THE DIAGNOSTIC CHALLENGE BETWEEN CHRONIC PULMONARY EMBOLISM AND PULMONARY ARTERY SARCOMA: A CASE REPORT

E. Franco, D.M. Iancu, E. Cavallone, A. Feneziani, M. Pasquero, S. Ruffini, M. Astegiano, A.M. D'Armini, C. Moretti

We report the case of a 63-year-old obese and hypertensive woman, admitted in hospital with worsening dyspnea for 2 weeks and syncope. A suspected chronic thromboembolism was diagnosed nine months before for a similar clinical picture; hematologic screening excluded thrombophilia and NOACs were prescribed.

Two-dimensional transthoracic echocardiography showed enlargement of right chambers, the interventricular septum shifted to the left ventricle, showing "D" sign. Color Doppler flow imaging showed moderate regurgitation in tricuspid valve, with indirect pulmonary pressure estimation 90 mmHg. Left ventricle was normal for dimensions and function.

ECG evidenced sinus tachycardia and no specific ST-T alterations.

Blood exams evidenced anemia (Hb 9 mg/dl), negative neoplastic markers and absence of occult blood in feces, mildly elevated inflammatory markers, T-Troponin and D-dimer. Naso-pharyngeal swab for SARSCoV2 infection was negative.

Chest computed tomography (CT) with contrast revealed a large filling defect within bilateral main pulmonary arteries. No lymph nodes or pulmonary pneumonia were detected.

Lower extremity venous ultrasound was positive for right popliteal deep vein thrombosis.

The global clinical picture suggested the diagnosis of chronic pulmonary embolism with recent acute event, severe right ventricle compromise, a concomitant deep vein thrombosis. Some factors were not completely clear: chronic anemia, only mildly increased D-dimer, no specific increase of inflammatory markers.

The patient was referred to Cardiac Surgery Unit of Fondazione IRCCS Policlinico San Matteo in Pavia, for pulmonary endarterectomy (PEA).

Unexpectedly the surgical finding was a bilateral pulmonary artery sarcoma, confirmed by histological exam. We thus clarified some uncertain clinical aspects, explainable in athe context of a severe neoplastic picture.

There were no immediate complications, and a chemotherapy was initiated after a period of cardio-pulmonary rehabilitation.

Pulmonary artery intimal sarcoma (PAS) is a very rare disease, its prevalence is about 0.001–0.003%, it can originate from the left and right pulmonary arteries and intimal layer of pulmonary arteries, forming a tumor growing in the nodular cavity or spreading along the intimal surface.

PAS is often misdiagnosed as acute or chronic pulmonary thromboembolism due to its clinical presentation and radiological findings. Thus, early diagnosis is very crucial and may improve patient outcome.