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CASE REPORT

Extraperitoneal laparoscopy-assisted percutaneous nephrolithotomy in a patient with osteogenesis imperfecta

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Abstract Osteogenesis imperfecta (OI) patients represent a challenge to all physicians, as they do for anesthetists and urologists, when they develop symptomatic stones in the urinary tract. We recently treated an OI patient with renal pelvic stone by extraperitoneal laparoscopy-assisted percutaneous nephrolithotomy (PCNL). To our knowledge, this combined treatment modality has not been reported previously in OI. An 18-year-old paraplegic girl with OI presented to our urology department because of right-sided flank pain. She pointed out that she had right kidney stone for the previous 2 years, and because of risks of general anesthesia and surgical procedures, surveillance was recommended. Intravenous pyelography was performed and an 11.9-mm stone at the pelvis of the right kidney and grade 1–2 hydronephrosis at the same side with normal kidney functions and severe left-sided scoliosis were detected. After explanation of risks of the treatment modality and general anesthesia to the patient, extraperitoneal laparoscopy-assisted PCNL was performed. No complications occurred due to general anesthesia or surgical procedure. The operation time was 95 min and no blood transfusion was required. The nephrostomy tube and retroperitoneal drain were removed 2 and 3 days after the procedure, respectively. The patient was doing well at a follow-up of 6 months. Extraperitoneal laparoscopy-assisted PCNL approach may decrease the risk of surgery as an alternative treatment modality for OI patients. Such cases should be operated on at centers with significant experience in the field of endourology, where all the equipment and specialized personnel are readily available.

Keywords Kidney stone · Laparoscopy · Osteogenesis imperfecta · Percutaneous nephrolithotripsy

Abbreviations

OI	Osteogenesis imperfecta
PCNL	Percutaneous nephrolithotomy

Introduction

Osteogenesis imperfecta (OI) is a rare congenital disease of connective tissue [1]. Increasing evidence points to a disturbance of type I collagen biosynthesis as the underlying defect [2, 3]. The main skeletal manifestations are bone fragility, osteopenia, skeletal deformities, short stature and dentinogenesis imperfecta. OI has some ocular complications such as blue sclerae, myopia, embryotoxon and keratoconus [1, 4, 5]. Joint laxity or contractures, hyperplastic scar formation and increased capillary fragility can be additional problems [6, 7, 8].

When OI patients present with symptomatic stones in their urinary tract, they represent a challenge to all physicians, as they do for anesthetists and urologists. General anesthesia can be risky because of high basal metabolism that may cause malignant or non-malignant hyperthermia; bleeding and fractures of the mandible and teeth also have been described [9]. Urologists must consider the altered

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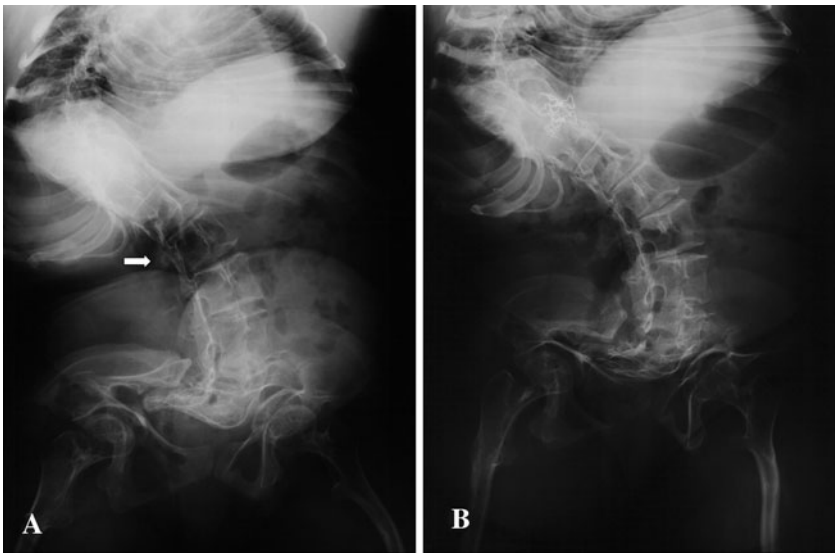

Fig. 1 Plain X-ray of kidney, ureter and bladder before (**a**) and after (**b**) the treatment. Patient had severe left-sided scoliosis. *White arrow* show the renal stone

anatomy of the urinary tract and the fragility of the bones before deciding about the treatment modality. Because of the potential complications mentioned above, reports about the surgical treatment of urinary stones in OI patients are rare in the literature [9].

We recently treated an OI patient with renal pelvic stone by extraperitoneal laparoscopy-assisted percutaneous nephrolithotomy (PCNL). The technique and the patient characteristics are reported. To our knowledge, this combined treatment modality has not been reported previously in OI.

Case report

An 18-year-old paraplegic girl with OI presented to our urology department because of right-sided flank pain. She pointed out that she had right kidney stone for last the 2 years and, because of the risks of general anesthesia and surgical procedures, surveillance was recommended. Blood nitrogen and creatinine, and urine analyses and cultures were obtained. Since these laboratory findings were normal, intravenous pyelography was performed and an 11.9-mm stone at the pelvis of the right kidney and grade 1–2 hydronephrosis at the same side with normal kidney functions were detected (Fig. 1a). Also, patient had severe left-sided scoliosis. After explanation of risks of the treatment modality and general anesthesia to the patient, extraperitoneal laparoscopy-assisted PCNL was planned.

During induction, fentanyl, sodium pentothal and vecuronium were given and orotracheal intubation was applied. Anesthesia was maintained with 1–2% sevoflurane in a mixture of 50% NO_2 and 50% O_2. After transurethral 14-F foley catheter placement, the patient was secured to the operating table carefully in the supine position. All bony prominences were meticulously padded and extremities carefully placed in the neutral position to minimize postoperative neuromuscular sequelae. The operation table deviated slightly to the left side to gain appropriate access to the right kidney.

We used the open (Hasson cannula) technique for obtaining initial access. A horizontal 1.5-cm skin incision was made just below the tip of the 12th rib. The flank muscle fibers were bluntly separated. Entry was gained into the retroperitoneal space by gently piercing the anterior thoracolumbar fascia with the fingertip. Limited finger dissection of the retroperitoneum was performed in a cephalad direction, immediately anterior to the psoas muscle and fascia, and posterior to the Gerota's fascia to create a space for placement of the balloon dilator. At this juncture, the finger can often palpate the tip of the lower pole of the right kidney. We inserted a trocar-mounted balloon dissection device for rapidly and atraumatically creating a working space in the retroperitoneum.

Following balloon dilatation and removal, a 10-mm trocar was placed as the primary port and secured to eliminate air leakage at the primary port site. The pneumoretroperitoneum was established at 15 mmHg, and a 10-mm, 30° laparoscope was inserted. The psoas muscle and Gerota's fascia were identified; 5 and 10-mm trocars were placed into the retroperitoneum under direct vision. By using laparoscopic dissectors through 5 and 10-mm trocars, Gerota's facia opened and the surface of the kidney dissected to form a safe area to reach the right kidney stone percutaneously without bowel injury. Under direct vision supplied by the laparoscopic approach, an 18-gauge percutaneous needle was inserted into the kidney through a 10-mm trocar and the tract was serially dilated with Teflon coaxial fascial dilators to accept a 30-F Amplatz sheath (Boston Scientific, Natick, MA). A 26-F nephroscope was introduced to reach the stone, which was fragmented with a pneumatic/

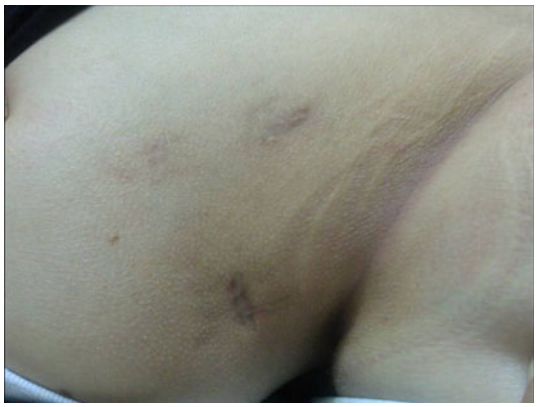

Fig. 2 Patient view, the appearance of trocar incision scars at the right side of the patient 6 months after the procedure

ultrasound lithotripsy device (Swiss Lithoclast Ultra, Boston Scientific). Stone fragments were removed with forceps extraction. At the end of the procedure, X-ray of the kidneys, ureters and bladder and nephrostogram were obtained and no evidence of residual fragments was detected (Fig. 1a). Then a 24-F re-entry nephrostomy tube was placed. A 14-F drain was placed in the retroperitoneal cavity close to the kidney through a 5-mm laparoscopic port. Hemostasis was confirmed and the laparoscopic ports were removed. No complications occurred due to general anesthesia or surgical procedure. The operation time was 95 min and no blood transfusion was required. The nephrostomy tube and retroperitoneal drain were removed 2 and 3 days after the procedure, respectively. Stone analysis revealed calcium phosphate. The patient was doing well at a follow-up of 6 months (Fig. 2).

Discussion

Osteogenesis imperfecta (OI), a heritable metabolic bone disease, is characterized clinically by blue sclera, onset of deafness during early adult life, abnormal dentition and recurrent fractures from osteopenia that often result in short stature and skeletal deformity [10, 11, 12]. Nevertheless, the clinical expression of OI is extremely variable and not all of the above features may be present in a given patient.

Due to the considerable clinical and genetic variability of OI, numerous classifications have been established. At present, the Sillence classification is mainly used in which four patient subgroups are identified. Types I and IV show a relatively mild clinical course and an autosomal dominant mode of inheritance [13, 14]. Type I patients have distinctly blue sclerae, whereas type IV patients have white sclerae. Type III is an autosomal recessive form with a severe clinical course. Type II is lethal. Although this classification has found wide acceptance, there is a considerable number of patients who cannot be assigned to any of the four subtypes.

The often severe scoliosis in OI patients may lead to an obstruction of the urinary flow favoring kidney stone formation. While the risk of stone disease seems to be increased, occurring in about 20% of these patients, in some studies its incidence was reported to be 4.7–6.9% in children with OI (6/127 and 4/58, respectively), which does not appear to differ from that seen in the general population [13, 15].

In cases of kidney stones, flexible ureteroscopy and PCNL are both acceptable options: in pediatric patients, PCNL has been successfully performed in two children with OI [16]. In a recent study, Argyropoulos et al. successfully treated ureteral stone in a patient with OI with an endoscopic approach [9].

Since the first report by Eshghi et al. [17] several authors have described techniques for laparoscopy-assisted percutaneous transperitoneal nephrolithotomy (PCNL) in ectopic pelvic kidneys [18]. Percutaneous access was obtained by retrograde nephrostomy in combination with continuous observation and displacement of bowel loops via a laparoscope. Holman et al. [19] reported a series of 15 patients treated with laparoscopy-assisted percutaneous transperitoneal nephrolithotomy.

Troxel et al. [20] described extraperitoneal laparoscopy-assisted percutaneous approach to access the lower pole calyx of a pelvic kidney for PCNL. Desai et al. [21] performed PCNL in nine patients with ectopic kidneys; however, the procedure was performed under ultrasound guidance alone. The aim of laparoscopy-assisted percutaneous approach was to access to the kidney under direct vision by preventing bowel and organ injuries.

The case we report here is to our knowledge the first in literature. We must emphasize that careful preoperative planning is required. The urologist should select an approach most likely to facilitate treatment in one stage in the minimum amount of time. While treating kidney stones, urologists must be careful about preventing other problems such as other organ injuries (e.g., bowel), spontaneous fractures, or malign or non-malign hyperthermia due to anesthesia.

The type of anesthesia used is an important factor influencing the choice of treatment modality, since general anesthesia carries several risks. Malignant or non-malignant hyperthermia, fractures of the mandible, teeth and cervical spine, and mucosal bleeding or bruising have been reported as a result of anesthesia in OL patients [22]. Patients must be informed of these risks associated with general anesthesia.

OI patients represent a challenge to all physicians, as also to anesthetists and urologists when they develop symptomatic stones in the urinary tract. Extraperitoneal laparos-

copy-assisted PCNL approach may decrease the risk of surgery as an alternative treatment modality for OI patients. Such cases should be operated on at centers with significant experience in the field of endourology, where all the equipment and specialized personnel are readily available.

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