

Acute or subacute cor pulmonale: When should we look for malignancies?

Raniero Di Giovambattista

Hospital of Avezzano, Italy

Recently we observed a case of a 51-year-old woman who died in our hospital for respiratory distress related to a widespread invasion of the pulmonary vessel by metastatic cells of a gastric cancer. Autopsy showed an undifferentiated carcinoma of the gastric fundus with diffuse permeation of the pulmonary vascular and lymphatic channels. Acute respiratory failure and severe pulmonary hypertension was the first clinical presentation of the malignancy. Echo 2D examination showed marked dilatation and D-shape right ventricular (RV) deformation. PAP max was 80 mmHg. D-Dimer 1230 mg/dl. Angio-CT scan of the chest permitted us to rule out our first clinical diagnosis of acute pulmonary thromboembolism (PE).

4 months ago a new, similar case came to our attention. A 62-year-old man who suffered in the past of COPD, was admitted to our hospital after a brief clinical observation in another facility, complaining of progressive, severe dyspnea and weakness. He dated the onset of progressive deterioration of his symptoms about 1 month earlier, in absence of fever, chest pain, palpitations or significant changes of the BP. Heart rate at entry was 110/min, BP 105/60 mmHg, D-Dimer value was 3650 mg/dl. Echo 2D showed dilatation and RV D-shape deformation. PAP max measured by echo was about 100 mmHg. Even in this case the chest CT-scan failed to demonstrate pulmonary thromboembolism. On the fifth day of the hospital stay he died of respiratory failure. Autopsy showed a signet ring carcinoma of the stomach with diffuse permeation of the pulmonary vascular and lymphatic channels.

In the last 12 months we observed 2 cases of subacute cor pulmonale as the first clinical presentations of massive, microscopic pulmonary tumor embolism (PTE) arising from gastric cancer. The clinical presentation as acute or subacute RV pressure overload and respiratory distress is a very rare but a fatal complication of cancer, and often a post-mortem diagnosis. When present, adenocarcinoma - more frequently arising from stomach, breast, lung, gallbladder, colon or prostate - is the most common histological tumor type. The tumor involvement of the pulmonary vessels develops from either lymphangitic and/or hematogenous spread. In both cases we observed, as well has been reported in literature, the first clinical diagnosis was pulmonary embolism. D-dimer values were very high. Echocardiographic examination showed signs of acute, severe right ventricular pressure overload and positivity of the Mc Connell sign. This instrumental findings together with the severity of respiratory distress and a shock index > 1 (defined as heart rate divided by systolic blood pressure) observed in both our cases, reinforced in our mind the first clinical suspect of massive P.E. Microscopic PTE and pulmonary thromboembolism are clinically almost indistinguishable, and PTE is often mistaken for thromboembolism. Oxygen desaturation is generally more severe in PTE's patients. D-Dimer values might be as high as they are actually seen in the course of PE. Chest angio CT-scan plays a pivotal role for ruling out the diagnosis of acute pulmonary thromboembolism. Large pulmonary arteries indeed are generally involved in the course of thromboembolic disease: filling defects in the large pulmonary vessels and/or pulmonary infarction are quite always demonstrated by CT-scan. By contrast, with the exception of choriocarcinoma or hepatoma which may provoke acute cor pulmonale by large vein invasion, the tumor emboli usually occlude small vessels and produce subacute cor pulmonale. The parenchyma can be normal or near to normal in these patients. The cases we described remind physicians to consider unknown malignancies as a direct (not thrombus-mediated) cause of acute or subacute cor pulmonale. Chest CT-scan is usually negative in this clinical scenario, so in this case we should look for malignancies. The 2 cases we observed represent in our view also a reminder for physicians and sonographers that echocardiographic examination is a very useful tool to demonstrate pulmonary hypertension and acute right ventricular pressure overload, but not always is able to put light on the etiology of pulmonary hypertension. Take home messages from our experience:

- » Unknown adenocarcinoma may have its first clinical presentation as acute or (more frequently) subacute cor pulmonale. It is a very rare but a fatal complication of cancer, and often a post-mortem diagnosis.
- » Microscopic PTE and pulmonary thromboembolism are clinically almost indistinguishable, and PTE is often mistaken for thromboembolism. AngioCT-scan is the tool of choice for ruling out the diagnosis of pulmonary thromboembolism as a cause of acute or subacute cor pulmonale
- » Echocardiography is quite always useful to evaluate the severity of pulmonary hypertension (PH) and right ventricular overload. Is not the right tool to establish the etiology of PH.

If chest CT-scan fails to demonstrate either the presence of emboli in the large pulmonary vessels as well parenchymal abnormalities which well fit the severity of PH and respiratory failure, we should always look for malignancies.