Abstract

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A Rare Case of Porto-Pulmonary Hypertension

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A 55-yr-old female, moderate alcoholic consumption, hospitalized for ascites and declining edema. For laboratory tests: alb. 2,8 mg/dl, AST 77-ALT 128 Ui, pCHE 2970 Ui, slightly increased α-FP and FER., no proteinuria. HBV-HCV absent, negative autoimmunity. Normal chest X-ray, ECG, echo. In echo and CT: ascites, hepatosplenomegaly, portal hypertension. To EGDS congestive gastropathy. It establishes therapy: canrenone 200 mg/day, furosemide 50mg/day, albumin; at discharge canrenone 100mg/day and low-sodium diet. Months of well-being follow, then episodes of decompensation responsive to therapy. After 3 years she returns for worsening wheezing, declining edema, ascitic layer. To A.B.G. no respiratory failure. In echo right cavities dilation, PAPs 110 mmHg. To HR chest and angio CT pulmonary hypertension confirmed with right cardiac catheterization (PAPm 70mmHg), not-responder to vasoreactivity test. HIV negative. Spirometry: no ventilatory deficit. Walking test:<300m. Porto-pulmonary hypertension (PoPH) is diagnosed.

PoPH has prevalence of 2-6% in portal hypertension; female sex appears associated with high risk. It should be suspected in cirrhosis with no obvious cause of dyspnea. Right cardiac catheterization is required for diagnosis. The prognosis for untreated disease is severe. The therapy consists of PDE5-I, ERA, prostanoids. Liver transplantation shows improvement in selected patients, the severe forms represent a contraindication. Our patient is on therapy with sildenafil 60mg/day.