PRIMARY PULMONARY CHORIOCARCINOMA MIMICKING CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: CASE REPORT

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ABSTRACT

Tumors of the pulmonary artery are often incorrectly diagnosed as chronic thromboembolic pulmonary hypertension (CTEPH) and represent a rare clinical emergency. We report a case of a 46 year-old woman who presented choriocarcinoma of right pulmonary artery (PA) mimicking CTEPH arising five years after a complete hydatidiform mole (HM). The diagnosis was made by pulmonary endarterectomy (PEA), procedure of choice for CTEPH. The patient died soon after complete PEA.

HISTORY AND PHYSICAL

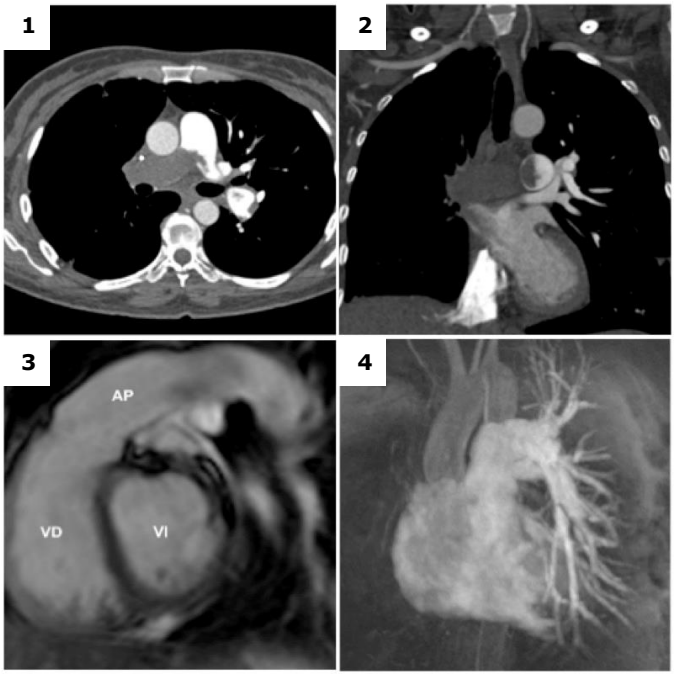
A 46 year-old woman with history of molar pregnancy treated with hysterectomy in 2009. In 2013 she began complaining of oppressive thoracic pain at rest, irradiated to her back, non associated with physical activity which was resolved with NSAIDs. One month later, the patient presented syncope and progressive dyspnea. With a continuing functional class deterioration, she decided to come to our Institute.

The patient was admitted to the hospital on February 4th 2014, and was hospitalized in the cardiopulmonary department. Initial laboratories revealed NT-proBNP 10971pg/ml, high sensitivity CRP: 15.3mg/L, LDH 640.8U/L, TroponinI 0.1ng/ml, Sodium 135mmol/L. Coagulation tests, antibodies for antiphospholipid syndrome, complete blood count, blood chemistry, hepatic function tests, D dimer, C and S protein and antithrombine III were within normal range.

IMAGING

Electrocardiogram showed occasional ventricular extrasystoles, RBBB and RV hypertrophy. Echocardiogram showed mild dilation of RV, free wall of RV hypertrophy, paradoxical septal motion, moderate tricuspid regurgitation and preserved EF. Angiotomography **(Panel A:1-2)** reported right PA occlusion, partial thrombus in left PA with multiple segmentary and subsegmentary defects in upper and inferior lobe.

Angioresonance **(Panel A:3-4)** concludes bilateral pulmonary embolism, predominantly right, RV dilation and biventricular systolic dysfunction (main PA 26mm, RPA 22mm, LPA 24mm). At this time, urine pregnancy test was performed with positive result.
Three days later, Right Heart catheterization revealed: CWP medium 7mmHg. PA: 83/25/45mmHg. RV: 86/2mmHg, RVD2: 8mmHg. RAP: medium 4mmHg, CO(Fick): 2.7L/min, PVR: 1121dyn*s/cm-5. Angiography showed total occlusion of right PA and eft vascular system without perfusion defects.
The management in cardiopulmonary department with inotropic therapy stabilized the patient with a last NT-proBNP 867pg/ml.



Panel A. *1,2.*Contrast-enhanced CT scan, right pulmonary artery occlusion, partial thrombus in left pulmonary artery; 3,4. Angioresonance, bilateral pulmonary embolism.

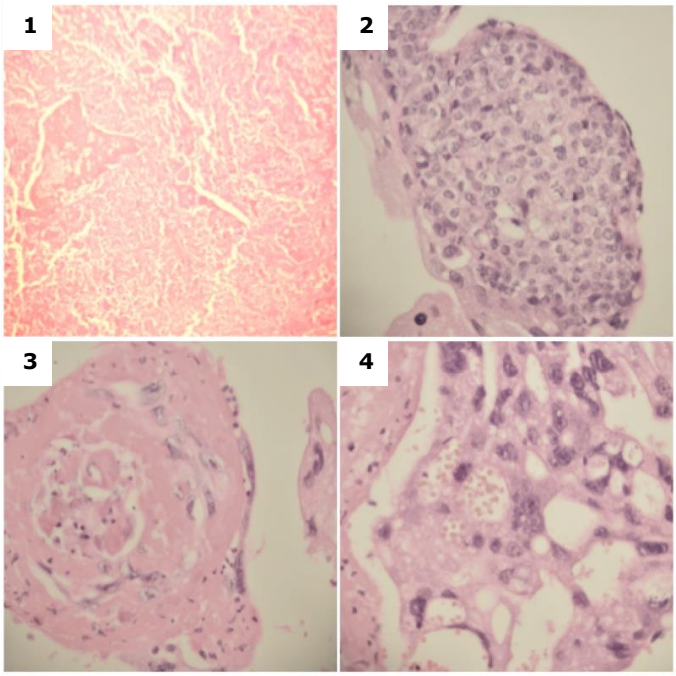
INDICATION FOR INTERVENTION

With a strong suspicion of CTEPH and accessible lesions to the intervention, it was decided to present the patient to PEA.

INTERVENTION

On February 27th 2014 thepatient underwent bilateral PEA with profuse pulmonary hemorrhage complication immediately after reperfusion, hepatization and edema of both lungs, four attempts to retire extracorporeal circulation pump, and pulmonary hypertensive crisis with hypoxemia. The findings were reported as organized thrombi in the left PA, occlusion of 80% of the lumen and total occlusion of right PA. The patient was admitted to the Intensive Care Unit (ICU) for cardiogenic shock.

Despite management with vasopressors, inotropic drugs, nitric oxide and volume therapy thepatient deceased 12 hours later. Post-surgical pathology findings **(Panel B)** reported neoplastic thrombus compatible with choriocarcinoma.



B. Histological images of the PEA specimen 1. Body of lesion, mostly necrotic tissue, HE 10x. 2. 10X Cyto- and syncitiotrphoblast with mitosis and giant multinucleate cells. 3-4. HE 40X Cyto and syncitiotrphoblast with mitosis and giant multinucleate cells.

LEARNING POINTS OF THE PROCEDURE

Choriocarcinoma is a highly malignant gestational trophoblastic disease associated with high β-hCG levels in serum and urine. Choriocarcinoma frequently metastasizes to the lungs, but primary pulmonary choriocarcinoma (PPC) is a very rare entity. PPC of the pulmonary arteries (PA) has been described as an atypical cause of pulmonary thromboembolism (PTE) and pulmonary artery hypertension (PAH). Such cases were reported and some represent a potencially fatal emergency due to cardiopulmonary failure. [1] We report a female patient with PPC of the right PA who was clinically misdiagnosed with PTE and died after embolectomy.

The most important risk factor is a history of HM. A poor prognosis has been established, but based on a systematic review that demonstrated a 1-year survival rate of 61% and with a better outcome in women. Women with history of gestational event less than 7 years had better survival than women without history of gestational a event. [3] Prompt diagnosis with optimal management have higher survival rate. Therefore, genetic examination could help to predict prognosis.

Although, the diagnosis is difficult in the early stages because for the tendency of these tumors to present characteristics of more common diseases such as CTEPH. Typically, serum β-hCG levels in patients with PPC are high, ranging from 1,000 mIU/mL to over 1,000,000 mIU/mL, and urine pregnancy tests are always positive. [2] There is no standardized treatment for PPC. Eetoposide, methotrexate, actinomycin D, cyclophospamide and vincristine has become the preferred regimen for initial treatment. PPC grows rapidly and has high propensity to disseminate to other organs, and due to undifferentiated nature of malignancy, PPC has poor response to radiation therapy.

In conclusion, PPC is now a curable disease and should be considered in the differential diagnosis of fertile women presenting with strong suspicious of CTEPH or PAH. Hence, measuring urine and/or serum B-hCG should be a frontline test to rule out the possibility of a choriocarcinoma. It is important to capture and treat patients in the early stages of the disease and offer surgical treatment combined with chemotherapy, because is the best treatment rendering survival.

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