Original Research Article

Emphysematous polycystic infection in autosomal dominant polycystic kidney disease – A rare presentation and successful conservative management

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Abstract

Autosomal Dominant Polycystic Kidney Disease a common hereditary disorders which leads to end-stage renal disease. Emphysematous infection of a polycystic kidney is a very rare presentation and is usually fatal. Hence an high index of suspicion is needed to diagnose it early. We report a case of a 50 years old diabetic female, who is a known case of Polycystic Kidney Disease, presented with fever, flank pain and burning micturition. On subsequent evaluation she was diagnosed to have Emphysematous cyst Infection. She was successfully managed with percutaneous drainage and intravenous antibiotics.

Key words: Emphysematous Polycystic Infection, Autosomal Dominant Polycystic Kidney disease

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INTRODUCTION

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is one of the most common hereditary disorders leading to end-stage renal disease. Approximately 30%–50% of patients with ADPKD have one or more renal infection during their lifetime[1]. However, cyst infections with gas-forming organisms are rare with few case reports in literature. Emphysematous polycystic cyst infection (EPCI) is life-threatening and needs to be differentiated from emphysematous pyelonephritis which is characterized by the presence of gas in the collecting system, peri renal tissues, and absence of gas in the cysts. To date, antibiotic therapy combined with nephrectomy was done for a majority of cases in published literature. We report a rare case of EPCI and how we successfully managed it conservatively with antibiotics and percutaneous drainage at our department.

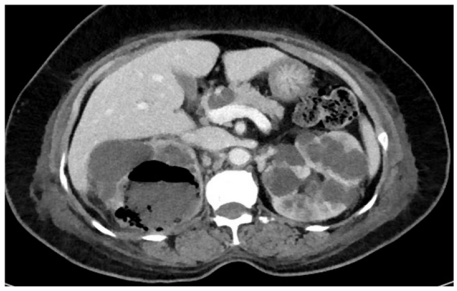

Figure 1: CT KUB showing polycystic kidneys with emphysematous infection

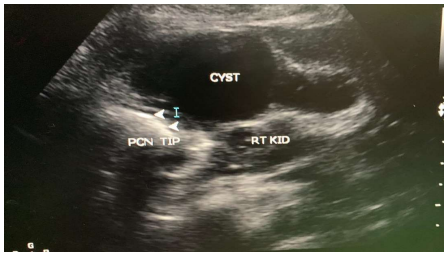

Figure 2: USG image showing location of percutaneous nephrostsomy tube in the cyst cavity

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CASE REPORT

A 50 years old diabetic female, presented to our outpatient clinic with history of fever, right-sided flank pain and burning micturition for 8 days duration. Examination revealed bilateral nodular abdominal masses with localized tenderness over the right flank. Investigations revealed an elevated leukocyte count of 27000, and urinalysis showed plenty of pus cells with a serum creatinine of 2.2mg/dL. Ultrasonography of the abdomen revealed bilateral enlarged kidneys with multiple cysts. Subsequently, contrast-enhanced computed tomography of the abdomen revealed bilateral polycystic kidneys with gas formation in the cyst of right kidney [Figure 1,2]. The patient was empirically started on Meropenam on day 1 of hospitalization. Subsequently, patient underwent percutaneous drainage of the cyst under ultrasound guidance. Cyst fluid analysis revealed numerous pus cells and culture grew Escherichia coli sensitive to piperacillin tazobactam. The patient improved symptomatically after the procedure, with the total count coming down to 9000 and creatinine coming down to 1. The catheter was removed after a week.

DISCUSSION

EPCI occurring with ADPKD is very rare with only 8 cases being previously reported [2]. The etiology of EPIC in ADPKD is not clear. It has been suggested that the preexisting damaged renal tissue and impaired vascular supply might be a precipitating factor, as certain bacteria (E.coli) can use the necrotic tissue as a substrate for gas formation[3,4]. Our patient appears to be at higher than normal risk due to the combination of diabetes and adult polycystic kidney disease. Mortality reportedly occurs in 20% of unilateral EPIC and 50% of bilateral EPIC, so appropriate management is essential[5]. Earlier reports showed a better survival with early aggressive surgical treatment in comparison to medical treatment[6] and decreased antibiotic perfusion was suggested to be the cause of failure of medical treatment.[3] Prompt diagnosis and surgical intervention was therefore suggested to be the most effective way of managing EPIC.[5,6] However an alternative medical approach with broad-spectrum antibiotics, maintenance of fluid balance, control of blood glucose and intensive circulatory support has been proposed recently[7,8]. But Intravenous antibiotic therapy alone may be insufficient to resolve emphysematous cyst infection considering disease severity and poor prognosis. A percutaneous cystostomy drainage of the infective foci along with intravenous antibiotics may be considered as an alternative therapeutic options to control emphysematous cyst infection instead of nephrectomy in patients with ADPKD. Huang and Tseng[9] suggested antibiotic treatment combined with percutaneous catheter drainage, wherever necessary, in patients with localized EPN and in patients with extensive EPN with a more benign manifestation (less than two of the following four risk factors including thrombocytopenia, acute renal function impairment, disturbances of consciousness, and shock). Our patient was managed conservatively with percutaneous cystosctomy and intravenous antibiotics and the outcome was good, probably because of early diagnosis, and appropriate antibiotic therapy.

CONCLUSION

We demonstrate that conservative management with percutaneous cystosctomy and intravenous antibiotics can be a successful modality in the treatment of EPIC, and is an acceptable alternative to surgical intervention. Till now, only eight cases have been described in literature of EPCI and only two of these patients improved with cyst drainage and antibiotics alone without requiring nephrectomy. To conclude, a high index of suspicion is required to diagnose this entity early, to reduce mortality.

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