Teaching File

Adult Polycystic Kidney Disease

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A case of asymptomatic adult polycystic kidney disease (APKD) in an experienced aircrew, detected on routine evaluation, is reported with its aeromedical disposal dilemma, follow up of two years and review of problems associated with APKD.

Part I : Initial Clinical Evaluation

A 41 year old fighter pilot with 3400 hours of flying experience, rated as QFI (A2) and Master Green was found to have nonspecific ECG abnormality (T-inversion in anterior leads), mild hypertension (140-150/100-104 mm Hg) and microscopic haematuria with several pus cells (Pyuria), suggesting urinary tract infection (UTI), during annual medical examination. He was asymptomatic and functional class I. He gave no history of colicky abdominal pain, fever, chest discomfort, difficulty in breathing, headache and icterus in the past. He used to smoke occasionally and had no family history of ischaemic heart disease (IHD) or sudden cardiac death. However there is a strong family history of hypertension and polycystic kidneys in the first degree relations and two of his cousins have undergone renal transplanation because of chronic renal failure due to severe bilateral polycystic kidney disease. Clinical examination revealed pulse 70/mt regular, normal and all peripheral pulses well felt, BP 150/100 mm Hg, no other abnormalities on general examination CVS, Respiration and CNS examination were normal. No carotid bruit was detected. Ocular fundi showed cupping of optic discs both eyes but no hypertensive changes or arterio- venous malformations were detected. Abdominal examination revealed both kidneys ballottable (left bigger than right), no hepatosplenomegaly and no tenderness in renal angles/urinary bladder area. Per rectal examination revealed normal prostate.

He was diagnosed to have ECG abnormality, hypertension and urinary tract infection (UTI). He also had enlarged kidneys, left more than right, the aetiology of which remained unclear. He was subjected to routine investigations, urine culture, biochemical investigations, X-ray chest and abdomen and Treadmill Exercise Test (TMT). Urine culture revealed E coli infection which was treated and he recovered. Other routine investigations and biochemical parameters were normal including blood urea (25 mg%) and uric acid (3.8 mg%). X-ray chest was normal but X-ray KUB revealed left kidney 2.0 cms larger than right and had irregular cortical outline. Treadmill Exercise Test (TMT) showed mildly positive response with normal heart rate and BP response during exercise and ST depression was mainly upsloping type which quickly reverted to baseline in post-exercise.

Question No 1 : What further investigations would you undertake at this time ?

Part II : Further Evaluation

He was subjected to abdominal ultrasound which revealed both kidneys enlarged and studded with multiple cysts of varying size filled with fluid and pelvi-ureteral system and urinary bladder were normal. Liver showed two small cysts in right lobe but liver size and parenchyma and vasculature was normal. Spleen showed no cystic structures. His urine output was stable but repeat evaluation after three months showed serum creatinine 1.2 mg% and creatinine clearance varying between 43 and 103 ml. Because of altered creatinine clearance with underlying APKD, he was kept under observation in non-flying category to assess & follow the progression of renal disorder over the time.

Question No 2 : How would you evaluate ECG abnormality in this case ?

Part III : ECG - Abnormality Evaluation & Disposal

Though TMT was mildly positive but he had associated hypertension and he was a smoker and hence the significance of ECG abnormality was found out by Thallium^{201} (TH^{201}) perfusion study and coronary arteriography. TH^{201} study showed no perfusion defects and coronary arteriography showed normal coronaries and normal LV functions. The ECG abnormality was considered nonspecific and benign and his BP was well controlled with diuretics alone and no target organ involvement was detected. He was adjudged fit to perform normal ground duties and restricted flying on this account. The Echo study showed mild concentric hypertrophy of left ventricle with normal systolic and diastolic functions. No mitral valve prolapse was detected.

Question No 3 : What is the Aeromedical disposal in this case regarding APKD?

Part IV : Aeromedical Disposal of APKD

This patient was serially evaluated from Dec 88 to Oct 90. His kidney size remained same and the renal functions stable. In Oct 90, the blood urea, uric acid and serum creatinine were 35, 4.3 and 1.0 mg% respectively and the creatinine clearance was 226 ml. There were no additional cysts in the liver, spleen was free of cysts, lungs had no cystic disease as X-ray chest was normal and his BP was adequately controlled this time even without the diuretics. No target organs involvement was detected. Resting ECG and response to exercise showed no deterioration. At this juncture considering his vast flying experience and keen interest to go back to flying, he was considered fit for restricted flying in medical category A3G3. A preliminary observation on fighter trainer aircraft revealed satisfactory performance without any symptoms. During re-evaluation, in addition to metabolic and clinical studies, he was subjected to centrifuge runs with anti-G suit and he could easily tolerate 4.5 G without symptoms. Considering no imminent danger from APKD, stable features on repeated evaluations and normal coronaries, he was recommended by medical board for full flying status in fighters in A2G2 (permanent) but approving authority at Air Headquarters converted it to co-pilot status in A3G2 (permanent) in transport or helicopters as there was unpredictable real danger of cyst rupture or subarachnoid haemorrhage causing sudden incapacitation in flight.

Question No 4 : What is the aetiopathogenesis and other problems associated with APKD ?

Part V : Aetiopathology of APKD

The APKD is an autosomal dominant disorder with high degree of penetrance, approaching 100% in those surviving through their 70s or 80s. In adults it is a relatively common condition, the incidence varying from 1/200 to 1/1000 and ultimately APKD cases account for about 10% of cases requiring chronic dialysis or renal transplantation. The genetic defect leads to multiple cyst formation in renal and extra-renal tissues (liver, spleen, lungs and pancreas). Microaneurysms in brain and mitral valve prolapse are often associated with the APKD[2]. The APKD may progress to turn both kidneys into bunch of cysts. The common renal problems in APKD are hypertension,UTI, renal stones, chronic renal failure (CRF) and higher incidence of renal cell carcinoma (hypernephroma). Unlike chronic glomerulonephritis, the CRF of APKD is associated with normal or raised Hb% because of increased erythropoietin induced erythrocytosis. The arteriovenous (A-V) malformations in cerebral circulation could really prove hazardous and may cause subarachnoid haemorrhage (SAH). The APKD is best diagnosed by abdominal ultrasound which can pick up even the smallest cysts too. However computerised tomography and magnetic resonance imaging may be used in doubtful cases to enhance diagnostic accuracy and image delineation. The cysts in other organs gradually progress and may cause functional derangement. The A-V malformations in brain remain silent till rupture and thus cause sudden SAH. However SAH is usually not massive to cause shock because of reflex spasm of vessels

distal to the site of rupture and the spasm itself later causes ischaemic cerebral infarction and neurological deficit and death. Also these cases tend to have recurrence of SAH due to rupture of multiple A-V malformations. The diagnosis can be made by CT/MRI or angiography, however smaller cysts may be missed.

Thus APKD is a systemic disorder with predominant involvement of kidneys and brain. Evaluation of APKD should be aimed to assess all the organ systems.

Question No. 5 : What is the current status on aeromedical disposal of APKD ?

Part VI : Aeromedical disposal of APKD

In a case of APKD with gross impairment of renal or cardiovascular functions, the aeromedical disposal is easy. But in an asymptomatic, normotensive, highly qualified, experienced and well motivated aircrew, the disposal raises several questions. Taking such an aircrew off flying permanently means loosing an experienced and trained aircrew. The risk of renal colic and potential for subarachnoid haemorrhage should preclude any category better than as or with a co-pilot[3]. This implied that the fighter pilot (as was the aircrew under reference) must undergo conversion to either a transport aircraft or a helicopter. It can be debated if the motivation of the fighter pilot for flying will remain high on conversion to the slower aircraft. The issue of flying a transport aircraft (military or civil) with passengers on board, also needs clarification. There have been no reports regarding grant of fighter flying with APKD and logically so because of potential danger of sudden mechanical incapacitation in flight due to SAH or acute abdomen. Another problem is mechanical i.e. possible rupture of large kidneys due to trauma. Therefore in uncomplicated cases of APKD, the disposal should be as in this case after thorough evaluation and serial follow up and periodic reviews are mandatory.

Question No 6 : What is the follow up information in this Case ?

Part VII : Follow up studies

This pilot has been under observation since reinduction into flying in Dec 90. His renal parameters remained stable, BP controlled without drugs and has been asymptomatic. He has been flying as co-pilot in transport aircraft. In Oct 92 he took premature retirement from service. In Dec 92, he developed sudden headache, giddiness and vomiting. He was found to have bradycardia (heart rate 45/minute), raised BP (200/120 mmHg, accelerated hypertension) and signs of meningeal irritation (neck rigidity) with drowsiness. Fundoscopy showed no papilloedema or vascular malformations. Lumbar puncture revealed uniformly haemorrahagic CSF and computerised tomography (CT) scan showed aneurysmal & tortuous left internal carotid artery with blood in 3rd and 4th ventricles. He was managed with Nimodipine (newer calcium channel blocker) 60 mg 6 hourly to prevent cerebral arterial spasm (causes cerebral infarction) alongwith diuretics, antibiotics, steroids and mannitol (to reduce intracranial tension). In view of high probability of recurrence of SAH (due to multiple A-V malformations) which may claim life, he was referred to neurocentre for four vessel angiography (bilateral carotids and vertrebrals) and possible surgery. He was subjected to cerebral angiography which revealed an aneurysm at the junction of middle cerebral artery and origin of posterior communicating. There was another aneurysm in the posteior cranial fossa. The posterior communicating aneurysm was clipped. On second post operative day, he developed sinking sensation, fall of BP, hyperventilation with bradycardia and clinically he had pulmonary congestion, which all suggested another bleed in cranium with neurogenic pulmonary edema and tachypnoea. Despite all supportive measures he died after two hours.

In light of these developments ie. fatal SAH in a case of APKD, we need to critically look back at our disposal regarding his flying status. Even if asymptomatic, the possible occurrence of unpredictable SAH excludes fighter flying but it may even be argued that what worth is achieved in granting him the transport flying when the real danger of SAH keeps lurking which may produce emergency in flight and compromise flight safety. Therefore it is felt that these individuals with

APKD should be permanently grounded. No amount of eccentric individual case reports should be used to mould a policy in APKD and flying.

This case of APKD with strong family history, detected on routing medical evaluation is reported to highlight this entity, associated problems (the prominent being fatal SAH) and the aeromedical disposal. The problem of SAH with APKD can not be overemphasised and this should be appreciated as a potential danger. The BP should be meticulously controlled. This also raises the question of undertaking CT brain in all cases of APKD to diagnose asymptomatic aneurysm to help further management or even undertaking four vessel angiography and prophylactic surgery to prevent SAM. The issues remains subjudice due to asymptomatic status of patients.

References

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