Trop J Obstet Gynaecol, 18 (1), April 2001.

Polycystic Kidney Disease In Pregnancy In A Nigerian Woman

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Abstract

Adult Polycystic Kidney disease (ADPKD) is a known but uncommon cause of haematuria in pregnancy in this environment. Other causes include, haemaglobinopathies, calculi, pyelonephritis, schistosomiasis, haemangiomata and neoplasms. Although ADPKD is the commonest single gene disorder of man affecting both sexes, it usually presents in adulthood. It often presents in pregnancy with features of renal impairment, with or without haematuria. We present this case of ADKPD with life-threatening haematuria, acute renal failure, spontaneous abortion and recovery following nephrostomy tube insertion.

Key Words: *Adult Polycystic Kidney; Autosomal Dominant; Pregnancy.* [Trop J Obstet Gynaecol, 2001, 18: 40-42]

Introduction

There are generally two main categories of polycystic kidney disease, the infantile type, which is inherited in an autosomal recessive fashion and is usually fatal in childhood; and the adult type with an autosomal dominant mode of inheritance and a milder clinical course.

Adult polycystic kidney disease (ADPKD) is the commonest single gene disease of man with an incidence of 1:1000 [1]. It accounts for 10 per cent of end stage renal failure [2]. There are 3 types of ADPKD based on the nature of the mutant gene inherited. Type I is present on chromosome 16 p 13–3, type 2 on chromosome 4q 13–23 and type 3 in an unknown gene locus. The phenotypic changes are however similar in all three. The cortex and medulla of both kidneys are filled with thin-walled spherical cysts lined by cuboidal epithelium of varying sizes. They enlarge the kidneys and compress the nephrons, causing localised obstruction and impairment of renal function. They contain straw-coloured fluid which may become haemorrhagic due to trauma or infection. The patients usually present in the third or fourth decades with flank pain, gross and microscopic haematuria, nocturia and, sometimes, renal calculi. They usually do well in pregnancy especially where renal damage is minimal and hypertension is absent [3]. Eventually, up to 75 percent of cases will progress to chronic renal failure. Early detection in pregnancy depends on obtaining relevant family history and ultrasonography.

This case is being reported because of the atypical presentation, life–threatening haematuria and acute renal failure which necessitated emergency nephrostomy followed by spontaneous miscarriage. Current management guidelines are also discussed.

Case Report

A 21 year old Nigerian in her fourth pregnancy, having had two uneventful deliveries at term and one induced abortion, presented with haematuria and generalised abdominal pain worse on the right flank at 24 weeks gestation. She was found to be pale with a grossly distended and tender abdomen about the size of a 36 week cyesis with positive fluid thrill and multiple masses. Vaginal examination was normal. Abdominal Ultrasonography confirmed a viable, singleton fetus compatible with her dates. There were bilateral renal cysts ranging from 2 to 20 centimeters in diameter with the right worse than the left kidney (figs 1 and 2).

The haematocrit was 22 percent, haemoglobin (Hb) electrophoresis revealed HbAA, electrolytes and urea, creatinine and liver function tests were within normal limits. Urine culture yielded no growth and serial test-tube collection of urine revealed worsening haematuria.

Her condition deteriorated, with increasing abdominal distension, dypsonea, anaemia and diminishing renal function. She was placed on intravenous ampicillin 250mg with cloxacillin 250mg (Ampiclox) 6 hourly and intravenous fluid therapy. She was also given intramuscular pethidine 100mg as required for pain relief. Indwelling catheterisation and hourly fluid intake and output monitoring were instituted. She had 2 units of compatible O Rhesus positive blood transfusion and was given liberal oxygen by face-mask.

Seven days after admission the patient became anuric with serum creatinine of 360mmol/L, potassium of 6.2mmol/L, uric acid of 0.68umol/L, and the haematocrit was 16 per cent. On the same day, a nephrostomy tube was inserted into her right kidney, which yielded about 8.0 litres of blood stained fluid within a short period of time. This was done using local infiltration with 1 percent lignocaine and inhalational anaesthesia (Nitrous oxide and oxygen by face mask) because she was regarded as a grave anaesthetic risk.

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Figure 1: ***Shows multiple renal cysts in both kidneys***

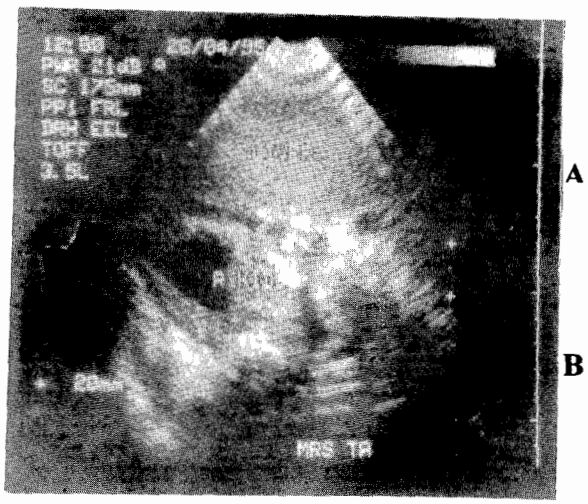

Figure 2: ***Post-operative ultrasound showing normal liver (A) and cystic right kidney(B).***

She had further blood transfusion as required. Within 48 hours, the effluent from the nephrostomy tube became clear. Haemodialysis was contemplated but was shelved because she progressed to the polyuric phase of acute renal failure.

Five days after the nephrostomy tube insertion, she had spontaneous complete miscarriage of a 300gm fetus, followed by gradual return of renal function. Biochemical analysis of effluent from the nephrostomy was similar to that of urine. Drainage from this tube continued to diminish while that from the urethral catheter increased. The nephrostomy tube was removed on the twenty-third day and the urethral catheter was removed shortly afterwards. Abdominal ultrasound confirmed a considerable reduction in the size of the right kidney with some remnant cystic areas, while the left kidney remained unchanged. She was allowed home 42 days after admission, when renal function had returned to normal. Although she was given an outpatient follow-up clinic appointment, she defaulted and has since been lost to follow up.

Discussion

This case of congenital ADPKD is being presented as an unusual example of a common condition, which became complicated and threatened the life of the patient. She is one of twins but her twin sister did not have polycystic kidneys on ultrasound. The appearance of gross haematuria for the first time in pregnancy should raise the possibility of ADPKD and prompt the exclusion of other causes such as spontaneous or traumatic kidney rupture, primary or secondary neoplasms, haemangiomata, calculi, pyelonephritis or fungal diseases involving the urinary system.

The presence of a positive family history, bilateral flank pain, increasing abdominal distension with encysted fluid should always lead to the suspicion of ADPKD. The diagnosis can easily be confirmed by abdominal ultrasound. In 30 percent of cases, there is co-existence with hepatic cysts or intracranial aneurysms [1].It has also been stated that only 17 per cent of cases are diagnosed by the age of 25 years [4].

Haematuria in this case was probably caused by rupture of some of the over-distended renal cysts in the right kidney. The anuria could be explained by the combination of outflow tract obstruction by some of the cysts and pre-renal hypovolaemic shock.. The only way to secure temporary drainage in the right kidney was through a nephrostomy, which decompressed the kidney and led to recovery from acute renal failure (ARF). A nephrectomy could be another option of management in similar circumstances but the parlous clinical condition of our patient precluded this major surgical option. Therefore nephrostomy was performed to tide the patient over the critical period of ill health.

Therapeutic termination of pregnancy was entertained as a management option prior to nephrostomy, but the patient's condition was so poor that it was shelved. However, the spontaneous miscarriage was the inevitable consequence of severe anaemia, hypovolaemic shock, ARF and the stress of anaesthesia and surgery. Other workers [5] have utilized prophylactic haemodialysis during pregnancy and meticulous fetal surveillance to improve outcomes in patients with ADPKD.

Currently, prenatal diagnosis and selective pregnancy termination of the affected fetus is available in developed countries where DNA probes and molecular genetic analysis are utilized for accurate diagnosis. This patient made satisfactory recovery and was able to retain functional kidneys.

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