

CLINICAL IMAGES

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Polycystic kidneys occupying entire abdominal cavity

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CASE REPORT

A 54-year-old female patient, pharmacist by profession, met her family doctor for annual health check-up. She was in her usual state of health. She is a known hypertensive, well controlled with anti-hypertensives. Patient had no other past medical or surgical history. Her family history was significant for polycystic kidney disease. Patient is a non-smoker denied alcohol consumption and drug abuse. The physical examination was unremarkable. Routine blood tests demonstrated elevated serum creatinine level of 2 mg/dL (normal 0.7–1.4 mg/dL). An ultrasound examination was performed which showed bilateral very large cystic kidneys consistent with polycystic kidneys. This was followed by a CT scan of the abdomen which showed that both kidneys are replaced by numerous cysts typical for polycystic kidney disease. The right kidney measures 22x17.6x24 cm, and left kidney measures 17x29x24 cm. The entire bowel was displaced anteriorly by the large kidneys. There was a 1 cm cyst seen in the right lobe of liver. The CT scan confirmed the diagnosis of very large polycystic kidneys occupying almost the entire abdomen and pelvis (Figures 1 and 2). Over the next one year the patient's renal function deteriorated to GFR of 6, serum creatinine level 7 mg/dL (normal 0.7–1.4 mg/dL), BUN 42 mg/dL (normal 7–26 mg/dL), serum potassium level 6.2 mmol/L (normal

3.5–5 mmol/L). Hemodialysis was initiated through a dialysis catheter then via a dialysis access AV fistula. She also started having early satiety. She underwent bilateral nephrectomy making room for the renal transplant and also to relieve the compressive gastrointestinal symptoms. Currently, she is on renal transplantation list and getting hemodialysis three times a week.

DISCUSSION

Autosomal dominant polycystic kidney disease (ADPKD) is a hereditary disorder characterized by multiple renal cysts and various systemic manifestations [1]. Massively enlarged kidneys with innumerable cysts characterize it. Grossly the kidneys are enlarged; the cysts are well defined round or oval with thin imperceptible or calcified wall. Cysts are filled with clear, serous, turbid, or hemorrhagic fluid. Microscopically: the cysts are lined by simple flattened or cuboidal epithelium with or without calcification [2].

ADPKD or adult PKD is one of the most common inherited disorders [1]. Abnormality in genes located on chromosome 16 and 4 leads to proliferation of renal tubular cells leading to formation of diverticulae of nephrons (collecting ducts) ultimately causing cystogenesis [1, 2]. It is a multisystemic disorder. The cystic manifestations of ADPKD are seen in kidneys (100%), liver (75%), pancreas (10%); spleen, ovaries, testis, and seminal vesicals [3]. Noncystic manifestations of ADPKD are cardiac valvular disorders (26%), hernias (25%), colonic diverticulae, cerebral "berry" aneurysms (5–10%) aorta and coronary arteries aneurysms [3].

It can present in any age group. It has equal preponderance in males and females. The incidence in white population is 1 in 400 to 1 in 1000. Patients can be asymptomatic or present with flank pain, hematuria, hypertension or renal failure. The disease process can be complicated with hemorrhage, infection, rupture, renal calculi and renal failure [1].

Ultrasound shows multiple, well-defined, round, anechoic areas in both enlarged kidneys. Ultrasound has

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Received: 20 March 2012

Accepted: 08 May 2012

Published: 01 October 2012



Figure 1: CT scan (coronal view) showing PKD occupying the entire abdomen.

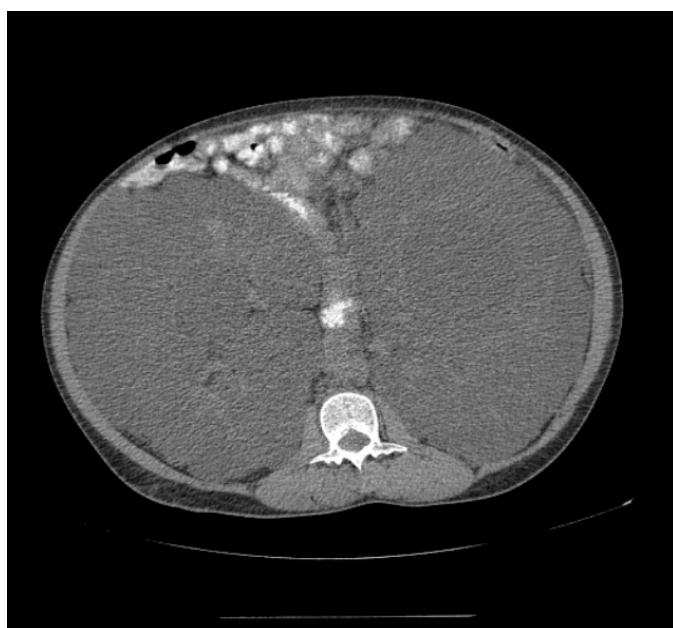


Figure 2: CT scan (transverse view) showing PKD occupying and anterior displacement of bowel.

sensitivity (97%), specificity (100%), accuracy (98%) [4]. It is recommended for screening family members.

CT scan shows multiple, well defined, round or oval cysts; variable in size; bilateral; with or without calculi. In the early stage, kidneys are normal in size and contour. In the later stage, there is increase in size and number of cysts, decrease in renal volume; with or without asymmetrical kidneys. Bosselated kidneys: is a term given to describe multiple cysts projecting beyond renal contours.

The treatment is usually symptomatic and managing complications like hypertension, pain, and renal infections. Once patients develop end stage renal failure dialysis or renal transplantation is necessary. The prognosis is fair, following renal transplantation like any other chronic renal failure [1]. The indications for pre transplant nephrectomy in ADPKD are for making space for the renal transplant, uncontrollable symptoms like pain, massive hematuria, recurrent infections and gastrointestinal compressive symptoms [5].

CONCLUSION

ADPKD causing massively enlarged kidneys occupying the entire abdominal cavity compressing adjacent visceral organs is rare. We should be aware that bilateral nephrectomy is indicated if the patient is symptomatic. The long-term prognosis for such patients is still good with renal replacement therapy, initially with dialysis and ultimately renal transplantation.

Gundlapalli VS, Ramirez C. Polycystic kidneys occupying entire abdominal cavity. *International Journal of Case Reports and Images* 2012;3(10):60–62.

doi:10.5348/ijcri-2012-10-203-CI-16

Author Contributions

Vinay S Gundlapalli – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Carlo Ramirez – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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