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LATE PRESENTATION OF PLATYPNEA-ORTHODEOXIA SYNDROME – A RARE CAUSE OF HYPOXIA AND PULMONARY HYPERTENSION – A CASE REPORT

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Background: Platypnea-orthodeoxia syndrome (POS) is a rare condition of postural hypoxemia and dyspnea which disappears or diminishes in a lying position.
Case report: A 67-year old man was hospitalized due to dyspnea worsening and hoarseness. Several days before admission, he became dyspneic with face and distal parts cyanotic. One year prior to admission, transthoracic echocardiography (TTE) was without visible anatomic pathology with mild pulmonary hypertension (RVSP 40 mmHg). During hospitalization, the patient was dyspneic and cyanotic at rest, worsening in a sitting position. Blood gas analysis on room air revealed partial respiratory insufficiency with orthodeoxia (ABG in lying position: PaO2 34 mmHg, PaCO2 32 mmHg, SaO2 69%; ABG in sitting position: PaO2 28 mmHg, PaCO2 30 mmHg, SaO2 59%). Perfusion scintigraphy, MSCT angiography and thorax HRCT were done to rule out pulmonary embolism, pneumonia or interstitial lung disease. Lung function tests revealed mild to moderate airflow limitation. DLCO was significantly reduced at the level of the alveocapillary membrane. A bronchoscopy found a formation in the epiglottis and cervical lymph nodes biopsy revealed squamous cells laryngeal carcinoma. In the bronchial catheter aspirate Strongyloides stercoralis was isolated. TTE verified severe pulmonary hypertension (RVSP 80 mmHg) and normal systolic function of the right heart, and contrast TTE in lying position found no evidence of shunt. In contrary, contrast TTE in sitting position revealed occurrence of contrast in the left atrium within 3 contractions indicating a presence of shunt. Transesophageal echocardiography (TEE) showed patent foramen ovale (PFO) with an interatrial septal aneurysm, and a test with bubbles proved right-left shunt which was increased in a sitting position. Right heart catheterization was also performed. Initial treatment was with high doses of systemic corticoids with bronchodilators. Albendazole therapy was applied for Stongyloides infection, followed by ivermectin. Radiotherapy of the larynx cancer was planned and the patient was referred for tracheotomy. Few hours after the procedure patient developed respiratory arrest, was reanimated and placed on mechanical ventilation, and after the initial recovery he passed away three days later.
Conclusion: The patient's dyspnea during sitting in bed led to the diagnosis of POS. A careful assessment of the patient's history and the demonstration of a decrease in the arterial O2 saturation and an increase in the right to left shunt through the PFO in sitting position proved by transesophageal echocardiography were essential for the diagnosis.