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A Rare Case of Flash Pulmonary Edema

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ABSTRACT

Autosomal dominant polycystic kidney disease(ADPKD) is a common disorder occurring 1 in 1000 live births .ADPKD is also the fourth most common cause of kidney failure. ADPKD is a systemic disease with extrarenal manifestations involving the cardiovascular system. Cardiovascular disease (CVD) is the major cause of mortality in ADPKD. Abnormalities in the intracellular calcium pathway have been linked to cardiomyopathy in ADPKD.FLASH PULMONARY EDEMA is a severe form of acute LV failure that occurs when there is a sudden increase in left ventricular end diastolic filling pressure in DCMP.

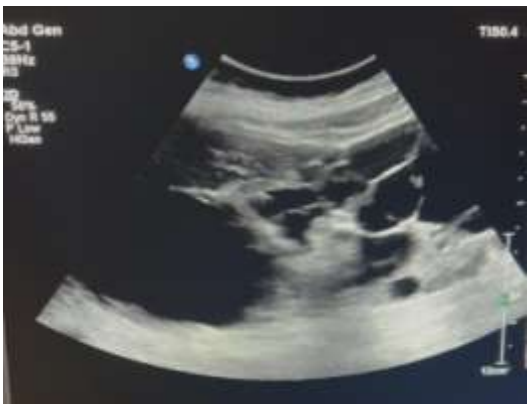
KEYWORDS: Autosomal dominant polycystic kidney disease, Dilated cardiomyopathy, Flash pulmonary edema, Erythropoeitin , End stage kidney disease.

I. CASE REPORT

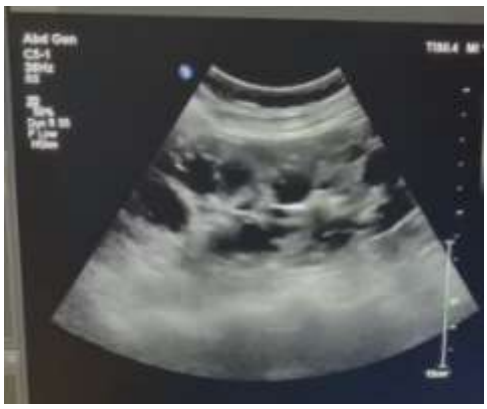
A 59 year female , known hypertensive and diabetic , presented to ER with complaints of shortness of breath on mild exertion NYHA III-IV since one month , insidious in onset, gradually progressive, no postural and diurnal variation and aggravated suddenly since few hours before presenting to casualty . Patient gives no history of cough and expectoration , or hemoptysis or chestpain or any decreased urine output or swelling of feet.

On examination patient was fully conscious, oriented, afebrile, no signs of pallor, PR 97/min, BP 140/100, bilateral air entry with basal fine crepts on auscultation, S1 S2 heard; with oxygen saturation 89% @room air; per abdomen was soft , bowel sounds heard.

Basic investigations done in ER; Hb 12g/dl, Sr creat/blood urea 3.1/88.5, grbs 107: Complete urine examination showed 25-30 RBC per HPF . USG abdomen was done which showed bilateral bulky kidneys ; right kidney measuring 187*66mm with largest cyst measuring 70*57 mm in upper pole and left kidney of 145*60 mm with largest cyst measuring 55*30 mm in lower pole with cystic pattern suggesting polycystic kidney disease and also liver with multiple cysts.



Rt kidney



Lt kidney

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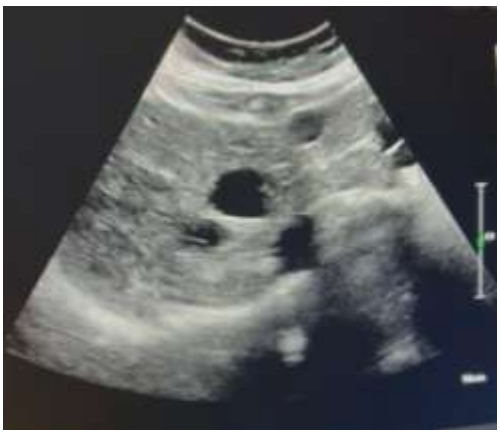
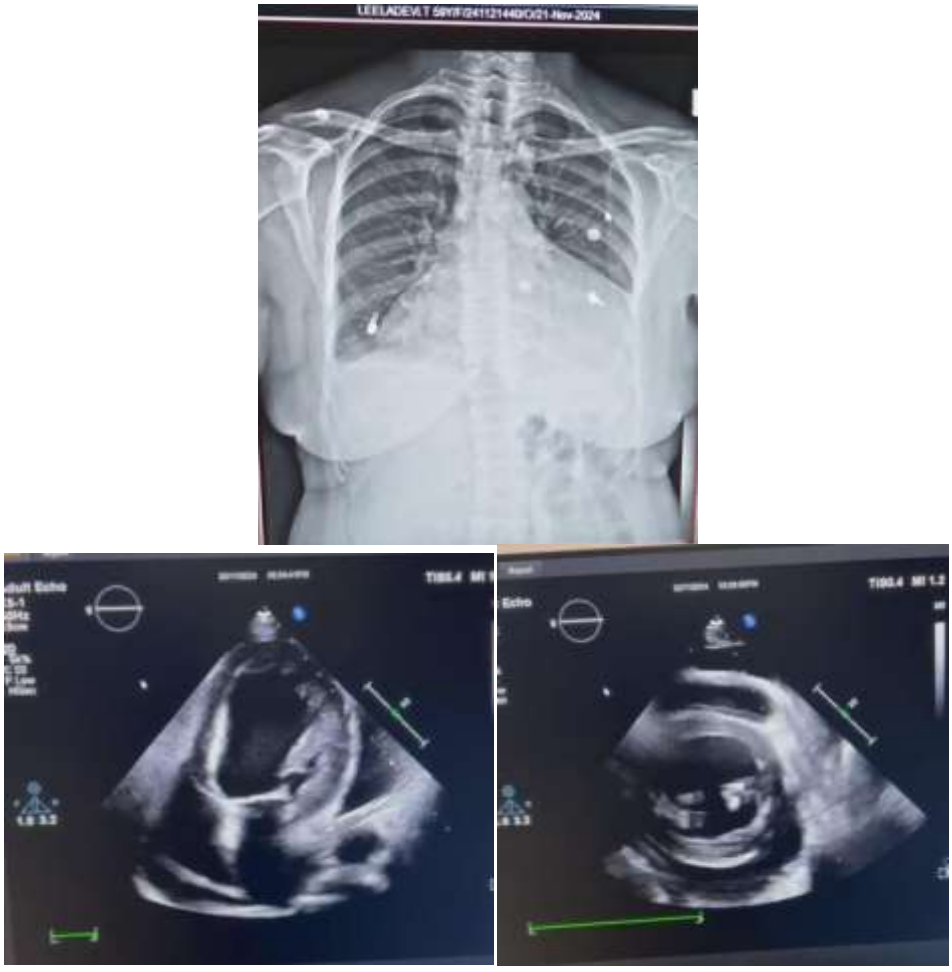


Hepatic cysts

Routine chest xray showed huge dilated heart.

2DECHO was done which showed global hypokinesia of LV, moderate PE, Concentric LVH, Dilated LV, Severe LV dysfunction with EF 30%.



During the hospital course, the patient was treated with iv diuretics ,beta blockers and SGLT2 inhibitors according to the heartfailure treatment protocol. Erythropoeitin levels (EPO) were 40.30 mIU/ml

Her both sons were also screened for any inheritance of polycystic kidney disease. But

ultrasound abdomen of both of them showed normal kidneys.
Patient was discharged on day 3 on the request of patient's attendants.

II. DISCUSSION

Autosomal dominant polycystic kidney disease(ADPKD) is a common disorder occurring 1 in 1000 live births.ADPKD is also the fourth most common cause of end stage renal disease (ESRD). It is characterized by excessive fluid secretion in the kidneys leading eventually in to end stage renal disease (ESRD) . It is predominantly caused by mutations in one of the two genes : PKD 1 (which encodes polycystin -1) on chromosome 16 and PKD 2 (which encodes polycystin-2) on chromosome 4. Polycystin-1 is a transmembrane protein in the cell membrane and primary cilia Polycystin 2 is a member of transient receptor protein channel that regulates calcium from intracellular stores .

Kidney cyst growth in ADPKD is associated with regional hypoxia mostly due to mismatch between the enlarged cysts and the peritubular capillary blood supply and compression of peritubular cells in cyst walls. This hypoxia activates hypoxia inducible transcription factors in two of its isoforms : HIF1 is present in the cyst-lining and HIF 2 in the pericystic interstitial cells. HIF 2 stimulates erythropoietin (EPO)production and thus mitigating the anemia which is present in chronic kidney disease. HIF 1 promotes cyst growth. HIF-dependent cyst growth is primarily due to an increase in chloride- dependent fluid secretion into the cyst lumen. Also, hypoxia can influence cyst growth through the generation of reactive oxygen species. Since cyst expansion further aggravates regional hypoxia, a feed forward loop is established that accelerates cyst expansion and disease progression. Patients with ADPKD have higher levels of EPO in their serum than patients with other causes of end- stage renal disease.

ADPKD is a systemic disease with extrarenal manifestations involving the GASTRO INTESTINAL AND NON GASTROINTESTINAL

GI manifestations include livercysts(94% over age 35), diverticular disease(50-83% in ESRD patients), hernias(45%), pancreatic cysts (9-36%), Common bile duct dilation(40%), splenic cysts(2.7%), choledochal cysts(rare).

Non-GI manifestations include cardiovascular system abnormalities ,cerebral aneurysms(9-12%), arachnoid cysts (8-12%), spinal meningeal cysts (1.7%), seminal vesicle cysts(40%), bronchiectasis(37%), thyroid cysts.

Cardiovascular system manifestations include left ventricular hypertrophy , intracranial aneurysms, valvular heart diseases, arrhythmias and cardiomyopathies . It has been shown in various studies that PKD genes are expressed in cardiac myocytes. Abnormalities in the intracellular calcium pathway have been linked to cardiomyopathy in ADPKD. Mutations in PKD2 are linked with impaired intracellular calcium flux leading to decreased cardiac contractility, thin ventricular walls and dilated cardiomyopathy. While mutations in PKD1 can lead to cardiac hypertrophy and reduced myocyte fractional shortening, predisposing to cardiomyopathies. Idiopathic cardiomyopathy is characterized by LV dilatation and systolic dysfunction and hypertrophic obstructive cardiomyopathy is characterized by asymmetrical LVH and diastolic dysfunction. Diastolic dysfunction is generally reported in late stages of ADPKD with established kidney failure. Polycystin –related dysfunction of cardiomyocytes and vascular cells are likely involved in the pathogenesis of diastolic dysfunction. Diagnosis of cardiomyopathies are made by echocardiography.

FLASH PULMONARY EDEMA is a severe form of acute LV failure that occurs when there is a sudden increase in left ventricular end diastolic filling pressure in DCMP. It is characterized by sudden onset of respiratory distress related to accumulation of fluid in the lung interstitium over minutes to hours.

ADPKD is an autosomal dominant disease. So there is a 50% chance for the child to inherit the disease and the family therefore should be screened for the occurrence of this disease. Ultrasound is the most common, non invasive and affordable way to screen the disease. BP monitoring and urine tests for proteinuria are also helpful

The National Institute for Health and Care Excellence (NICE) recommends tolvaptan as a medication to treat ADPKD in adults. It can slow down the growth of cysts and preserve kidney function.

III. CONCLUSION

We report a case of a 59 year old female ,who is known hypertensive and diabetic presenting with flash pulmonary edema : On evaluation she has been diagnosed with an underlying Dilated cardiomyopathy (DCMP) with severe LV dysfunction .Her renal function tests were simultaneously deranged which rose suspicion and a USG abdomen was done which revealed bilateral polycystic kidney disease along with multiple

cysts in liver. She was treated according to the heartfailure treatment protocol in the hospital course and discharged on day 3 on the request of patients' family.

This case highlights the existence of cardiomyopathy in undiagnosed ADPKD which presented to ER with flash pulmonary edema.

Her EPO levels were also on the higher side supporting the incidence of higher EPO levels in ADPKD. Screening of the family members was also done as this is an autosomal dominant condition and it runs in families.

Early detection can reduce the morbidity among the patients and can promote also early screening among the family members and thus helping in delaying the adverse affects of this devastating disease.

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