

AN EXPERIENCE OF SUPINE PCNL ON BILATERAL STAGHORN AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

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ABSTRACT

Objective: The aim of this article was to reports our experiences PCNL on ADPKD patients. **Case(s) Presentation:** We report the case of a 38-year-old male with autosomal dominant polycystic kidney disease associated with bilateral staghorn diseases. We performed a supine PCNL on left kidney. **Discussion:** ADPKD may arise sporadically, the developmental abnormality that resulting in multiple cysts, hypertension, hematuria, polyuria, plank pain, and are prone to reccurent urinary tract infection and renal stones. ADPKD is one most commonly genetics disorder than frequently results in End Stage Renal Disease (ESRD). Population of ADPKD has a higher risk to have a nephrolithiasis, thus it may hastened the onset of ESRD. Open surgeries was one of treatment of choices of ADPKD with staghorn stones, but considering of high rates of morbidity. Recently there's consistent switching the trend on minimal invasive treatment such as ESWL and PCNL. **Conclusion:** PCNL may considered as an effective and safe procedure in managing nephrolithiasis in ADPKD, further studies with larger sample and longer periods observation is needed to confirmed role of PCNL in preserving kidney on ADPKD with staghorn kidney.

Keywords: Supine PCNL, ADPKD, kidney disease.

ABSTRAK

Tujuan: Melaporkan pengalaman PCNL pada pasien ADPKD. **Presentasi Kasus:** Kami melaporkan kasus seorang pria berusia 38 tahun dengan penyakit ginjal polikistik dominan autosom dengan pemberat berupa batu staghorn bilateral. Kami melakukan PCNL dengan posisi supine. **Diskusi:** ADPKD dapat muncul secara sporadis sebagai kelainan perkembangan yang mengakibatkan beberapa kista, hipertensi, hematuria, poliuria, nyeri punggung dan rentan terhadap infeksi saluran kemih dan batu ginjal. ADPKD salah satu kelainan genetika yang paling umum dan sering terjadi pada End Stage Renal Disease (ESRD). Populasi ADPKD memiliki risiko lebih tinggi pada nefrolitiasis, sehingga dapat mempercepat timbulnya ESRD. Operasi terbuka adalah salah satu pengobatan pilihan ADPKD dengan batu staghorn, tetapi mempertimbangkan tingkat morbiditas yang tinggi. Baru-baru ini terdapat perawatan invasif minimal seperti ESWL dan PCNL. **Simpulan:** PCNL sebagai prosedur yang efektif dan aman dalam manajemen nefrolitiasis ADPKD, studi lebih lanjut dengan sampel yang lebih besar dan pengamatan periode yang lebih lama diperlukan untuk mengkonfirmasi peran PCNL dalam menjaga ginjal pada ADPKD dengan ginjal staghorn.

Kata Kunci: PCNL supine, ADPKD, penyakit ginjal.

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INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is associated with an increased incidence of nephrolithiasis, the probability of these patients is 5-10 higher than general population (RR: 5-10) with higher risk of obstruction, infection, and altered function due it's anatomical abnormality, with concurrence of Staghorn stone facilitated the onset of Chronic Kidney Failure (CKD).¹⁻⁴

Due the distorted and the difficult anatomy and associated chronic kidney disease PCNL feasibility and safety feared among most urologist. However, the condition with Staghorn stone it's self may hastened the onset of end stage of renal Disease (ESRD), due increase risk of RCC, nephrectomy and kidney transplant is recommended among these patients.⁵ These options are not available for most cases, further making the management more challenging. CKD-associated coagulation defect

increases risk of bleeding during or after the PCNL procedure. On this article we share experience of using this procedure in patient in ADPKD with staghorn kidney, the pitfalls of this procedure also discussed.

CASE(S) PRESENTATION

A 38 years old male presented with chronic flank pain, pain describe as dull pain with discomfort that are not relieved by analgesic, history of passing stone was admitted. History of fever, gross hematuria, and nausea was denied, his father has history of ESRD, but the diagnosis of causes was unconfirmed. Previously, he used to treat the pain with commercial pain killer, no significant medication previously was noted. On physical examination, hypertension was noted (Blood Pressure: 160/100), flank mass was palpated on both of his plank, tenderness wasn't found. The Creatinine was mark on 2.3 mg/dl on USG examination the kidney enlarged bilaterally with multiple cyst and multiple acoustic shadow.

On admission, routine blood test was mark normal, on urinalysis we found that specific gravity of urine <1.005, PH 6, blood 3+ leukocyte esterase 1+, and protein 1+. Urine culture didn't shown any positif culture. On MS-CT Scan, we found that both bilateral kidney has multiple cysts bilaterally, and stone was found over calyxs (Figure 1). Supine PNCL was performed on left kidney with single

puncture was obtained and retained stone was found 5 mm and ESWL was performed for the remained stones. Patient has single periods of fever 24 hours after procedure and reliefs after. The second PCNL will performed later. The stone analysis found that the structure of stone was made by uric acid and oxalate.

Initial puncture was made on left infra-costovertebral angle, initial puncture was punctured on cyst that shown leaked urine-like from needle puncture, methyle blue wasn't leaked from urethra catheter and guidewire insertion was out of the track (Figure 2). C-arm was rotated 20 degree to gain more to depth of the needle, and reinsertion of access needle. Methylene blue was leaked, and guidewire was on position to confirm initial puncture was in calyx system (Figure 2).

Distorted anatomy related to false puncture to Cysts puncture, methylene blue dye installation through the ureteric catheter was conducted (Figure 3). Once the puncture was confirmed was on calyceal, subsequent dilatation was done with help of Amplatz sequential facial dilator. A 24 Fr nephroscope was used, and a single puncture was used on the left kidney. False puncture also can be confirm by insert the wire and the shown the false track. Patient has 2 days onset of fever (38' C), and didn't experienced a hematuria, and pain after the procedure. A precise puncture needed to avoided further loss of nephrons and altering kidney function. And patient was discharge on seventh days.

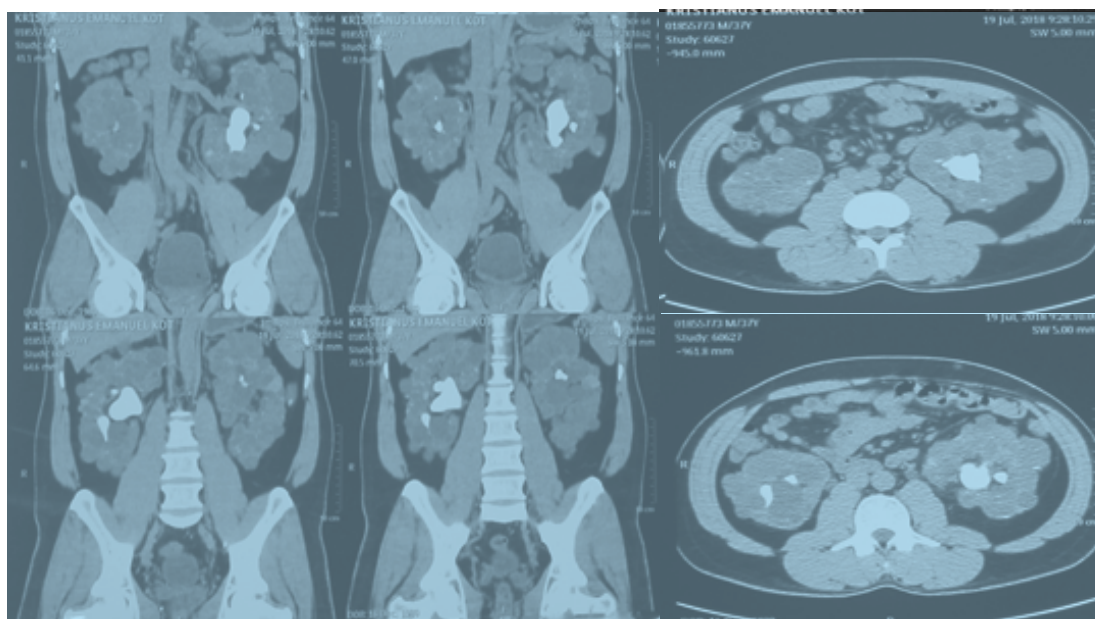


Figure 1. Pre-operative CT of an ADPKD patient showing multiple cyst and staghorn stone bilaterally.

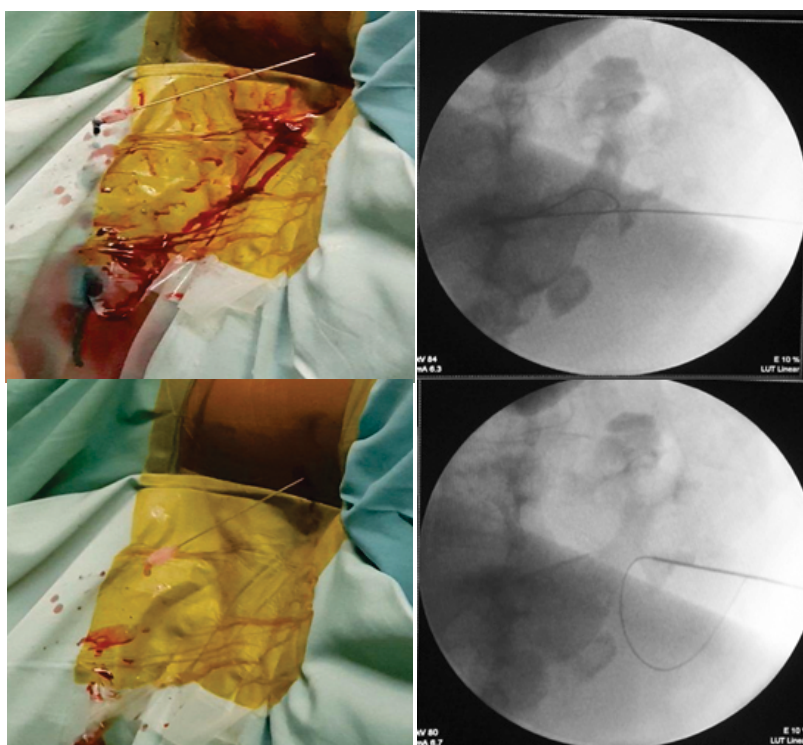


Figure 2. Urine-like extracted from puncture needle (left top) and false track shown by guide-wire to is common pitfalls (right top). Methylene blue from puncture needle (left below) and guide-wire on calyx to confirm the puncture (right below).



Figure 3. Retained Stone do not appear on plain Abdomen after ESWL.

DISCUSSION

ADPKD may arise sporadically, the developmental abnormality that resulting on multiple cysts, hypertension, hematuria, polyuria,

and plank pain and are prone to reccurent urinary tract infection and renal stones. ADPKD is one most commonly genetics disorder than frequently results in End Stage Renal Disease (ESRD). Population of ADPKD has a higher risk to have a nephrolithiasis. Thus it may hastened the onset of ESRD.⁶⁻⁹ Open surgeries was one of treatment of choices of ADPKD with staghorn stones, but considering of high rates of morbidity. Recently there's consistent switching the trend on minimal invasive treatment such as ESWL and PCNL.¹⁰⁻¹²

PCNL on ADPKD becomes more challenging that general population, due distorted pelvicaliceal sistem anatomy, and calcification of parenchymal misleading to presence of the stone under fluoroscopy, the anatomy complexity required a multiple puncture, a precise anatomical reconstruction and calyxs preference is important step on PCNL.¹³⁻¹⁴ On this case, we performed a methylene blue installation through the ureteric catheter and aspiration of it's from jet efflux to confirm the correct puncture, and false tract also confirmed by guide-wire, a pitfalls for false puncture can be anticipated. Small retained stoned was treated with ESWL a week later. A trauma resulting from the

puncture site was inevitable, in this cases creatine was slightly rose 2 day after procedure due loss of nephrons. However, the renal function stable and obstructed system was markly improved postoperatively.

On this case, patient was diagnoses based on ADPKD diagnostic criteria and algorithm diagnosis published by a grant-in-Aid for Progressive Renal Diseases Researches, Ministry of Health, labour and welfare of japan. On this case, patient has history of non-confirmation of family history, multiple cyst that assessed by USG and CT scan, the cysts observed multiple bilaterally with staghorn kidney. Currently, advanced management of ADPKD do not exist, ideally, the treatment should start early; the goal of management ADPKD are attenuate the progression of ESRD, control of complication, and changing the life style.^{5,9,15}

CONCLUSION

In conclusion, despite anatomically distorted and renal function is impaired, PCNL may considered as an effective and safe procedure in managing nephrolithiasis in ADPKD, further studies with larger sample and longer periods observation is needed to confirmed role of PCNL in preserving kidney on ADPKD with staghorn kidney. On the same times, to perform this procedure urologist should avoid pitfalls on false dilatation to the cyst cavity and make further nephron lost.

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