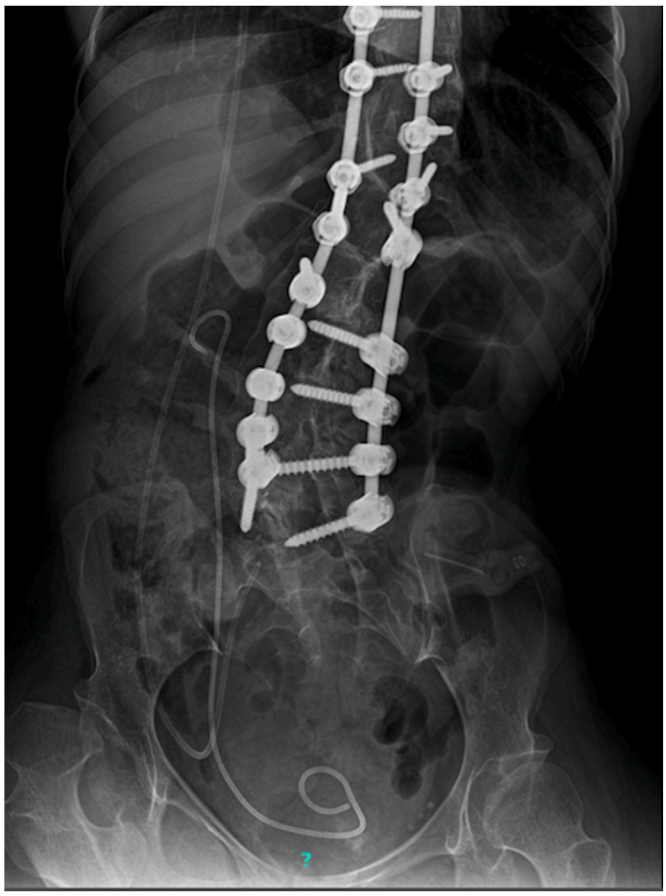

Fig. 5. Postoperative KUB X-ray.

were employed for transurethral cystolitholapaxy by the other surgeon. At the end of the procedure, a 26 cm, 6 Ch double J stent was placed retrogradely. Nephrostomy tube was not inserted. Local injection of diluted bupivacaine was administered to the percutaneous tract to decrease postoperative pain. The operative time was 65 minutes, and the estimated blood loss was approximately 590 ml (the López-Picado's formula was used for the calculation). There were no anesthetic complications during the surgery.

Postoperatively, anemia (Hgb: 84 g/L) was observed, which was expected, and 2 units of packed red blood cells were transfused. Postoperative creatinine level was normal (58 μmol/L). On the first postoperative day, a kidney-ureter-bladder (KUB) X-ray was performed to confirm the correct position of the double J stent (Fig. 5). The patient was discharged home without any complaints on the second postoperative day. The double J catheter was removed after 2 weeks. Stone-free status was confirmed with low-dose CT after 1 month.

2. Discussion

Considering that spina bifida patients are not standard patients, performing PCNL in these cases can be challenging with many potential pitfalls starting from the anesthesiological management, to patient positioning and technical aspects of percutaneous access. Surgery planning is particularly crucial for patients with spinal deformities. Despite accurate planning, the results can be suboptimal, and the complication rate can be higher. According to a PCNL series in patients with spinal deformities published by Goumas-Kartalas, the complication rate was higher (40 %), compared to the general population.[5] Additionally, there was a higher rate of severe renal hemorrhage, with a 20 % transfusion rate due to difficult puncture and access.

From an anesthesiological perspective, the presence of scoliosis and restrictive lung disease can complicate intubation and ventilation. Performing PCNL in the prone position exacerbates these issues, further reducing the lung capacity, causing ventilation difficulties, and increasing the risk of complications. Therefore the supine position is preferable as it offers better airway control and a more natural position.[6]

From a urological perspective, the supine position also has several benefits in such cases. There is no need for repositioning during surgery, which can be particularly beneficial in patients with limited mobility and anatomical deformities. PCNL and cystolytholapaxy can be performed simultaneously, which can decrease the overall operative time.

The technical aspects of percutaneous access in spina bifida patients are also more complex.

Unfortunately, in the supine position, the access space between the rib cage and the iliac crest is narrower, compared to the prone position. In this case, the staghorn stone was on the convex side of the scoliotic spine, which made the renal access slightly easier. If the stone had been on the left side, which is the concave side of the spine, the space for access would have been more limited. In such a case, prone PCNL might be beneficial due to the wider space between the iliac crest and the rib cage, therefore the choice of position in such complex cases should be always individualized.

In our case, ultrasound-guided fluoroscopic adjustment was crucial for accurate puncture. Ultrasound-guided puncture has several advantages, especially in patients with atypical anatomy. Ultrasound can verify the renal and perirenal anatomy, improve the visualisation of the adjacent organs and distinguish between anterior and posterior calyx, reduce radiation exposure and provide real-time imaging of the collecting system. Additionally, applying Doppler flow imaging can help avoid vascular injury. Due to these advantages, ultrasound increases the safety of the puncture.[7] The fluoroscopic image quality was another difficulty, as it was compromised by the presence of spinal hardware and the ventriculoperitoneal shunt.

Based on our literature review, this technique appears to be novel. We did not find any articles describing PCNL and cystolitholapaxy being performed simultaneously by two surgeons. There are case reports about combining prone PCNL and cystolitholapaxy for the removal of encrusted double J stents, but these were carried out sequentially by one surgeon.[8,9] Given that this is a single case involving a non-standard patient, we cannot draw definitive conclusions about the safety of the procedure. However, we believe that if both parts of the surgery are performed by experienced surgeons, the operating time can be significantly reduced, potentially lowering the complication rate.

In the end the patient became stone-free in one session. We plan a close follow-up to prevent the formation of such a big stone-burden in the future.

3. Conclusion

Our case clearly demonstrates the versatility of the supine position. Performing simultaneous PCNL and cystolitholapaxy in a patient with spina bifida is feasible, but requires meticulous planning. The unique challenges by the anatomical and physiological characteristics of spina bifida patients necessitate careful anesthesiological management, and a well-chosen patient positioning.

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CRediT authorship contribution statement

Zoltán Kiss: Writing – original draft. **Gyula Drabik:** Writing – review & editing. **János Dócs:** Writing – review & editing. **Tibor Flaskó:** Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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