

Case report

“ADPKD presenting as congestive cardiac failure secondary to dilated cardiomyopathy” - a case report

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Abstract:

ADPKD presenting as congestive cardiac failure due to dilated cardiomyopathy is rare. ADPKD is an inherited systemic disorder with major renal manifestation and in some cases extrarenal manifestation or combination of both. In this report 45 year male patient presented with complains of dyspnoea, abdominal distension, pain right hypochondriac region. He was hospitalized, examined clinically and advised various bio-chemical and imaging tests. The finding was suggestive of ADPKD with dilated cardiomyopathy with congestive cardiac failure. He was managed with diuretics, ACE inhibitors, digoxin, Moist oxygen inhalation and he responded to the treatment.

Key words: ADPKD (Autosomal Dominant Polycystic Kidney Disease); CCF (Congestive Cardiac Failure)

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Introduction: ADPKD is an inherited systemic disorder that predominantly affects the kidney but may affect liver, pancreas, brain, cardiac and arterial blood vessels¹. The symptom or sign of ADPKD typically occurs between ages of 30-50 years². ADPKD disease occurs world wide and affects 1 in 400 to 1 in 1000 people³. Presentation of ADPKD as congestive cardiac failure due to dilated cardiomyopathy is rare.

Case Report: A 45 year old man patient presented as an outpatient with complains of dysnoea, abdominal distension and pain abdomen right hypochondriac region for last 15 days. Past history of similar episodes on and off for last 6-7 months was present for which patient use to take medications which subsided the symptoms.

Examination: Patient had bilateral pitting oedema, engorged but non pulsatile neck veins. Blood pressure was 138/76 mm Hg. On auscultation heart sounds were normal, chest revealed bilateral basal crepts. Abdominal examination revealed enlarged and tender liver measuring 4 cm in mid clavicular line.

Investigations: Routine biochemical test were within normal limits except serum creatinine which was 1.62 mg / dl. Chest x-ray PA view revealed

Cardiomegaly with bilateral increased broncho-vascular marking. Ultrasonography whole abdomen findings were suggestive of multiple cystic lesion of varying size with enlarged both kidneys. Multiple cyst in both lobes of liver with enlarged size measuring 17.2 cm with dilated inferior vena cava was present. Ultrasonography findings were also suggestive free fluid in peritoneal cavity with mild right pleural effusion. ECG suggested left ventricular hypertrophy. Echocardiography revealed dilated cardiomyopathy with hypokinesia with systolic dysfunction (EF= 44.34 %)

Management: Patient was given diuretics, ACE inhibitors, antiplatelet, inotropes (digoxin) to which he responded, his oedema and engorged neck veins subsided. Liver became non tender but was palpable. Later on he was advised beta-blocker (carvedilol) after 10 days when his crepts disappeared. At follow-up 3 weeks later patient was completely symptoms free except dysnoea on exertion. Patient was continuing medications as advised. His serum creatinine was 1.46 mg/ dl.

Discussion: ADPKD is the most common form of polycystic kidney disease characterized by presence of cysts throughout cortex and medulla. Most cases are identified between 30-50 years of age(2). Due to early

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screening, hypertension has become the most common form of presentation, in patients with ADPKD(4),⁵. ADPKD is an important cause of Renal failure requiring hemodialysis. Lion or back pain is seen in 50 to 70 % of patients with ADPKD⁶. The pain may be colicky, acute or chronic. Colicky pain is attributed to passage of stones or clots. Gross & microscopic haematuria is common and present in 60 % of patients overall⁷. Haematuria may be preceded with or without Abdominal trauma. Renal stones and urinary tract infection (Pyelonephritis) is seen in 20-30 % patients⁸. ADPKD also causes extrarenal manifestation like cerebral Aneurysm, Colonic diverticuloses, Abdominal inguinal hernia, cardiac manifestation and liver cysts^{9,10}. Among cardiac manifestation mitral valve abnormalities including Mitral valve prolapse and mitral regurgitation are common in patients with ADPKD¹¹. Non compaction cardiomyopathy

(NCC) association with ADPKD is reported in 3 cases of polycystic kidney disease¹².

Conclusion:

Hypertension, protienuria, microscopic haematuria and flank pain are the most common clinical presentation. Although most cases are identified between 30-50 years age, the condition has been recognized in children and utero(13). Upto 50 % patient with ADPKD require renal replacement therapy by 60 years of age, and accounts for 10-15% of patients who receive haemodialysis. Besides the above presentation, ADPKD can also present itself as C.C.F. in normotensive individuals secondary to cardiomyopathy. From above description it is evident that ADPKD despite being common but its presentation of C.C.F. which is rare, made it essential that all other cause of C.C.F. being ruled out establishing the diagnosis of ADPKD with congestive cardiac failure with dilated cardiomyopathy in patient with normal renal function.

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