BRONCHIOLITS OBLITERANS SYNDROME A MANIFESTATION OF A GRAFT-VERSUS-HOST DISEASE IN RECIPIENTS OF ALLOGENETIC HEMATOPOIETIC STEM CELL TRANSPLANT. CLINICAL CASES FROM A PULMONOLOGIST'S EXPERIENCE

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Abstract

Bronchiolitis obliterans syndrome (BOS) occurs in allogeneic hematopoietic stem cell transplant recipients as a manifestation of a graft-versus-host disease (GVHD-BOS) and is characterized by progressive fibroobliteration of the terminal bronchioles, the clinical manifestations of which include progressive dyspnea on exertion, cough, wheezing, recurrent lower respiratory tract infections, reduced quality of life, and an increased risk of non-recurrent mortality. GVHD-BOS is characterized by an asymptomatic course in the early stages of the disease, with a persistent decline in spirometric parameters showing an obstructive pattern, hyperinflation, and reduced diffusion lung capacity. GVHD-BOS should be considered in every patient following allogeneic hematopoietic stem cell transplantation, with mandatory pulmonary function testing (spirometry and plethysmography with DLCO) on day 100 after transplantation, at the time of initial diagnosis of a GVHD, 1 year after transplantation, and at 6-month intervals during the first 2 years following the initial diagnosis of chronic GVHD. According to the recommendations of the European Society for Blood and Marrow Transplantation, the treatment of GVHD-BOS involves immunosuppressive therapy, management of extrapulmonary GVHD-BOS manifestations, treatment of infections, management of bronchial obstruction with long-acting bronchodilators, and a combination of fluticasone, azithromycin, and montelukast, with pulse steroid therapy. The definitive treatment for GVHD-BOS is lung transplantation, with early placement of the patient on a waiting list for a potential lung transplant in the future during the terminal stage of this disease. Given the diagnostic complexity of this disease, this publication presents two clinical cases of GVHD-BOS, which, based on their clinical course and spirometric "flow-volume" loop findings including hyperinflation and reduced lung diffusion capacity resemble other chronic obstructive pulmonary diseases. Consequently, GVHD-BOS should be differentiated from other chronic obstructive lung diseases, however, the presence of GVHD-BOS does not rule out another diagnosis, which may take the form of a comorbid respiratory condition a fact that hematologists, transplant specialists, and pulmonologists should take into account.

Key words: bronchiolitis obliterans syndrome, graft-versus-host disease, GVHD-BOS, chronic obstructive pulmonary diseases, pulmonary function testing, spirometry, plethysmography, diffusion lung capacity.

Ukr. Pulmonol. J. 2026;34(3):56–61.

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