

Kussmaul's Sign- In a patient with Severe PAH

Journal of Clinical Case Reports,
Medical Images and Health
Sciences

Volume 5 Issue 2, 2023

Article Information

Received date: 24/06/2023

Published date: 07/08/2023

***Corresponding author**

*Javaid Ahmad Dar, Assistant Professor in Cardiology, Christian Medical College, Vellore India.

Javaid Ahmad Dar¹, MBBS, MD, DM, Shah Tauseef Arjamand², MBBS, MS

¹Assistant Professor in Cardiology, Christian Medical College, Vellore India.

²Fellow in Surgery, Indraprastha Apollo Hospital, Jasola Vihar, New Delhi, India.

Case Report

The patient is a 33 years old male who had a history of acute pulmonary embolism three and a half years back. He had received thrombolysis with alteplase and was subsequently on oral anticoagulants. After one year of the index episode, the patient started to experience progressive worsening of breathlessness. Further evaluation led to the diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH). Patient was offered pulmonary endarterectomy which he declined and preferred medical management. He did not show clinical improvement and eventually developed refractory heart failure. And patient presented to Emergency department (ED) with worsening of breathlessness. On presentation to the ED, patient was in distress with pulse of 110/min, BP of 100/60 with no evidence of pulsus paradoxus, Respiratory rate of 22/min and chest was revealing bilateral basal crepts and CVS examination revealed pansystolic mummer at left parasternal edge. JVP was elevated and showed only one prominent outward crest, which is a prominent CV wave and one dominant downward trough, a prominent Y descent, and there was a paradoxical rise in the JVP on inspiration (video 1) which is an important clinical sign in heart failure commonly known by the eponym Kussmaul's sign. The prominent CV wave in this patient reflected severe Tricuspid regurgitation (TR). On echo, patient had severe RV dysfunction with severe TR with Pulmonary hypertension. Patient was treated with intravenous diuretics, and pulmonary vasodilators and improved symptomatically and was referred for work up for heart-lung transplantation.

Discussion

Kussmaul's sign is characterized by paradoxical increase in right atrial pressure on inspiration due to decrease in RV compliance as in pericardial diseases like chronic constrictive pericarditis, cardiac tamponade, advanced heart failure and pulmonary hypertension. In this patient's JVP, only prominent upstroke and downstroke is against the constrictive pericarditis which is the most common condition in which Kussmaul's sign is seen. Constrictive Pericarditis has prominent X and Y descends in contrast to only prominent Y descend here. In this patient with a history of previous history of CTEPH, Kussmaul's sign reflects advance disease with severe RV dysfunction. In patients with pulmonary hypertension, Kussmaul's sign is thought to result due to decreased RV compliance, however in a study in patients with severe PAH, Kussmaul's sign was shown to reflect severe pulmonary vascular physiology and correlated independently as a poor prognostic factor.¹ In a meta-analysis in patients presenting with acute myocardial infarction, Kussmaul's sign has been found to be very specific for RV involvement and portends an increased preload requirement with intravenous fluids.² Correctly identifying these clinical signs in a patient presenting to ED, adds in the appropriate management of the patient. This would be most appropriate in patients presenting with inferior wall MI's where Kussmaul's sign identifies a subset of patients with RV involvement who have a much sinister prognosis. And in heart failure

population, Kussmaul's sign is common in patients referred for heart transplantation and is associated with adverse cardiopulmonary hemodynamics.³

Conflicts of interest: None

Funding source: None

Patient consent for the publication: Yes

References

1. Alkhunaizi FA, Harowicz MR, Ireland CG, et al. Kussmaul's Sign in Pulmonary Hypertension Corresponds with Severe Pulmonary Vascular Pathology rather than Right Ventricular Diastolic Dysfunction. *Circ Heart Fail.* 2021;14(1):e007461. doi:10.1161/CIRCHEARTFAILURE.120.007461
2. Dubé E, Crozier M, Middleton A, Best B, Ohle R. Kussmaul's sign for the diagnosis of right ventricular myocardial infarction: a systematic review and meta-analysis of diagnostic test accuracy studies. *CJEM.* 2021;23(2):185-194. doi:10.1007/s43678-020-00012-8
3. Nadir AM, Beadle R, Lim HS. Kussmaul physiology in patients with heart failure. *Circ Heart Fail.* 2014;7(3):440-447. doi:10.1161/CIRCHEARTFAILURE.113.000830

Video: <https://jmedcasereportsimages.org/articles/video-1210.mp4>