CASE REPORT

Haematuria in ADPKD: not always benign. Be aware!

Manav Agarwal, Ashok Kumar Sokhal, Manoj Kumar, Sunny Goel

Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India

Correspondence to

Dr Manav Agarwal,
agrawalmanav@gmail.com

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SUMMARY

Haematuria in patients with autosomal dominant polycystic kidney disease (ADPKD) is a very common manifestation. The cause of haematuria is often benign with the most common cause being haemorrhage within the renal cyst. But haematuria may also be caused by a coincident malignancy, the diagnosis of which may be missed if not investigated thoroughly. Herein, we present a case of ADPKD who presented to us with haematuria and was later found to have bladder cancer.

BACKGROUND

Autosomal dominant polycystic kidney disease (ADPKD) is a common inherited condition, occurring in 1 out of 400 to 1000 live births.¹ The inherent pathophysiology involves the formation of multiple cysts leading to progressive destruction of functional renal parenchyma and eventually end stage renal disease (ESRD).² About 50% of patients suffer from episodic haematuria during their lifetime, which is mostly having benign aetiology.³ However, sometimes haematuria may have an unrelated cause which needs proper evaluation and management. We present a case of urinary bladder carcinoma in a patient with ADPKD.

CASE PRESENTATION

A 38-year-old man, farmer by occupation, had an episode of gross haematuria 1 year back. It resolved spontaneously within a week. He was found to have multiple cystic lesions in both kidneys on ultrasonography and was subsequently diagnosed to have ADPKD at a community health centre. Patient did not attend clinic appointments for regular follow-up. He now came to us with complaints of nausea, vomiting, decreased appetite, generalised weakness and breathlessness for 15 days and painless gross haematuria for 3 days. He was having oliguria (<0.5 mL/kg/hour) for the past 1 week. He had no significant family history. Patient had no history of smoking or any chemotherapeutic exposure. On general physical examination, patient was afebrile, pulse rate 96/min, respiratory rate 36/min and blood pressure 168/94 mm Hg. On local examination, a non-tender, firm, bosselated lump was palpable over the left flank region, which moved with respiration. Coarse crepts were audible in the bilateral basal lung fields. The rest of the systemic examination was within normal limits.

INVESTIGATIONS

Blood investigations revealed deranged serum urea 358 mg/dL, creatinine 13.80 mg/dL and

haemoglobin 9.4 g/dL. Liver function tests were normal. Ultrasound of the abdomen revealed bilateral enlarged polycystic kidneys (figure 1) with liver cysts and urinary bladder filled with clots. On cystoscopy, he was found to have large clots and a small pedunculated bladder mass of 1.5×0.5 cm on the right lateral wall. Non-contrast CT scan of the abdomen and pelvis revealed multiple cysts in both kidneys and liver and a urinary bladder mass on the right lateral wall with no evidence of extravesical invasion (figure 2).

DIFFERENTIAL DIAGNOSIS

Haemorrhage within the renal cyst, coincident infection, nephrolithiasis and bladder tumour.

TREATMENT

In the setting of renal failure with uraemia, nephrology consultation was done after which the patient underwent haemodialysis. After three sessions of haemodialysis, his breathlessness and general well-being improved. We did a transurethral resection of bladder mass under general anaesthesia, and perioperatively, mitomycin C was instilled intravesically.

OUTCOME AND FOLLOW-UP

The patient is on maintenance haemodialysis and is under regular follow-up in nephrology and urology departments. Histopathology of the transurethral resection of bladder tumor (TURBT) specimen revealed low-grade, non-muscle invasive transitional cell carcinoma. He had no recurrences in the last 6 months.

DISCUSSION

Haematuria has myriad causes, some of which may be treatable, whereas others are life-threatening.⁴ Male gender and increasing age are important risk factors for malignancy. Recently, a large prospective study of 4020 patients with haematuria reported 12.1% overall prevalence of malignant disease. In men, a malignant cause was found in 4.7% (23/488) and 15.65% (335/2140) below and above 50 years, respectively.⁵

Patients with ADPKD are educated and advised to be on regular follow-up with their nephrologists, who monitor and prevent further deterioration of renal function and its complications. Haematuria is often encountered in such patients, and its cause is usually haemorrhage into a renal cyst communicating with the pelvicalyceal system.⁶ Other causes are infection and coincident nephrolithiasis (occurs in around 20% of patients). Haematuria typically

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Figure 1 Ultrasound images showing multiple renal cyst. (A) Right kidney. (B) Left kidney.

resolves within a week with conservative measures such as bed rest, hydration and discontinuation of offending drugs, if any (eg antiplatelets or anticoagulants).⁷ However, as in our case, an underlying malignancy may be the causative factor, which needs aggressive management but may have been easily overlooked.

After extensive literature search, we were able to find an article reporting two such cases of bladder cancer in patients with ADPKD.⁸ Both cases were detected by transabdominal ultrasonography. The first case was a 49-year-old man with a 3 cm polypoidal mass at the base of the bladder, whereas the second case was of a 34-year-old pregnant woman (size and location of tumour not mentioned). The histopathological examination of the resected tumours revealed them to be pT1 G2 and pTa G2, respectively.

There is no documentary evidence of increased frequency of neoplasms in patients with ADPKD.⁹ Moreover, tests to exclude neoplasms in patients with ADPKD with haematuria are generally advised in cases of persistent haematuria beyond 1 week or when the patient's first haemorrhagic episode is above 50 years.¹ All three patients of bladder cancer in ADPKD were less than 50 years, and it also

well-known that bladder cancer may itself present with intermittent gross haematuria/microscopic haematuria.¹⁰

In light of these statements, it becomes very difficult to chalk out an infallible algorithm to avoid missing such coexisting underlying neoplasms in a patient population with ADPKD.

Learning points

- ▶ Haematuria in autosomal dominant polycystic kidney disease (ADPKD), though mostly benign, should be evaluated with high index of suspicion to rule out malignancy.
- ▶ Early diagnosis and treatment can be very beneficial for the patient.
- ▶ Further genetic and molecular studies may be needed to reveal which subset of patients with ADPKD may be having a greater risk of malignancy.

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REFERENCES

- 1 Chebibe FT, Torres VE. Autosomal dominant polycystic kidney disease: core curriculum 2016. *Am J Kidney Dis* 2016;67:792–810.
- 2 Chapman AB, Devuyst O, Eckardt KU, et al. Autosomal-dominant polycystic kidney disease (ADPKD): executive summary from a Kidney Disease: Improving Global Outcomes (KDIGO) controversies conference. *Kidney Int* 2015;88:17–27.
- 3 Gabow PA. Autosomal dominant polycystic kidney disease. *N Engl J Med Overseas Ed* 1993;329:332–42.
- 4 Feldman A, Hsu C, Kurtz M, et al. Etiology and evaluation of hematuria in adults. *UpToDate*, 2013. version 170.
- 5 Edwards TJ, Dickinson AJ, Gosling J, et al. Patient-specific risk of undetected malignant disease after investigation for haematuria, based on a 4-year follow-up. *BJU Int* 2011;107:247–52.
- 6 Suwabe T, Ubara Y, Sumida K, et al. Clinical features of cyst infection and hemorrhage in ADPKD: new diagnostic criteria. *Clin Exp Nephrol* 2012;16:892–902.
- 7 Peces R, Aguilar A, Vega C, et al. Medical therapy with tranexamic acid in autosomal dominant polycystic kidney disease patients with severe haematuria. *Nefrologia* 2012;32:160–5.
- 8 Dedi R, Bhandari S, Turney JH, et al. Causes of haematuria in adult polycystic kidney disease. *BMJ* 2001;323:7309–86.
- 9 Jilg CA, Drendel V, Bacher J, et al. Autosomal dominant polycystic kidney disease: prevalence of renal neoplasias in surgical kidney specimens. *Nephron Clin Pract* 2013;123:13–21.
- 10 Schmidt-Hansen M, Berendse S, Hamilton W. The association between symptoms and bladder or renal tract cancer in primary care: a systematic review. *Br J Gen Pract* 2015;65:e769–775.

Figure 2 Non-contrast CT scan image showing bladder mass.

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