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Letter to Editor

Anaesthesia Section

Iatrogenic Carotid-jugular AV Fistula in a Patient with Polycystic Kidney Disease and Mitral Valve Prolapse: A Case of Diagnostic Complexity

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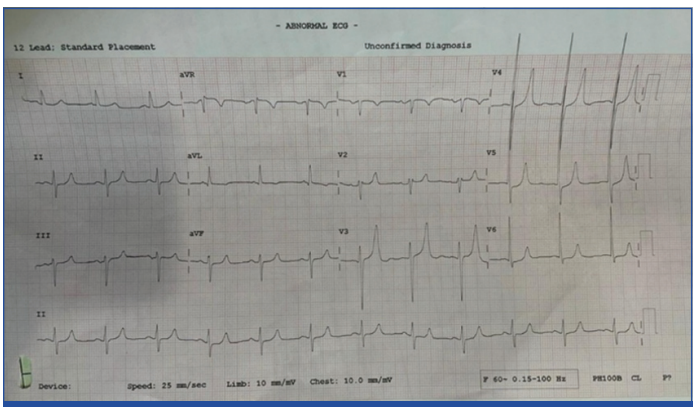
Dear Editor,

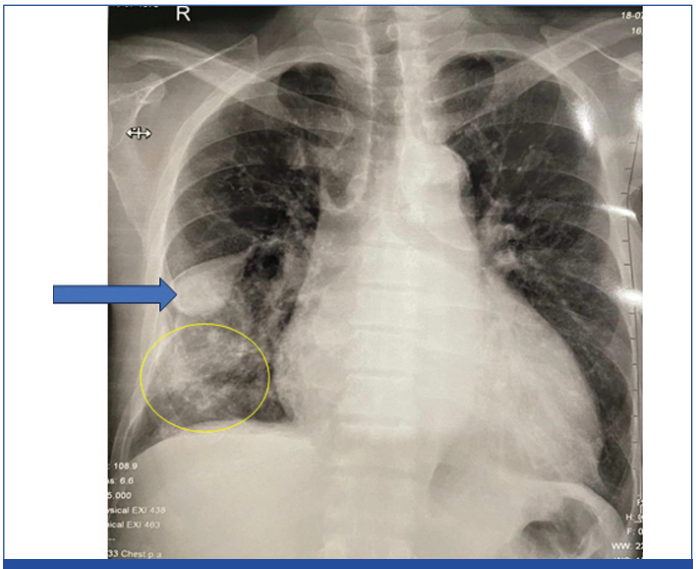
Autosomal Dominant Polycystic Kidney Disease (ADPKD) is characterised by multiple renal cysts, progressive kidney enlargement, and dysfunction [1], eventually leading to End-Stage Renal Disease (ESRD). Extrarenal manifestations include hepatic cysts, hypertension, Left Ventricular Hypertrophy (LVH), valvular heart disease, intracranial and extracranial aneurysms, pancreatic cysts, and diverticulosis [2]. Arteriovenous fistulae (AV fistulas) can be congenital, idiopathic, or acquired following trauma or iatrogenic procedures, such as Internal Jugular Vein (IJV) catheterisation [3,4]. This report highlights the need for clinician awareness and routine vascular imaging in high-risk ADPKD patients with a history of jugular catheterisation.

A 46-year-old male presented with bilateral leg swelling, oliguria, exertional dyspnoea, and fatigue. He was a known case of ADPKD and had been on maintenance haemodialysis for the past 10 years. His last session was two days prior to surgery via a right forearm AV fistula. A right internal jugular permanent catheter, which is a tunneled central venous catheter used for long-term haemodialysis access, had been placed eight years prior. There was no significant dental history.

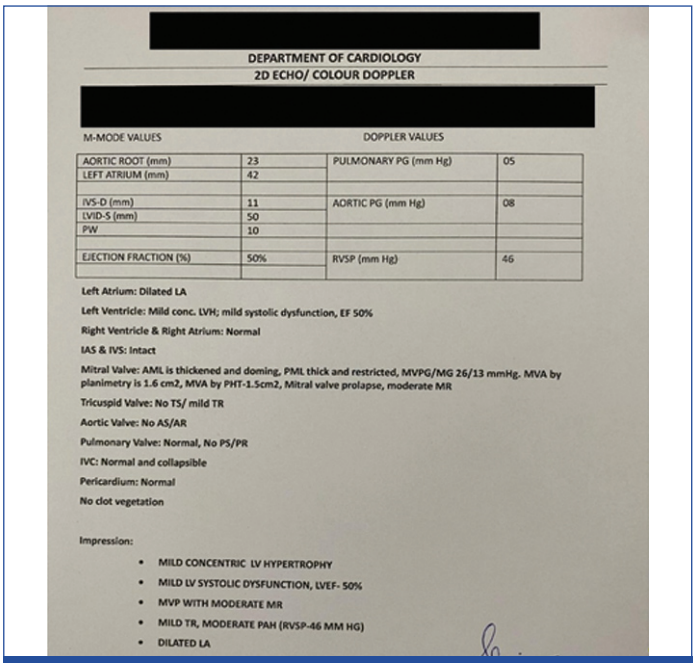
On examination, he exhibited moderate anaemia with facial puffiness, grade 3 pitting oedema, mild ascites, a heart rate of 105 beats/min, and blood pressure of 168/97 mmHg. An Electrocardiogram (ECG) revealed sinus tachycardia, tall T waves in leads V3-V5, and LVH [Table/Fig-1]. A chest X-ray showed right lung consolidation, pleural effusion, and cardiomegaly [Table/Fig-2]. Echocardiography demonstrated a left ventricular ejection fraction of 50%, LVH, dilated left atrium, Mitral Valve Prolapse (MVP) with moderate Mitral Regurgitation (MR), moderate pulmonary artery hypertension, and mild tricuspid regurgitation [Table/Fig-3]. Auscultation revealed a systolic murmur and basal crepitations. Ongoing medications included ramipril 2.5 mg once daily, metoprolol 25 mg once daily, tolvaptan 45 mg in the morning and 15 mg in the evening, lasilactone 50 mg once daily, torsemide 5 mg + spironolactone 50 mg, and aspirin 75 mg at night.



[Table/Fig-1]: Pre-operative electrocardiogram.



[Table/Fig-2]: Pre-operative chest X-ray showing right lung consolidation and pleural effusion.

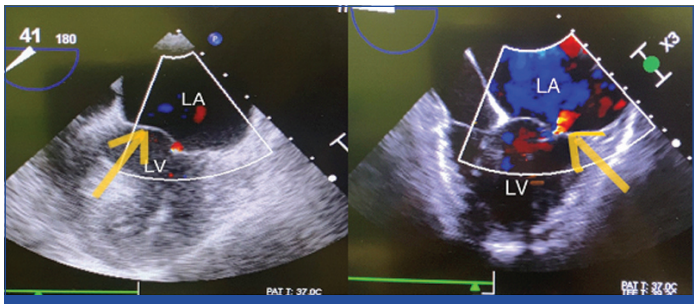


[Table/Fig-3]: Pre-operative echocardiogram.

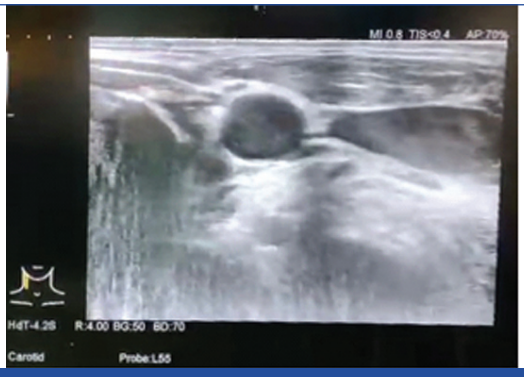
Based on clinical and imaging findings, a provisional diagnosis of chronic kidney disease stage 5 on regular haemodialysis and underlying valvular heart disease was made.

The treatment plan involved immediate dialysis, adjustment of cardiac and antihypertensive medications, and close haemodynamic monitoring. He was scheduled for right deceased-donor renal transplantation following standard preoperative clearance.

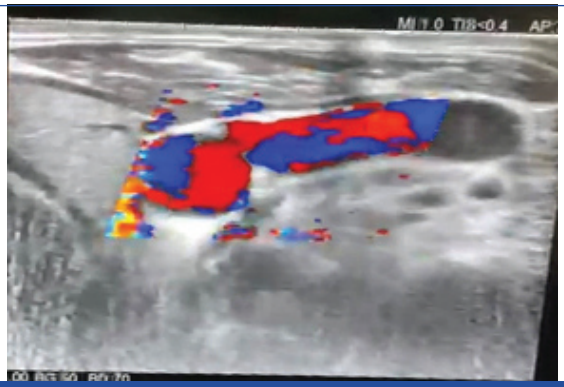
On the day of surgery, Transesophageal Echocardiography (TEE) was used in addition to standard intraoperative monitoring [Table/Fig-4,5]. A left radial artery line was cannulated under local anaesthesia. General anesthesia was induced with intravenous midazolam 0.02 mg/kg, fentanyl 2 µg/kg, etomidate 0.3 mg/kg, and cisatracurium 0.15 mg/kg, followed by intubation with an 8.0 mm cuffed endotracheal tube. While preparing for right IJV cannulation, a pulsatile neck swelling with engorged veins was observed. Ultrasound and color Doppler confirmed a high-flow AV fistula between the right common carotid artery and IJV [Table/Fig-6,7]. It was presumed that the permanent catheterisation of the right IJV performed eight years earlier caused the AV fistula. Cannulation was safely planned on the left side, and the transplant surgery proceeded uneventfully.



[Table/Fig-4,5]: Transesophageal Echocardiography (TEE): showing mitral valve leaflet billowing, colour Doppler in TEE showing flow between left atrium and left ventricle showing Mitral Regurgitation (MR).



[Table/Fig-6]: Ultrasound showing a fistula between the right common carotid and the Internal Jugular Vein (IJV).



[Table/Fig-7]: Colour Doppler showing turbulent flow between the right common carotid and the Internal Jugular Vein (IJV).

To manage hypotension and blood loss, volume resuscitation was initiated along with phenylephrine infusion at 1 µg/kg/min to maintain Mean Arterial Pressure (MAP), control tachycardia, and ensure adequate tissue perfusion. The transplanted kidney produced 250 mL of urine within four hours after ureteric anastomosis and reperfusion. Intraoperative analgesia included intravenous paracetamol 15 mg/kg and fentanyl boluses of 0.5 µg/kg twice in response to tachycardia and a rise in blood pressure.

The patient was transferred intubated to the transplant recovery unit, with extubation planned for the next morning. Multidisciplinary consultation with a cardiologist, vascular surgeon, interventional radiologist, and the consulting nephrologist was initiated, and the patient was informed about the AV fistula postoperatively. Endovascular closure with a covered stent was scheduled at another hospital following discharge.

While many iatrogenic AV fistulas remain asymptomatic, larger fistulas can cause significant haemodynamic instability and may be fatal without timely detection. A carotid-internal jugular fistula leads to high-output heart failure due to direct arterial flow into the vein, which increases venous return and cardiac workload over time [5]. Clinical signs include pulsatile neck swelling, bruit, tachycardia, palpitations, arrhythmias, elevated jugular venous pressure, fatigue, pulmonary congestion, peripheral oedema, orthopnoea, and paroxysmal nocturnal dyspnoea. Pulmonary congestion may present as basal crepitations. To maintain perfusion, the heart compensates by increasing heart rate and stroke volume [6]. However, blood diversion into the venous system may impair cerebral circulation, causing syncope or stroke. In patients with pre-existing MVP, a fistula can worsen MR, increasing preload and afterload, leading to left atrial dilation, ventricular overload, and eventually systolic dysfunction, which clinically presents as worsening heart failure and a new or louder systolic murmur [7].

Essential investigations include echocardiography to assess MR severity and ventricular function, ECG for arrhythmias, chest X-ray for pulmonary oedema, and Brain Natriuretic Peptide (BNP) levels as a marker for heart failure. First-line therapy includes Angiotensin-Converting Enzyme (ACE) inhibitors, with Angiotensin Receptor Blockers (ARBs) as alternatives [2], followed by diuretics to manage volume overload. If blood pressure remains uncontrolled, calcium channel blockers or beta-blockers may be added [2]. Sacubitril/valsartan, an Angiotensin Receptor-Neprilysin Inhibitor (ARNI), has shown benefits in heart failure, while ivabradine is beneficial for patients with sinus rhythm and a resting heart rate >70 beats/min [8]. In MVP with MR, bradycardia should be avoided to prevent worsening of regurgitation.

Anaesthetic goals include maintaining haemodynamic stability by cautiously titrating anesthetic agents, balancing fluid management to optimise preload without causing pulmonary congestion, and using invasive monitoring and intraoperative TEE guidance [8]. Endovascular closure with a covered stent or coil embolisation is the first-line treatment for AV fistulas; surgical repair is reserved for cases deemed unsuitable for endovascular approaches [9]. MVP requires ongoing monitoring, with echocardiography every 6-12 months if MR remains mild to moderate. Surgical intervention is indicated for severe MR or if ventricular dysfunction develops [7].

In this case, an undetected carotid-internal jugular AV fistula compounded the cardiovascular burden in a patient predisposed to ADPKD and MVP. Similar conclusions were made in other studies, stating that MVP is an established cardiac manifestation in ADPKD and contributes to the overall cardiovascular burden in affected individuals by compounding risks already elevated by hypertension and other structural heart changes [10,11]. This incident prompted a revision of our institutional protocols to mandate Doppler ultrasonography of the neck and peripheral vessels in transplant recipients with prior central venous access. Essential investigations, including echocardiography and Doppler ultrasound, are crucial for assessing haemodynamic status and detecting abnormal flow patterns [12]. Therefore, routine preoperative vascular imaging should be emphasised, particularly in transplant recipients, to identify

undetected anomalies, minimise intraoperative risk, and optimise anesthetic and surgical planning. Additionally, we recommend heightened awareness among anaesthesiologists, nephrologists, surgeons, radiologists, and cardiologists to anticipate such findings and communicate them effectively with the multidisciplinary team and patients.

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