Hepatic Cirrhosis Associated with Arterial Pulmonary Hypertension

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Abstract

The association between hepatic cirrhosis and arterial pulmonary hypertension is mentioned in the literature. The authors report the case of a patient with hepatic cirrhosis, who developed in time an arterial pulmonary hypertension with a fatal outcome. They discuss the pathogenesis of arterial pulmonary hypertension in patients with hepatic diseases, and the therapeutic options in these patients.

Key words

Hepatic cirrhosis - portal hypertension - arterial pulmonary hypertension

Rezumat

Este cunoscută asocierea dintre ciroza hepatică şi hipertensiunea arterială pulmonară. Prezentăm cazul unui pacient in evidența noastră cu ciroză hepatică alcoolică, la care s-a instalat în timp o hipertensiune arterială pulmonară, cu evoluție fatală. Discutăm etiopatogeneza hipertensiunii arteriale pulmonare la pacienți cu boli hepatice cu hipertensiune portală secundară, cât şi unele aspecte terapeutice actuale.

Introduction

The association between hepatic cirrhosis, or portal hypertension and arterial pulmonary hypertension (APH) is quite rare. We report the case of a patient with hepatic cirrhosis, who developed arterial pulmonary hypertension with a fatal outcome.

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Case report

A 58 year old man, a heavy drinker, was known for about four years with hepatic cirrhosis. During all these years he was repeatedly admitted with digestive symptoms, severe asthenia, ascites and peripheral edema. His past medical record revealed a mild essential arterial hypertension, discovered 9 years ago, treated with betablockers and diuretics, and a nonspecific pleural effusion in adolescence. For about 5 months before his last admission he noticed exercise dyspnea, chest pain, cough and in the last month the cough was associated with blood – streaked sputum. Also, the dyspnea became severe in the last month, even with minimal effort (NYHA III). At presentation the patient was in orthopnea, with perioral cyanosis and a high ventricular rate. A systolic murmur of grade III was present at the apex. The initial diagnosis at admission was lung embolism .

The ECG revealed sinus rhythm, a right bundle branch block, with right ventricular hypertrophy, left atrium enlargement, axis right deviation.

The chest X rays in postero-anterior, lateral, RAO and LAO projections showed an enlarged pulmonary artery arch, a dilated right descending pulmonary artery (diameter 22 mm), a high arterio-bronchial index, enlargement of the right heart cavities (Fig.1).

The echocardiography documented a dilated right ventricle (40 mm antero –posterior diameter), paradoxal septum movement, tricuspid regurgitation. Utilizing the tricuspid systolic gradient we calculated a pulmonary systolic arterial pressure of 60 mm Hg, which confirmed the existence of pulmonary arterial hypertension.

The blood oximetry was within normal limits. Limb venous Doppler excluded an embolic APH, and also the chest X rays and spirometry ruled out pulmonary fibrosis, or chronic obstructive lung disease, other causes of secondary pulmonary hyper-tension.

The abdominal ultrasound revealed findings of hepatic cirrhosis, splenomegaly, and moderate ascites, whereas the fibroscopy documented esophageal varices (grade II).

The laboratory analyses showed minimal cytolysis, a reduced prothrombin level (55%), hypergammaglobulinemia, mild anemia. The immunological examinations (antinuclear antibodies, LE cells, rheumatoid factor, cryglobulins) were negative. The HBs antigen and HCV antibodies were also absent.

The final diagnosis was hepatic alcoholic cirrhosis associated with primary arterial pulmonary hypertension. The treatment consisted of silymarin, betablockers, diuretics, iron and folic acid supplementation. The APH was treated with calcium channel inhibitors (nifedipin) and nitrates. After a minimal symptomatic (but not hemodynamic) improvement, the patient left the hospital, being followed–up at home by the family practitioner. He died after 3 months with symptoms of right ventricular and respiratory failure.

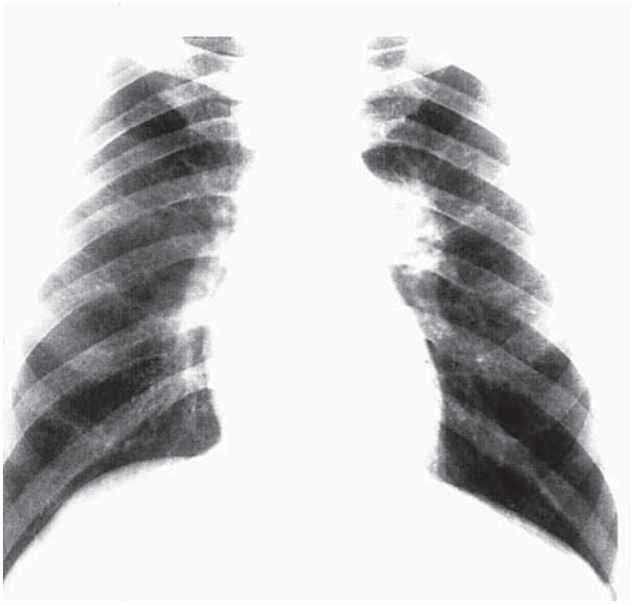

Fig.1 The thoracic radiography reveals an enlarged pulmonary artery arch, dilated right cavities, enlargement of right descending pulmonary artery.

Discussion

The association between hepatic cirrhosis and primary arterial hypertension is quite rare, under 1% (1,2). In a right heart catheterization study with 507 patients with portal hypertension, Hadengue diagnosed the presence of APH in 2% of them. He also found that more than half of the patients had no symptoms of pulmonary hypertension (3). This association is more frequently encountered in females (4,5). The arterial pulmonary hypertension may occur in a patient with cirrhosis or portal hypertension with a variable delay, from months to a few years (2,5). In our case it was 4 years. In many patients the pulmonary hypertension is asymptomatic for a few years.

The occurrence of the APH in our patient was signaled by recent , severe, aggravating dyspnea , chest pains and hemoptysis. The initial diagnosis was lung embolism and for this reason we started an anticoagulant treatment, but the complementary investigations (chest X rays, Doppler examination of veins), ruled out the embolism.

Not only hepatic cirrhosis may be associated with primary arterial hypertension, but also hepatic tumors or portal hypertension secondary to agenesia or thrombosis (4). Different studies found no correlation between the stage of cirrhosis and the degree of pulmonary hypertension (3,6). There is a general agreement that the occurrence of APH is due to the presence of porto –caval shunts. Hadengue found no differences concerning the pulmonary hypertension in patients with surgical porto –caval shunts compared to those with spontaneous shunts. (3). Pulmonary pathological findings in these patients showed a pulmonary plexogenic arteriopathy with micro-thromboemboli.

Noninvasive methods have emerged nowadays to diagnose and follow-up patients with APH. Right heart catheterization is an invasive technique, incurring some risks especially in patients with cirrhosis or portal hypertension (7-9).

Radiological examination uses some indices to quantify the degree of APH (the right descending pulmonary artery diameter, the arterio-bronchial index, the pulmonary artery arch). Cardiac ultrasound not only certifies the diagnosis but also measures very accurately the pulmonary artery pressures (systolic, diastolic, mean). Previous studies have documented a very strong positive correlation between these noninvasive parameters (obtained at echo-cardiography, radiology) and the catheterization data (10-14).

Different hypotheses try to explain the origin of the APH in patients with hepatic diseases and portocaval shunts (2-4). One of them insists on the presence of splanchnic nonmetabolised vasoactive substances, getting directly into the pulmonary circulation, bypassing the liver. Another hypothesis indicates that the pulmonary hypertension is secondary to an overflow generated by the portocaval shunts. The third hypothesis sustains that a recurrent splanchnic embolism arriving directly into the pulmonary circulation (entering the porto - caval shunts), may in time generate the APH.

The patients with hepatic cirrhosis and APH have an bad prognosis, death occurring in months to a few years after diagnosis. Death is due to right cardiac or hepatic failure or complications secondary to portal hypertension (15).

The treatment of AHP consists of calcium channels' blockers, ACE inhibitors, nitrates, other vasodilators, and more recently i.v. prostacycline, nitric oxide or trombo-moduline. In young patients with severe disease, a liver - heart – lung transplantation may save the patient.

In conclusion, the association between hepatic cirrhosis, or portal hypertension and APH is quite rare.

The occurrence of the AHP in patients with cirrhosis and portal hypertension is indicated by recent, severe, aggravating dyspnea, chest pains and hemoptysia. These patients have a bad prognosis, death occurring in months to a few years after diagnosis. Death is due to right cardiac

or hepatic failure or complications secondary to portal hypertension. In patients with severe disease and no contraindications, a liver - heart – lung transplantation may be salvatory.

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