CLINICAL IMAGE

Postcapillary pulmonary hypertension in the course of a fulminant neoplastic disease

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A previously healthy 36-year-old woman was admitted to our department with an unusually fulminant clinical course of a neoplastic disease and an atypical clinical picture of severe pulmonary hypertension (PH). She was transferred to our department owing to a suspicion of idiopathic PH after exclusion of pulmonary embolism. Seven weeks before hospitalization, she suffered from influenza, followed by severe chickenpox. On admission, her condition was severe. She presented with dyspnea at rest, New York Heart Association class IV, but no chest pain. A physical examination revealed sinus tachycardia of 130 bpm, blood pressure of 100/80 mmHg, normal breath sounds, as well as slightly enlarged left supraclavicular and axillary lymph nodes on both sides. Transthoracic echocardiography (TTE) showed right ventricular dysfunction and dilatation (FIGURE 1A) as well as the echocardiographic signs of PH (calculated right ventricular systolic pressure, 93 mmHg; calculated mean pulmonary artery pressure [mPAP], 47 mmHg; acceleration

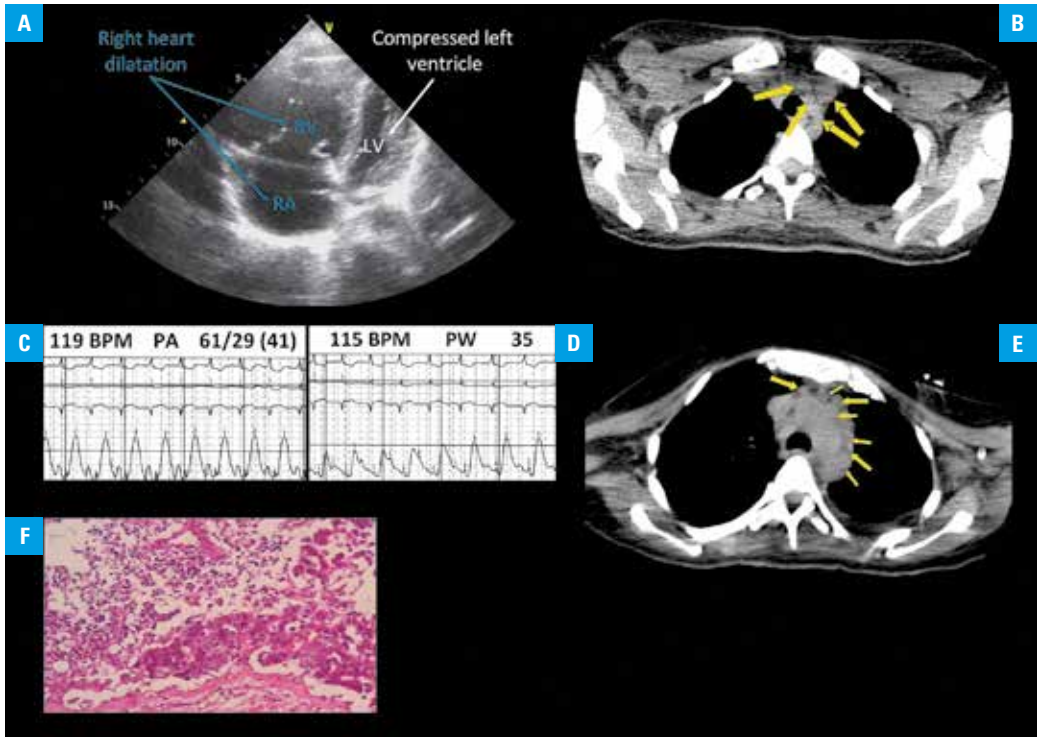

FIGURE 1 **A** – transthoracic echocardiogram, apical 4-chamber view; **B** – computed tomography, enlarged thoracic lymph nodes; **C** – right heart catheterization, pulmonary artery pressure curve; **D** – right heart catheterization, pulmonary wedge pressure curve; **E** – computed tomography, enlarged thoracic, paraaortic, and paracaval lymph nodes (computed tomography performed 2 weeks after the previous one); **F** – histopathological examination, glandular tissue in the metastatic axillary lymph node

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Received: September 3, 2014.
Revision accepted: September 8, 2014.
Published online: September 9, 2014.
Conflict of interest: none declared.
Pol Arch Med Wewn. 2014; 124 (11): 637-638

time, 67 ms; and tricuspid annular plane systolic excursion, 15 mm) and a small amount of fluid in the pericardium; the left ventricle (LV) was compressed with preserved systolic function (LV end-diastolic diameter, 33 mm; LV end-systolic diameter, 16 mm; and LV ejection fraction, 55%) and a normal size of the left atrium. There were no signs of a shunt on TTE. High-resolution computed tomography (CT) revealed scattered packets of mediastinal lymph nodes (**FIGURE 1B**). An abdominal and pelvic CT scan showed enlarged paraaortic and paracaval lymph nodes and an unclear focal liver lesion. Considering lymphadenopathy, we suspected PH coexistent with neoplastic disease or sarcoidosis. Right heart catheterization was performed and revealed postcapillary PH (mPAP, 41 mmHg; pulmonary capillary wedge pressure, 35 mmHg; right atrial pressure, 3 mmHg; and pulmonary vascular resistance, 2.21 Wood units); LV cardiac output was 2.71 l/min and cardiac index, 1.92 l/min/m^2, probably due to LV compression (**FIGURES 1CD**). These findings were surprising because postcapillary PH is believed to be caused by well-known left heart diseases.[1] An acute vasoreactivity test was not performed because of high pulmonary capillary wedge pressure, as it could have provoked fatal pulmonary edema. Based on the laboratory results, infections with cytomegalovirus, Ebstein–Barr virus, and AH1N1 virus were excluded. We also assessed tumor markers with the following results: cancer antigen 125, 45.1 U/ml (normal value, <35); carcinoembryonic antigen, 20.01 ng/ml (normal value, <3.8). A sample of an axillary lymph node was obtained for histopathological examination, which revealed micrometastases of adenocarcinoma of an unknown origin (the gastrointestinal tract or reproductive system). Despite steroid treatment, control TTE and CT revealed progression of the lesions (**FIGURE 1E**), and balloon septostomy was considered. Because the patient's cardiopulmonary condition worsened and histopathology provided unequivocal results (**FIGURE 1F**), she was scheduled for palliative radiotherapy but died of pulseless electrical activity. At the request of the family, a postmortem examination was not performed.

In our patient, the observed abnormalities were most likely caused by the compression of the pulmonary veins due to lymph node enlargement. They could also result from the simultanous presence of a rare type of PH, namely, pulmonary venoocclusive disease. Another potential cause could be an increase in the filling pressure of the compressed LV; however, this is rather unlikely owing to the absence of left atrial enlargement.

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