

Extracorporeal Shock Wave Lithotripsy (ESWL) for Kidney Stone Treatment in People with Hemophilia A

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Introduction

Acquired hemophilia A is a rare bleeding disorder that presents as gross hematuria, with or without an initiating factor such as stone or cancer in urinary tract. People with hemophilia may require some surgical procedures in their live. Surgery in patients with congenital or acquired coagulation defects has always been challenging and requires special care with a multidisciplinary approach. Extracorporeal Shock Wave Lithotripsy (ESWL) is a standard procedure performed in patients with kidney stones.

Case Description

We report a 48 years old man who was admitted to the emergency room with chief complain left flank pain. Pain was felt two months before entering to the hospital. Dull pain, comes and goes and radiates to the front of the stomach. Two days before entering the hospital the patient complained of hematuria. There was no history of passing stone and previous surgery. Patient is a Hemophilia A patient with routine treatment.

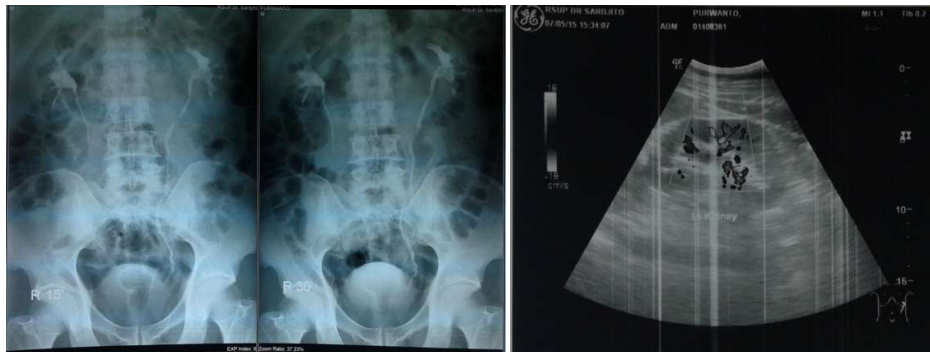


Figure 1. BNO-IVP and USG results showing left kidney stones.

On physical examination, the general condition of the patient was good, no abnormalities were found on the physical examination and urological status. We performed ultrasound and intravenous pyelogram (IVP) examinations on patients. On examination, a lucent stone was found in the patient's left kidney with a size of 10x11 mm (Figure 1). Laboratory examination showed an increase in APTT values (45.7/32.5) and a decrease in blood factor VIII levels (5.4%).

We performed Extracorporeal Shock Wave Lithotripsy (ESWL) on the left kidney. The lithotripsy was performed with ESWL with a focus 1-2, power set 19-20 joules, the frequency of 60 shocks per minute, for total 6000 shocks. Before the ESWL procedure is carried out, factor VIII preparation (Koate®) 1500 units is given 12 hours before the procedure. Administration of factor VIII to optimize coagulation status in patients. During the ESWL procedure, no signs of acute bleeding were observed in patient. However, patient complained of pain in the left flank. Left kidney stones were visible on USG and X-ray before lithotripsy (Figure 2).

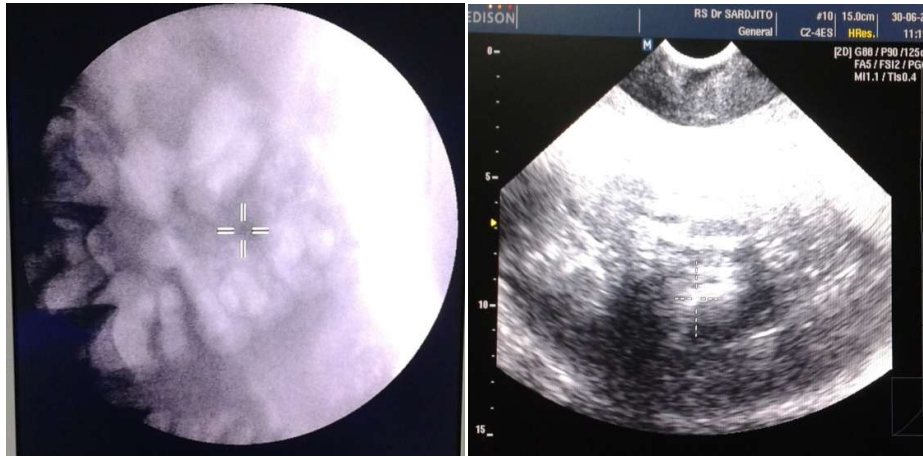


Figure 2. X-ray and ultrasound before ESWL show stones in the left kidney

The patient underwent hospitalization after the ESWL procedure. The results of laboratory examinations after the procedure showed that Hb levels had decreased from 16.2 gr/dl post to 15.7 gr/dl. The patient's hemostasis status showed APTT lengthening from 45.7 control 32.5 before surgery to 50.7 control 32.5 after surgery. The patient complained of pain in the left hip after the procedure. The patient also complained of hematuria after the ESWL procedure. A urinalysis examination revealed three positive erythrocyturia. The patient discharged three days after the procedure.



Figure 3. X-ray and ultrasound images after ESWL

The patient is planned to undergo a kidney ultrasound examination to evaluate the stones and the condition of the kidneys. Ultrasound examination revealed a perirenal hematoma of the left kidney with a diameter of 12.5 cm. The patient came for control to the polyclinic with complaints of fever three days before the control. There were no complaints of hematuria while the patient was at home. The patient also complained of hematoma in the left flank (Figure 5). The reddish lump feels painful. We did a blood test and found that the Hb level had decreased to 6.3 gr/dl. The patient's APTT level was 45.8 with the control 31. We re-admitted the patient for a blood transfusion. During the five days of treatment, the patient received three blood transfusions Packet Red Cell (PRC). The patient was discharged after five days of treatment.



Figure 4. Image of hematoma on the left flank seven days after ESWL.

Discussion

Managing urinary tract stone in patient with hemophilia is a real challenge to the urologist. Haematuria is a recognized sign of bleeding in urinary tract. Adult persons with hemophilia (PWH) have a higher prevalence of renal disease than the general population (Kyle et al., 2019).

Hemophilia is a rare bleeding disorder that may present as gross hematuria, with or without an initiating factor such as urinary tract stone. This condition should be considered when isolated prolonged Partial Thromboplastin time (PTT) is observed. Early recognition and prompt treatment are crucial, as they can be lifesaving. Although rare, acquired hemophilia A seldom presents as isolated gross hematuria. It is a serious condition with a high mortality rate, making clinical suspicion and prompt diagnosis essential for effective management (Max Schmidt-Bowman et al., 2018).

The patient in this report was a 48 years old man diagnosed with left kidney stones with hemophilia A. In this patient, left ESWL was performed. Operative management of hemophilia patients requires special preparation. The goal of prophylaxis is to reduce the occurrence of bleeding and is generally chosen in severe hemophilia to intermediate/mild hemophilia given regularly (typically 2–3 times per week) (Wenfeng Xu et al., 2023). In this case, the patient was administered factor VIII (Koate®) 1500 units 24 hours and 12 hours before the ESWL procedure. Giving factor VIII preparations to increase factor VIII levels thereby reducing the risk of bleeding. With effective substitution of deficient clotting factors, ESWL is a safe and low morbidity method in the treatment of urinary calculi in hemophiliacs (Saata et al., 2015).

With adequate preoperative preparation, close observation during surgery, and post-operative monitoring will improve outcome which is good for hemophilia patients undergoing surgery. Turoctocog alfa pegol appears to be efficacious for perioperative hemostatic treatment of all kind of surgical operations (minor and major) in PWH A ([E. Carlos Rodríguez-Merchán](#), 2021). During invasive surgical procedures, patients with hemophilia require additional treatment to maintain haemostasis; however, due to the complexity of hemophilia there is a lack of consensus on the optimal management (Wolfgang Miesbach, 2020).

After ESWL was performed on this patient, complications of bleeding in the urinary tract were found. Hematuria occurred in patients after the ESWL procedure. Complaints of hematuria decreased one day after the procedure. A complication that often arises after ESWL is hematuria. Hematuria occurs due to irritation of the urethelium due to stone fragments. Severe bleeding complications after ESWL procedures are rare. The most common side effects of ESWL procedures are hematuria associated with microtrauma and microscopic bleeding in the kidneys, significant bleeding occurs in 0.6% of patients without coagulation abnormalities (Sallami et al., 2015).

In this study, a decrease in preoperative and postoperative hemoglobin (Hb) values was found. The pre-operative Hb of 16.2 gr/dl decreased to 15.7 gr/dl after ESWL was carried out. Study systematic review by Sallami et.al 2015 showed that ESWL is safe and effective for use in patients suffering from hemophilia. The study reviewed 12 studies with 25 kidney stone patients. There were 4 (16%) patients experiencing severe bleeding complications (Sallami et al., 2015). Impaired hemostasis is a contraindication for surgery, so correction of preoperative hemostasis status is necessary. Research by Czaplicki, et al in 2000 observed 25 patients with urinary tract stones with hemophilia. In this study, there were no bleeding complications found in patients after ESWL. Research by Bastounis et al in 2000 looked at major operations performed over 10 years on hemophilia patients. It was found that 68 patients underwent major surgery. There were no deaths found in all patients and overall morbidity rate by 6%. Surgical care for patients with hemophilia disorders should only be given by hospitals where cooperation of a multidisciplinary team exists, including the urologist and an experienced hematologist. Hospital must ensure the availability of adequate replacement therapy and continuous monitoring to manage these patients effectively (Ali Ersin Zumrutbas et al, 2016).

Conclusions

Surgical care for patients with hemophilia A should provided only in hospitals where multidisciplinary team available, including urologist and experienced hematologist. Adequate replacement therapy and monitoring must always available. We suggest that ESWL can be performed safely in kidney stone patient with hemophilia A. Whenever possible, less invasive options should be considered as the priority or first option to decrease the risk of bleeding.

Acknowledgement

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