A 40-year-old Patient with Autoimmune Hepatitis and Portopulmonary Hypertension with Miliary Tuberculosis. Challenges of Drug Treatment: A Case Report

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Introduction

Autoimmune hepatitis (AIH) is a rare disease with prevalence of 1.07-1.9 per 100,000 in Europe, affects preferentially females and is characterized by idiopathic hepatitis, increased seric gamma globulins, and circulating non-organ-specific antibodies and the cornerstone of the therapy is immunosuppression. The complications of AIH are the same as in any other chronic liver disease including hepatopulmonary syndrome (HPS) and portopulmonary hypertension (PPH). We discuss a case report of PPH and milliary tuberculosis (TB) and interactions between sildenafil and antituberculosis treatment.

Methods

We review the Clinical History data

Case Report

A 40-year-old woman with AIH, develops cirrhosis, portal hypertension and severe pulmonary arterial hypertension (PAH) and PPH. Treated with Rituximab, diuretics, beta blockers, Sildenafil. Presents with 4 weeks of constitutional symptoms, low-grade fever, nausea, dyspnea, cough, thoracic pain, edemas, with a thoracic x-ray and thorax CT with countless centrolobular micro nodules <2mm in alveolar distribution and an increase in the caliber of the main pulmonary artery (32mm), highly suggestive of milliary TB. Fibrobronchoscopy with bronchoalveolar lavage was performed: GenXpert positive for M. tuberculosis sensitive for rifampin, standard antituberculosis treatment was started. Patient persisted with dyspnea being treated with sildenafil and increased hepatic enzymes, treatment was modified without rifampin, and Moxifloxacin was started. The patient tolerated treatment, clinical, laboratory and radiological improvement, bacteriological control at the first month.

PPH is a pulmonary complication characterized by the presence of liver disease, pulmonary vascular dilatations, and arterial hypoxemia. It occurs in 10-32% of patients with cirrhosis and may lead to the need for liver transplantation, which is the only effective and definitive treatment. Sildenafil, a phosphodiesterase-5 inhibitor is used for idiopathic pulmonary hypertension; however, it should be used carefully in patients with portal hypertension as it may increase portal hypertension by splanchic vasodilation. Sildenafil has a hepatic metabolism by CYP450 3A4, and has potential interactions with rifampin, its concomitant use decreases the plasmatic level and effect of Sildenafil. Moxifloxacin in a non-resistant TB treatment, improves the clinical results, avoids treatment interactions and decreases recurrence TB rates.

In patients with active TB being managed with rifampin and sildenafil, therapeutic drug monitoring must be done, if it is not available, Moxifloxacin can be used. It’s interesting that in murine models of TB, sildenafil reduces tissue pathology and faster bacteriological control, reducing TB treatment time, as evidenced in this case.

Discussion

Figure 1. A. Chest X-ray with extensive micronodular infiltrate of random distribution. B, C, D. Chest CT with countless bilateral diffuse micronodules, less than 2 mm with a centrolobular and perilymph distribution, some of them with greatest size present irregular contour.

Figure 2. Chest X-Ray with important improvement of micronodular infiltrates, magnification of cardiac silhouette and prominent pulmonary arteries are appreciated.

Bibliography


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