Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 4 (Issue 2) : February 2017 ISSN: 2394-9414
DOI- 10.5281/zenodo.291830 Impact Factor- 3.109

AN INTERESTING CASE OF RAISED RIGHT SIDED PRESSURE – WHAT IS IT?

Dr. Shahid Mahdi*[1], Dr. Raja Suhail[2], Dr. Darpanarayan Hazra[3] & Dr. Sanjeev Gera[4]
*Cardiac Intensive Care, BLK Super specialty Hospital

Keywords:
Pulmonary renal syndrome, small vessel vasculitis, pulmonary embolism, procalcitonin in small vessel vasculitis.

Abstract

We report case of a 47-year-old lady who presented to the emergency room with sudden onset breathlessness for 4 hours. The clinical, laboratory, EKG and 2D Echo were all suggestive of pulmonary embolism and hence was taken up for immediate thrombolysis, however treatment modalities were inverted as she developed headache, epistaxis and worsening of preexisting hematuria. In view of rapidly rising creatinine, worsening breathlessness possible diagnosis made were acute respiratory distress syndrome (ARDS), pulmonary edema (Cardiogenic/ non Cardiogenic) and pulmonary renal syndrome. Finally after narrowing down the diagnosis of renal vasculitis and thoroughly ruling out sepsis/ septic focus, patient was started on IV pulse steroid therapy and immune-modulators. The condition of the patient improved substantially with improvement in clinical and lab parameters.

I. Introduction

Small vessel vasculitis presenting as pulmonary renal syndrome is a clinico- pathological entity and can closely mimic clinical symptoms of pulmonary embolism. The role of thrombolysis in a fluid responsive hypotension patient with clinical features (Well's score), 2D Echo, EKG and laboratory investigations suggestive of pulmonary embolism without radiological evidence can be risky. Moreover serum Procalcitonin can be raised even in small vessel vasculitis. Small vessel vasculitis leading to pulmonary renal syndrome is a serious life threatening condition, which closely mimics pulmonary embolism and needs prompt management. Early use of I/V steroids would be an effective management and hence needs a strong clinical judgment.

II. Case report

A 47 years old normotensive, non-diabetic lady presented to emergency room with complaints of sudden onset shortness of breath for about 4 hours associated with decreased urine output. She had no complaints of chest pain, palpitation,fever or previous history of chronic obstructive pulmonary disease (COPD), hemoptysis, bronchial asthma, similar episode, DVT or any thrombo-embolic events (stroke, MI, amaourosisfugax, Limb ischemia, recurrent pregnancy loss)in other systems in the past. General examination of the patient, revealed an alert and conscious patient with tachycardia (108/min), B.P (140/80 mmHg), tachypnoea (32/min) and hypoxia (oxygen saturation on room air 85%). Chest examination revealed bilateral basal fine crepts. Other systemic examination was unremarkable. Initial blood investigations revealed deranged leukocyte count (N-95,L-4,M-1), TLC – 12.7, Blood Gas analysis (pH:7.30, pCO2:30mmHg, pO2:55mmHg,Lac:1.3mmol/L, Hct:32%, HCO3-: 14.8mmol/L,BE(B): -10.5 MMOL/l, BE ecf :-11.6mmol/L) and rapidly worsening kidney function (Creatinine - 2.97, 4.25, 3.97, 4.7), Troponin I -249.7 and NT-PRO BNP – 10628, D-DIMER- 3.68ug FEU/ml. Urine routine and microexamination revealed active urinary segment, (RBC ++, protein ++).

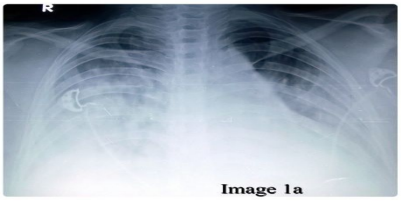


Image 1a

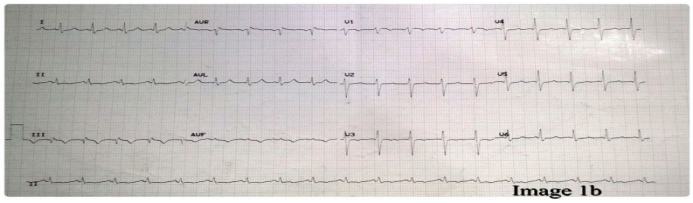
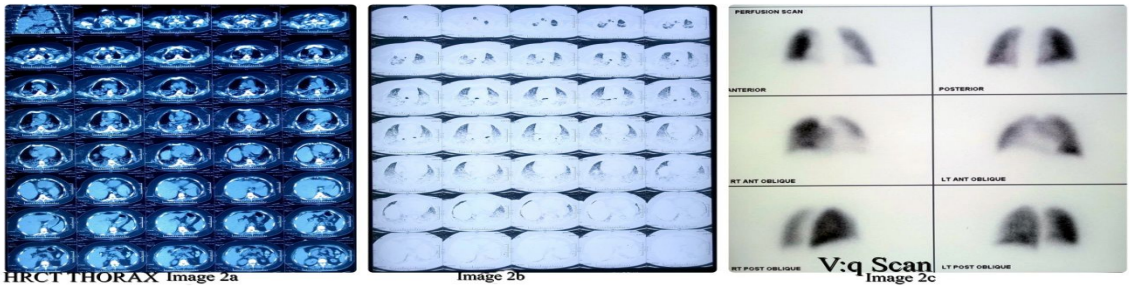


Image 1b

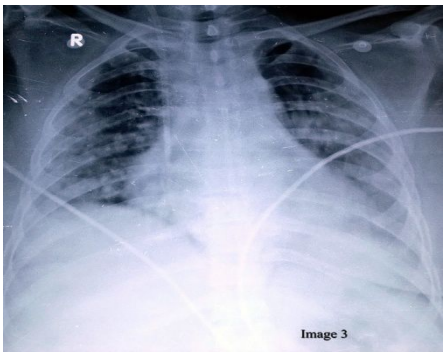
CXR revealed hazy bilateral mid zone, lower zone and CP angle (image 1a). ECG revealed sinus tachycardia with ST-T changes with S1, Q3, T3 pattern (image 1b).

ECHO revealed RA/RV dilatation with PASP 90mmHg, LV global hypokinesia with EF – 55%, no intra-cardiac clot, vegetation, pericardial effusion and IVC collapsing more than 50% associated with mild MR/TR. CTPA was avoided in view of worsening kidney function. Modified Well's score and Clinical Probability of Pulmonary Embolism (PERC) for Clinical Probability of Pulmonary Embolism (5) was calculated and in high suspicion of PE patient was started on systemicthrombolysis(I/V.Inj. Streptokinase 25000IU infusion). However patient developed severe headache, epistaxis and hence thrombolysis was stopped after 15 minutes. NCCT Head ruled out any intra-parenchymableed, epistaxsis and was managed conservatively however hematuria persisted (before and after thrombolysis)**.** There was no improvement in clinical scenario as well as in serial blood investigations. Parameters in context of PE associated withimpending respiratory arrest secondary to respiratory fatigue and increasing trend of creatininepersisted and hence patient was revised for other differentials which were acute onset MODS with worsening of pulmonary, renal and cardiac system, ARDS, pulmonary edema (Cardiogenic/NonCardiogenic), pulmonary renal syndrome with underlying capillaritis associated with underlyingvasculitis.



HRCT (Image 2a, 2b) revealed significant diffuse alveolar opacities associated with ground glass hazeand air bronchogram in bilateral lung right from the apex up to the base with borderlinecardiomegaly and mildly dilated pulmonary artery features suggestive of pulmonary hemorrhage and pulmonary edema. Lung ventilation-perfusion scan (Image 2c) was within normal limit.

Vasculitic – blood work up revealed elevated ESR, ANA (anti nuclear antibody) - weak positive (sample: serum, Dilution: 1.100, Technique: Indirect immune fluorescence assay) and pANCA – positive (sample: serum, Dilution: 1.20, Technique: Indirect immune fluorescence assay). cANCA anti GBM ab, anti phospholipids antibodies (IgG, IgM) , cardiolipinab (IgG,IgM), Lupus anticoagulant C3, C4 levels were normal and hence Wegener's granulomatosis, Good pasteur syndrome, and Antiphospholipid syndrome were ruled out . (4) Serial serum Procalcitonin (PCT) was done whichwas initially normal and then showed an increasing trend. However all blood investigations werenegative in favor of sepsis and therefore we reiterate the fact that elevation of serum PCT level is well documented in cases of inflammation (vasculitis) as well. The patient was started on intravenous pulse Cyclophosphamide 50mg once daily, with I/V Solumedrol 250mg once daily for 3 days and then changed to T. Prednisolone 30mg twice daily. In view anuria and rising creatinine and uremic symptoms she was started on hemodialysis. Serialserum PCTfollowup revealed a decreasing trend. Plasmapheresis was initiated as per protocol (ref) and patient showed remarkable improvement. She started producing urine with improvement in clinical symptoms, hematological, kidney function (creatinine), ESR values and ***radiological imaging (image 3).***



III. Discussion

The triad of hemoptysis, diffuse alveolar infiltrates and low hematocrit, defines pulmonary renal syndrome (1,2), however the presentation can have a wide spectrum and can range from typical to atypical presentations commonly considered as Isolated RPGN, Diffuse alveolar hemorrhage, Pulmonary artery hypertension, Acute Corpulmonale(1).

The underlying cause of pulmonary renal syndrome in usuallysystemic vasculitis, (4) which is predominantly small pulmonary and renal vessels. In the lungs Diffuse alveolar hemorrhage (DAH) is caused by pulmonary cappilaritis and rapidlyprogressiveglomerulo nephritis (RPGN) involves damage of capillaries and basement membrane with leakagesof erythrocytes followed by influx of macrophages, fibrinogen and formation of extra capillary cell proliferation called crescents. In 80% of ANCA associated pulmonary renal syndrome hasdetectable ANCA by ELISA and by immune- fluorescence. In our case the diagnosis of RPGN was made in view of rapid progressive renal failure,proteinuria, hematuria, high ESR, pANCA positivity withnormal compliments and normal anti GBM. A renal biopsy would have been gold standard however our patient and relative did not consent for the procedure. NegativeCTPA and VQ scan (Image 2a, 2b, 2c) ruled out pulmonary arteryhypertension and pulmonary embolism. Modified Well's score and clinical probability of pulmonary embolism (PERC) are internationally accepted forclinical algorithm of pulmonary embolism (5). In our case the calculated score was positive and hence was started on thrombolysis.However in patients with normal hemodynamic (normal BP, HR, mental status, capillary refill, absent mottling, skin temperature, urine output), high PCT values (3) in pulmo-renal syndromes might rather reflectsevere organ damage of lungs and or kidneys than sepsis and hence blunt the initiation of pulsesteroid therapy and redirect our diagnosis towards sepsis, ARDS, (non Cardiogenic edema) MODs (6, 7, 8) if not interpreted in context to the case. Hence timelyadministration of steroid,immuno suppression and plasmapheresis can save the kidney.

IV. Conclusion

Approach to patient OF sudden onset breathlessness should not be limited to ARDS,pulmonary edema, pulmonary embolism, non-Cardiogenic/Cardiogenic shock. Small vesselvasculitis involving renal and pulmonary vessels should also be considered as early diagnosis and prompt management is extremely rewarding in such cases (2). Moreover witha rising number of renal transplants in the world the chances of occurrences and recurrence in transplanted kidneyis not well documented in literatures and therefore needs further study to elucidate the role of ANCA levels in the evaluation and treatment of small vessel vasculitis involving renal and pulmonary vessels. (9,10)

V. Acknowledgements

Dr. Subhash Chandra (Chairman and HOD Cardiology, BLK SS Hospital), Dr. NeerajBhalla (Director and Senior consultant- Cardiology, BLK SS Hospital), Dr. Sunil Prakash (Director and Senior Consultant-Nephrology and Renal transplant).

REFERENCES

1. *Differential Diagnosis of the Pulmonary-Renal Syndrome. Martin Kimmel, Niko Braun and Mark DominikAlscher Department of internal medicine, Division of Nephrology, Robert-Bosch-Hospital, Stuttgart, Germany*
2. *Pulmonary-Renal Vasculitic Disorders: Differential Diagnosis and Management Luis J. Jara, MD*, Olga Vera-Lastra, MD, and Maria C. Calleja, MD*
3. *Diagnostic Value of Procalcitonin in ANCA-Associated Vasculitis (AAV) to Differentiate Between Disease Activity, Infection and Drug Hypersensitivity K. Herrmann*,1,2, S. Schinke2 , E. Csernok2 , F. Moosig2 and J.U. Holle2 1 Division of Rheumatology, Department of Medicine III, University Medical Center Carl Gustav Carus at the TU Dresden, Dresden, Germany 2 Department of Rheumatology, Klinikum Bad Bramstedt, Germany*
4. *Diagnosis and evaluation of vasculitis W. L. Gross, A. Trabandt and E. Reinhold-Keller Department of Clinical Rheumatology, University of Lu¨beck and Clinic of Rheumatology, Oskar Alexander Strasse 26, 24572 Bad Bramstedt, Germany*
5. *Nordenholz KE, Naviaux NW, Stegelmeier K, et al. Pulmonary embolism risk assessment screening tools: the interrater reliability of their criteria. Am J Emerg Med 2007;25:285-90.*

6. *Brunkhorst R., Eberhardt O.K., Haubitz M., Brunkhorst F.M. Procalcitonin for discrimination between activity of systemic autoimmune disease and systemic bacterial infection. Intensive Care Med. 2000;26(Suppl. 2):S199–S201. doi: 10.1007/s001340051144.*
7. *Wu J-Y., Lee S-H., Shen C-J., Hsieh Y.C., Yo P.H., Cheng H.Y., Chan R.C., Lee C.C., Chang S.S. Use of serum procalcitonin to detect bacterial infection in patients with autoimmune diseases: a systematic review and meta-analysis. Arthritis Rheum. 2012; 64(9): 3034–3042. doi: 10.1002/art.34512.*
8. *Joo K., Park W., Lim M-J., Kwon S-R., Yoon J. Serum procalcitonin for differentiating bacterial infection from disease flares in patients with autoimmune diseases. J. Korean Med. Sci. 2011; 26(9): 1147–1151. doi: 10.3346/jkms.2011.26.9.1147.*
9. *Bench-to-bedside review: Pulmonary–renal syndromes – an update for the intensivist Spyros A Papiris, Effrosyni D Manali, Ioannis Kalomenidis,Giorgios E Kapotsis, Anna Karakatsani and Charis Roussos*
10. *Pulmonary-renal syndrome: a life threatening but treatable condition*
Stephen C West1, Nishkantha Arulkumaran1, Philip W Ind2, Charles D Pusey1
Renal Section, Department of Medicine, Imperial College London, Hammersmith Hospital, London, UK
Respiratory Section, Department of Medicine, Imperial College London, Hammersmith Hospital, London, UK

AUTHOR BIBLIOGRAPHY

	Dr. Shahid Mahdi DNB (Respiratory medicine), FRCCM (SINGAPORE), MNCCP, MISCCM. Associate Director and Head Cardiac Intensive Care BLK Super specialty hospital, New Delhi
	Dr. Raja Suhail MBBS, MD (Medicine) Senior resident Cardiac Intensive Care, BLK Super specialty hospital, New Delhi
	Dr. Darpanarayan Hazra MBBS Junior resident Cardiac Intensive Care, BLK Super specialty hospital,New Delhi
	Dr. Sanjeev Gera MBBS, MD (Medicine), DNB (Cardiology) Senior consultant and head of non invasive cardiology, BLK Super specialty hospital, New Delhi