Case Report

Chronic loin pain in a 65-year old adult polycystic kidney disease patient

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ABSTRACT

Background: Autosomal Dominant Polycystic Kidney Disease (ADPKD) is often characterized clinically by acute/chronic pain, hypertension and variable progression to end-stage renal disease. About 60% of all ADPKD patients are estimated to have chronic pain, and often this is the initially presenting symptom of an ADPKD patient that brings them to medical attention. Aim: This study reports a case of a 65-year old with polycystic kidney disease and chronic loin pain. Findings: Our patient presented with history of chronic loin pain which we believed was due to increased cysts sizes/increase in weight of the kidneys. A good history was taken from him on presentation, and also laboratory investigations were carried out. However, the cause of the pain was believed to be due to increase in cysts sizes/weight of the kidneys. The patient was educated on the possible cause of the pain and various treatment options for the condition. He was placed on intermittent use of paracetamol with good response. Conclusion: It is important to educate patients suffering from chronic loin pain that the pain may not be relieved permanently by any intervention method adopted, and also to offer them the treatment modalities available to enable them to cope with the pain.

Key words: Polycystic kidney disease, chronic loin pain, hypertension, paracetamol

INTRODUCTION

Polycystic kidney disease is the most common life-threatening genetic disease, affecting approximately 7 million people worldwide.1,2 Autosomal dominant and autosomal recessive polycystic kidney diseases (ARPKD) are the two forms of the disease. The former is usually of late-onset and characterized by progressive development of cyst and bilaterally enlarged kidneys with multiple cysts, while the later is much rarer than ADPKD and is often fatal in...
utero or during the first month of life. The signs and symptoms of the condition are usually apparent at birth or in early infancy. ADPKD affects up to 1 in 1000 people, while the ARPKD is estimated to occur in approximately 1 in 20,000 people. The genetic defect is located on chromosome 16 and 6 respectively for the autosomal dominant and the autosomal recessive polycystic kidney disease.

Autosomal dominant polycystic kidney disease (ADPKD) is the commonest renal disorder involving a single gene and the fourth leading cause of end-stage renal disease in adults. There are two types: type 1, which is caused by mutations in the ADPKD1 gene and accounts for 85 to 90 percent of cases, and type 2, which is caused by mutations in the ADPKD2 gene and accounts for 10 to 15 percent of cases. The protein products of these two genes, polycystin-1 and polycystin-2, occur on renal tubular epithelia. Polycystin-1 is a membrane receptor capable of binding and interacting with many proteins, carbohydrates, and lipids and eliciting intracellular responses through phosphorylation pathways, whereas polycystin-2 is thought to act as a calcium-permeable channel.

The two types of autosomal dominant polycystic kidney disease have similar pathological and physiological features, but type 2 disease has a later onset of symptoms and a slower rate of progression to renal failure; thus, patients have a longer life expectancy (69.1 years) than those with type I disease (53.0 years). Some patients with typical features of autosomal dominant polycystic kidney disease have no mutations in ADPKD1 or ADPKD2 gene, suggesting that there may be a rare third form of the disease, although the proposed gene, ADPKD3, has not been identified.

Patients with mutations in both the ADPKD1 and ADPKD2 genes (transheterozygotes) have a more severe clinical course than those with mutations in only one of the genes.

The common renal features of ADPKD include hypertension, haematuria (macroscopic and microscopic), loin pain, urinary tract infection, and nephrolithiasis. Pain, (including abdominal pain and low back pain) is a common problem in patients with this kidney disease.

CASE REPORT

The patient is a 65-year old retired civil servant, with history of chronic left loin pain. He was apparently well until about 4 months prior to his presentation in the nephrology clinic when he started experiencing left loin pain. The pain is usually dull and persistent; it does not radiate to any other part of the body, and does not prevent him from engaging in his daily activities. It is temporarily relieved by taking analgesic. There is no known aggravating or relieving factor. He was diagnosed as having ADPKD while being managed for the pain in a private hospital and referred to our renal clinic for expert management. He is a known hypertensive for the past three years prior to presentation. His 62-year-old junior sister is also hypertensive. The mother had hypertension before her death, however the cause of her death is not known. The patient children (4 in number), the youngest being 23 years, refused screening, however none of them was hypertensive, or had any feature suggestive of renal disease.

His weight was 60.5 kg. The kidneys were bilaterally palpable, not tender. The blood pressure was 200/110mmHg. The apex beat was displaced and heaving. He also had grade 2 hypertensive retinopathy.

Abdominal ultrasound also showed that both kidneys were enlarged, measuring 21.5cm x 9.9cm (left) and 20.7cm x 7.92cm (right) with multiple cysts (more than 6 cysts in each kidney), distorting the normal corticomedullary distinction. Intravenous Urography (IVU) did not show any stone. Urinalysis was normal. The electrolyte, urea, and creatinine values were within normal range. The calculated glomerular filtration rate using the Cockroft-Gault equation formula was 63ml/min. the 24-hour creatinine clearance was 56ml/min. The 24-hour urine protein was 0.32g. Urine culture yielded no growth. The electrocardiography showed left axis deviation, left atria enlargement, and left ventricular hypertrophy. The echocardiograph showed concentric hypertrophy of the left ventricle, there was no
valvular abnormality detected. The results of a complete blood count, uric acid, phosphate, serum creatinine level and measurements of glucose, were normal. The diagnosis was ADPKD and hypertension.

He was treated empirically for urinary tract infection using tablet ciprofloxacin 500mg twice daily for 14 days, however there was no improvement. The hypertension was controlled with tablet atenolol 50mg daily, tablet valsartan 80mg daily and amlodipine 10mg daily. He was given paracetamol tablets for the pain which only relieves the pain when he is continuously on it for some days.

**DISCUSSION**

Pain is a common complaint in patients with ADPKD, affecting about 60% of patients with an established diagnosis. In differentiating the cause of the pain and defining an approach to its management, a systematic approach is necessary. A good history taking, and investigation coupled with a good understanding of the innervation of the kidney is also vital in understanding the source of the pain. Pain, located in the abdomen, the flank, or the back, is the most common initial complaint, and it is almost universally present in patients with ADPKD. Flank or abdominal pain usually accompanies renal infection, cyst hemorrhage, or renal stones; however, severe pain may also occur in the absence of other apparent renal complications.

It is a need to differentiate between cystic pain, and pain due to other renal pathology. Pain associated with hemorrhage into the cyst is usually acute and self-limiting, while pain due to infection is usually diagnosed by a good history, urinalysis, and urine culture. However, it is a known fact that in ADPKD patients with urinary tract infection (such as cystic infection), urinalysis may be relatively benign and the urine culture may be persistently negative. This is the reason the patient was treated with antibiotics when he was first seen despite a negative urine culture. Highly ionized water-soluble drugs (such as penicillins and cephalosporins) may have a low ability to penetrate this barrier, whereas the use of non-ionized lipid soluble antibiotics (such as trimethoprimsulfamethoxazole, fluoroquinolones, metronidazole) have better penetration into the cyst and are generally preferred in the treatment of urinary tract infections in ADPKD. Cystic infection in ADPKD patients is not a very common occurrence in ADPKD patients. A study carried out by Sallee et al. among 389 identified patients with ADPKD revealed that 33 (8.4%) had 41 episodes of cyst infection, including eight definite and 33 likely cases. The incidence of cyst infections in patients with ADPKD was 0.01 episodes per patient per year. Positron emission tomography scan will probably make the diagnosis of cyst infection more precise and accurate to make. However, we do not have such facilities in our centre.

Nephrolithiasis is another possible cause of pain. It has been noted to occur in about 20% of patients with ADPKD. However, renal stones symptoms are due to obstruction, with resultant pain, infection, nausea and vomiting. Asymptomatic haematuria, macroscopic or microscopic, or repetitive urinary infections are also common presentations. IVU, abdominal Ultrasound, and urinalysis did not give any evidence of renal stone in this patient. Pain in nephrolithiasis usually acute, however the patient had chronic pain.

The size and number of renal cysts can also be the cause of renal pain in a patient with ADPKD. Also, the increase in size of the kidney may also be the cause of the pain. Stretching of the renal capsule and/or traction on the renal pedicle secondary to increased renal weight causes chronic abdominal pain. Taking into consideration that other common causes of loin pains have been excluded, we do believe that the most likely cause of loin pain in this patient may be increased size of the cysts and stretching of the renal capsule and/or traction on the renal pedicle consequent to increased renal weight.

Treatment of chronic loin pain in ADPKD patients usually involves finding the possible cause of the pain and treating the condition. However, ADPKD with chronic pain may be difficult to manage. It is important to educate patients suffering from chronic loin pain that the pain may not be relieved permanently by...
any intervention method adopted, and also to offer them the treatment modalities available to enable them to cope with the pain. In our patient no specific cause was found however, we educated the patient about the possible cause of the pain which we have stated above. We also informed him that the management of this sort of pain in most cases involves an input from a pain management specialist; however we do not have one. He was informed to use none NSAIDs analgesic when he feels the pain is becoming distressing. Fortunately in his case the pain does not bother him much and he said he could cope without the analgesic now that he knows about the possible cause of the pain. In patients with severe pain some other methods of treatment include surgical decompression of the cysts, autonomic plexus blockade, de-innervation of the kidney and nephrectomy.\[^{[15]}\] The criteria for surgical intervention is failure of conservative management, as defined by one of the following: chronic pain that interferes with activities of daily living or decreases the quality of life, disability, and/or narcotic dependence.\[^{[11]}\]

Two years after we started managing the patient, he still had the dull chronic left loin pain, but this did not bother him. His worry was that if he develops End Stage Renal Disease (ESRD), will he be able to afford the cost of renal replacement therapy? A worry shared by millions of patients all over the world living with polycystic kidney disease. We spared a though for his worry. As at the time of reporting this case, his 24-hour creatinine clearance was 43ml/min/1.73m.\[^{[3]}\] ADPKD ultimately leads to the destruction of renal parenchyma in more than 50 percent of patients.\[^{[16, 17]}\]

CONCLUSION

ADPKD with chronic loin pain may be difficult to manage. A concept that must be employed in the treatment is to educate the patient that the pain may not be relieved permanently by any intervention method adopted. In patients with chronic pain, behavioral modification approaches can help them adapt so that their lifestyle will not be affected. Physicians should be aware of chronic loin pain problems in ADPKD patients so an approach to the management can be pursued, whenever they present with such problems.

REFERENCES


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Conflict of Interest: None declared

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