

From Hospice to Home: Re-emphasizing the role of PCI in Pickering Syndrome

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Introduction

Pickering syndrome (PS) is a clinical syndrome of recurrent flash pulmonary edema (FPE) in the presence of bilateral renal artery stenosis (RAS), in those without underlying cardiac disease. Interestingly, it can also occur with unilateral RAS if there only one functioning (solitary) kidney. In patients with valvular disease or heart failure, recurrent FPE from true PS can be overlooked.

We present this diagnostic dilemma, in a woman who was initially admitted for diastolic heart failure destined for hospice given her recurrent hypoxic respiratory failure and dependence on ultrafiltration (UF). However, with appropriate intervention of this rare clinical entity allowed her to be discharged home with an excellent prognosis.

Case Presentation.

77-year-old female admitted for a one-week history of dyspnea. She has a pertinent cardiac history for heart failure with preserved ejection fraction (HFpEF) with a several admissions over the past few months, moderate aortic stenosis and mitral regurgitation. Prior to admission she was started on isosorbide mononitrate 30 mg PO daily and losartan 50 mg PO daily due to uncontrolled blood pressure. Other pertinent home medications included: Aspirin 81 mg PO daily, Metoprolol tartrate 25 mg PO BID, Furosemide 40 mg PO daily, and Amlodipine 10 mg PO daily.

On admission, her vital signs were 190/110 mmHg, 64 (sinus) beats/min and SpO₂ 50% requiring non-invasive mechanical ventilation (NIMV). Her clinical exam was consistent with congestive heart failure. Initial ECG at baseline was normal sinus rhythm with non-specific ST-segments changes. At that time, 2D Trans-thoracic ECHO showed an ejection fraction of 65% with moderate AS and MR. She had a previously normal coronary angiogram. Initial labs showed a Creatinine of 1.0 mg/dL. She was managed with blood pressure control and IV diuretics.

Over 1-week her serum creatinine increased to 4.7 mg/dL. Despite hypervolemia and further diuresis her serum creatinine increased to 4.7 mg/dL and progressed to anuric AKI requiring HD removing two-to-three L of ultrafiltration. Over the next two weeks, her hospital stay was complicated by recurrent episodes of acute pulmonary edema, hypoxic respiratory failure in the setting of uncontrolled hypertension, despite having normal volume status and normal blood pressures the day prior to each episode.

At this juncture, her prognosis was poor as she developed recurrent pulmonary edema that required NIMV and HD. Given her treatment was refractory to all therapies, hospice was discussed until a rare, alternative diagnosis was entertained.

A pre-dialysis renal ultrasound had showed normal renal echogenicity with a size difference of about 1 cm. This coupled with the episodes of recurrent flash pulmonary edema prompted consideration of reno-vascular hypertension and a renal artery duplex scan was performed. This revealed an atrophic left kidney and renal artery, and significant stenosis of the proximal right renal artery. Renal angiography confirmed the duplex scan findings and right renal artery received percutaneous intervention (PCI) with an 7.0 x 18 mm Herculink Elite bare-metal stent. 48-hours post PCI, her renal function returned to baseline and she was discharged home.

Discussion and Conclusion

The diagnostic dilemma in this case was that her symptoms were thought to be related to her underlying valvular disease and diastolic dysfunction. However, the recurrent episodes of severe FPE with uncontrolled hypertension, was not entirely explained by a primary cardiac pathology, therefore prompting an alternative diagnosis.

The prevalence of PS is not well defined, but according to Messerli et al, 14.3% of those with bilateral experience FPE compared to 3.5% with unilateral RAS. RAS should be suspected in refractory hypertension, recurrent congestive heart failure, intolerance to aldosterone receptor blockers and angiotensin inhibitors.

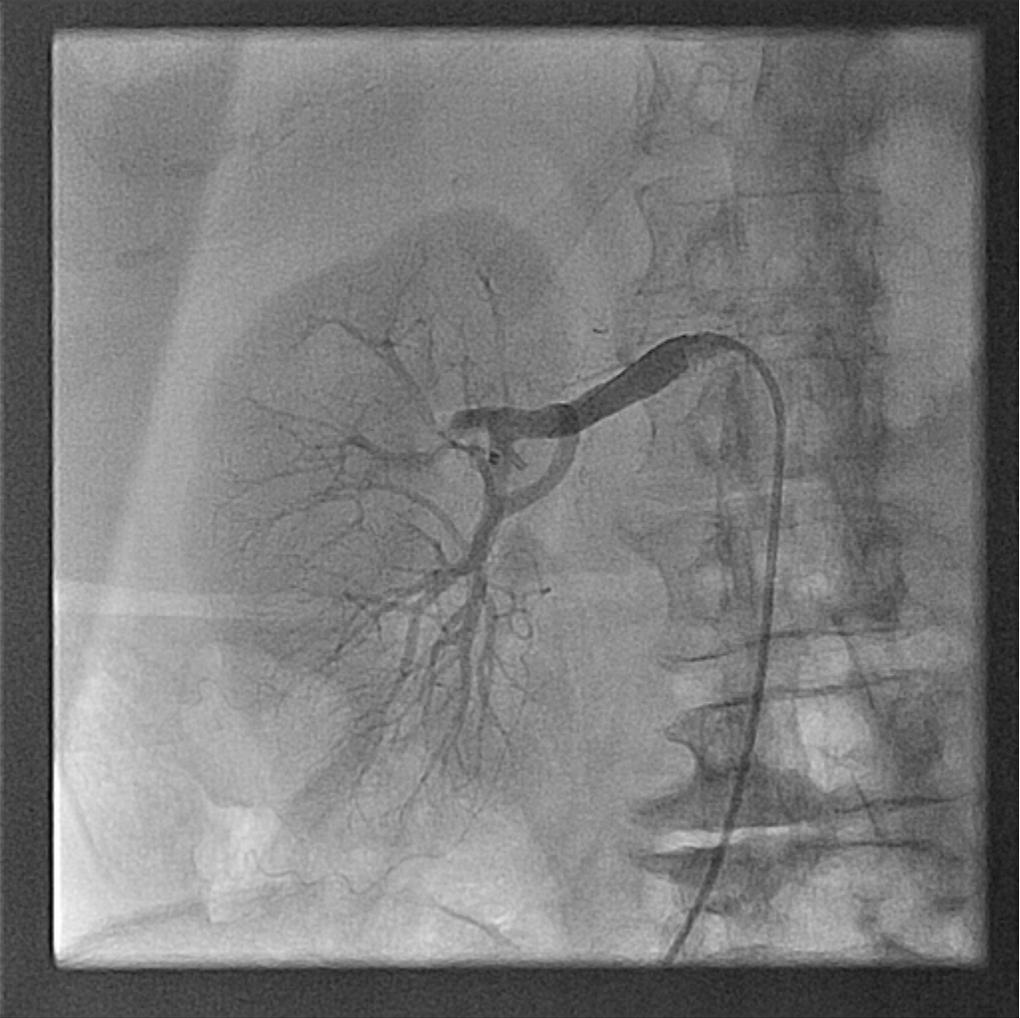
A few pathophysiologic mechanisms have been proposed in the development of FPE in the setting of uni and/or bilateral RAS. Renal ischemia from hemodynamically significant RAS stimulates the sympathetic nervous system and the renin-angiotensin-aldosterone system (RAAS) causing sodium and fluid retention. In context of unilateral disease, this can be countered by pressure natriuresis in the functionally normal kidney. This may explain the marginally lower prevalence of FPE in patients with unilateral RAS when compared to bilateral RAS [4]. In our case, with mild-moderate valvular disease, diastolic dysfunction in the setting of a solitary kidney and RAS, she was unable to maintain a normal volume balance and experience rapid fluid shifts.

Medical management of RAS is preferred given results ASTAL (2009) and CORAL (2014) trial, which showed no statistically significant benefit of revascularization vs. medical therapy [12],[13]. The ACC recommends PCI in patients with hemodynamically significant RAS, recalcitrant congestive heart failure and recurrent pulmonary edema (Class II).

Our case showcases a common presentation of an uncommon disease confounded by pre-existing cardiac disease and that patient-focused care, despite guidelines, can drastically change prognosis. A patient once hospice bound returned to her baseline following renal revascularization.







References

1. Miraie Wardi, Mohamed Teleh, Hasan Salameh et al. Pickering syndrome: a case report and review of literature. *Med Sci case rep*, 2015; 2: 17-20. DOI: 10.12659/MSCR.894208
2. Tolga Cimen, Engin Algul, Tolga Han Efe et al. Flash pulmonary edema: A rare cause and possible mechanisms. *Turkish journal of emergency*. 2017. Volume 17, issue 2, page 65-67. <https://doi.org/10.1016/j.tjem.2016.08.003>
3. H.J. Noh, H.C. Jo, J.H. Yang, *et al.* Flash pulmonary edema in a patient with unilateral renal artery stenosis and bilateral functioning kidneys. *Korean Circ J*, 40 (2010), pp. 42-45
4. Messerli FH, Bangalore S, Makani H et al. Flash pulmonary edema and bilateral renal artery stenosis: the Pickering syndrome. *European Heart Journal*. 2011. Volume 32, Issue 18, Pages 2231-2235. <https://doi.org/10.1093/eurheartj/ehr056>.
5. Saadian RD, Textor SC. Renal-artery stenosis. *N Engl J Med*. 2001 Feb 8; 344(6):431-42.
6. Scoble JE. Renal artery stenosis as a cause of renal impairment: implications for treatment of hypertension and congestive heart failure. *J R Soc Med*. 1999 Oct;92(10):505-10
7. Rundback JH, Murphy TP, Cooper C, Weintraub JL, Chronic renal ischemia: pathophysiologic mechanisms of cardiovascular and renal disease, *J Vasc Interv Radiol*. 2002 Nov; 13(11):1085-92.
8. Kariyanna PT, Jayarangaiah A, Adrah R et al. Pickering syndrome precipitated by angiotensin converting enzyme Inhibitor. *American Journal of Medical Case Reports*. 2016. Vol 4, No. 1, pp 31-34. doi: 10.12691/ajmcr-4-1-9.
9. Vashist A, Heller EN, Brown EJ Jr, Alhaddad IA. Renal artery stenosis: a cardiovascular perspective. *Am Heart J*. 2002 Apr; 143(4):559-64.
10. Hirsch AT, Haskal ZJ, Hertzner NR, Bakal CW, Creager MA, Halperin JL, Hiratzka LF, Murphy WR, Olin JW, Puschett JB, Rosenfield KA, Sacks D, Stanley JC, Taylor LM Jr, White CJ, White J, White RA, Antman EM, Smith SC Jr, Adams CD, Anderson JL, Faxon DP, Fuster V, Gibbons RJ, Halperin JL, Hiratzka LF, Hunt SA, Jacobs AK, Nishimura R, Ornato JP, Page RL, Riegel B; American Association for Vascular Surgery; Society for Vascular Surgery; Society for Cardiovascular Angiography and Interventions; Society for Vascular Medicine and Biology; Society of Interventional Radiology; ACC/AHA Task Force on Practice Guidelines; American Association of Cardiovascular and Pulmonary Rehabilitation; National Heart, Lung, and Blood Institute; Society for Vascular Nursing; Transatlantic Inter-Society Consensus; Vascular Disease Foundation. ACC/AHA 2005 guidelines for the management of patients with peripheral arterial disease (lower extremity, renal, mesenteric, and abdominal aortic): executive summary a collaborative report from the
11. Wierema TK, Yaqoob MM, Renal artery stenosis in chronic renal failure: caution is advised for percutaneous revascularization. *Eur J Intern Med* 2008; 19: 276-279.
12. ASTRAL Investigators, Wheatly K, Ives N, Gray R et al: Revascularization versus medical therapy for renal artery stenosis. *N Engl J Med*, 2009; 361: 1953-62

13. Cooper CJ, Murphy TP, Cutlip DE et al, CORAL Investigators: Stenting and medical therapy for atherosclerosis renal-artery stenosis. *N Engl J Med*, 2014; 370: 13-22