Case Report

Tumour Emboli And Thromboembolism -Induced Pulmonary Hypertension with Patent Foramen Ovale

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Introduction

Pulmonary hypertension induced by Tumor Emboli is a rare and often under diagnosed cause of severe dyspnea. Tumor emboli can come from various solid tumors, lung carcinoma being among the top ones. Our patient presented with progressive shortness of breath and marked oxygen saturation drop on mild exertion, the cause of which remained ambiguous until death. This emphasizes the importance of considering emboli, both by thrombosis and by tumors as a cause of otherwise undiagnosed and untreatable dyspnea.

Case Progression

A 63 year old man with history of coronary artery disease(with stent placement in 2003), diabetes mellitus, hypertension, hyperlipidemia, asbestos exposure and a 30 pack-year smoking history with cessation 20 years ago was admitted to hospital on 4/4/07. He had progressive dyspnea for the last 6 weeks and had been treated with amoxicillin and then azithromycin with no improvement. He was on outpatient evaluation when he complained of right leg pain and left sided chest discomfort getting worse with inspiration. Doppler examination showed saphenous vein thrombosis below right knee.

On admission, the D-dimer was raised to 6737. ECG findings were interpreted as possible uremia effect and septal fibrosis. Spiral CT on the same date showed a small 1.6x1.1 cm left apical mass lesion along with multifocal; pleural thickenings. He was started on I/V heparin. Pulse oximetry showed a pronounced O2 saturation drop on mild exertion. Transthoracic echocardiogram showed only slight increase in systolic pulmonary artery pressure. Adenosine stress test was done which was negative. The pulmonary function tests did not show any obstructive or restrictive disease but an isolated marked decrease of diffusion capacity was noticed. Bronchoscopy was not of much help.

After about a week, trans-esophageal echocardiogram was done. It showed clear evidence of a patent foramen ovale with a shunt from right to left on bubble study. In addition, the limited trans thoracic echo showed tricuspid regurgitation of 3.8ml/sec translating into systolic pulmonary pressure of 80mmHg(N 15-30). This prompted urgent shift to a university hospital.

On admission to that hospital late on 17/4/07 he was found to have normal vitals with mixed respiratory and metabolic alkalosis. ECG showed same nonspecific changes. His dyspnea was found to be relieved on lying flat (platypnea). Transthoracic echo showed systolic artery pressure as 91mmHg with biventricular hypertrophy. Cardiac catheterization showed diffuse coronary artery disease without high grade stenosis whereas severe pulmonary hypertension was found with transpulmonary gradient of 40mmHg and a probe patent foramen ovale with no evidence of right to left shunt.

The patient had continuous bilateral upper chest ache, which rated 2/10. He was shifted to ICU. His respiratory rate was 29/min with O2 saturation of 96% on 10L of O2 via non-rebreather. A repeat CT of the thorax with contrast, again showed no findings to indicate acute or chronic emboli. Rest were all the old findings along with hilar and thoracic lymph node enlargement. Granulomatous disease was being considered and there was a new left lower lobe opacity.

He remained profoundly hypoxemic and went into refractory hypotensive shock. It was thought to be due to the re-opened foramen ovale. He was
immediately taken to the cardiac catheterization laboratory for emergent closure. However, his condition deteriorated abruptly and he expired on 20/4/07.

Post-mortem examination revealed moderate emphysema. There were three pulmonary lesions that were taken as poorly differentiated adenocarcinoma of the lungs. Nevertheless, the most important finding was extensive tumor emboli and organizing thrombi occluding or nearly occluding the numerous pulmonary arteries in both lungs. In addition, we found multiple thoracic lymph nodes enlarged up to 3.7 cm with metastatic tumor which had moderately enlarged the thyroid gland. There was even a tumor embolus in a small intramyocardial coronary artery.

Discussion

It is shown by different studies and evident by our own case that the symptoms are usually more severe than the physical findings. Dyspnea is the most common finding i.e. in 57% to 100% of the patients.\textsuperscript{1-3} In fact that was the chief complaint of this patient while his chest was clear, abdomen was soft and only finding was significant right calf tenderness. Although considered “classic” we see the symptoms and signs of right heart failure only in 15 to 20%\textsuperscript{3} and our patient was lacking them too. Chest x-ray was clear, Spiral CT was not suggestive for emboli and thus thromboembolism was ruled out.

Spiral CT is considered the most useful imaging study for detecting thromboembolism but is most of the time typically non-diagnostic for tumor emboli. We did the CT three times but found nothing to suggest tumor emboli. Similarly, pulmonary angiogram, which is the gold standard for Thromboemboli has got poor sensitivity and specificity for tumor emboli. The V/Q scan, which is many times non-significant for thromboemboli, is the most promising diagnostic approach for tumor emboli where it can show the symmetric segmental or subsegmental defects in perfusion.

The incidence of pulmonary tumor emboli is between 3% and 26% among patients with solid tumors\textsuperscript{3-5}. Retrospective charts show that only 8% of those with pathological evidence of the tumor emboli actually have been documented for morbidity or mortality\textsuperscript{3,5}

It is unfortunate that most of the times the syndrome shows up only on autopsy. No doubt, treatment is only palliative but the early detection of this syndrome can save a lot of misery of the patient undergoing many different diagnostic and therapeutic procedures and can save a lot of money in terms of both patient and the health care system. The patient should be referred for Hospice care.

In conclusion, this rare and poorly understood entity is under recognized before death. Considering this possibility along with V/Q scan and pulmonary artery aspiration for cytology may be the best method to pick this deadly syndrome before death.

References