Combined heart-liver transplantation caused by transthyretin amyloidosis: a case report

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Introduction: Systemic amyloidosis is a diverse set of illnesses characterized by the misfolding and aggregation of over 30 distinct proteins, resulting in the extracellular deposition of amyloid fibrils throughout the body.[1] If amyloid fibrils build up in the heart, it causes cardiac amyloidosis, which leads to progressive cardiac dysfunction. There are two types of cardiac amyloidosis: light chain amyloidosis (AL) and transthyretin amyloidosis (ATTR), which is further subdivided into inherited (ATTRv) and acquired transthyretin amyloidosis (ATTRwt).[2] The clinical presentation is vague, and patients frequently arrive with signs of cardiac insufficiency such as fatigue, orthopnea, and peripheral edema. Today, treatment consists of symptom management, medicines that disrupt TTR's amyloidogenic pathway, and liver and heart transplants. This case study presents a young man who underwent combined heart and liver transplantation caused by transthyretin amyloidosis with severe restrictive cardiomyopathy.

Case report: This is a 48-year-old patient who was diagnosed with amyloidosis in 2019 as part of cardiology treatment for poor exercise tolerance. Following echocardiographic and radiographic evaluation, a bone marrow biopsy was done to rule out amyloid accumulation, and a fatty tissue biopsy confirmed several accumulations. The patient has been admitted to the University Hospital Centre for further treatment, where a genetic test will be performed in June 2020 to confirm a mutation on the transthyretin gene and a diagnosis. With symptoms and signs of heart failure (HF), echocardiography detects significant diastolic and beginning systolic dysfunction, whereas electromyoneurography detects a milder, distal axonal polyneuropathy, for which tafamidis treatment is initiated. Throughout 2020, the patient was observed for stationary findings of HF signs, as well as recurring pleural effusions, for which pleurodesis was performed twice in September 2021, complicating the procedure by the development of liquid pneumothorax. Further disease progression was noted in July 2022, when there was a clinical worsening of the condition in the form of progression of the right-sided pleural effusion and the emergence of ascites, for which paracentesis with albumin replacement was conducted on many occasions. Due to recurrent episodes of acute HF, the patient is registered on the emergency Eurotransplant list on November 8, 2022. On January 4, 2023, a combination heart and liver transplant were conducted, and the patient is being sedated, mechanically ventilated, and hemodynamically stable before being transported to the intensive care unit for cardiac surgery. After 18 hours of mechanical ventilation, the patient is weaned off the ventilator, and noradrenaline is removed from the therapy on the first postoperative day. On the fifth postoperative day, all thoracic and two abdominal drains were withdrawn, and the patient was transported to the Institute for Intensive Cardiac Care, Arrhythmias, and Transplant Cardiology. Because of the increase in transaminases, acute rejection of the liver transplant was suspected, and a biopsy was conducted on January 13, which confirmed mild acute cellular rejection of the liver. The patient follows up with corticosteroid boluses as well as gastroprotection, and the dose of immunosuppressive medication is raised, while the results show a regression of liver enzymes as well as normal graft function. Due to hypoalbuminemia and prolonged abdominal secretion, the patient required parenteral albumin replacement for 18 days, and due to extensive immunosuppression, intravenous immunoglobulins were administered. In addition to the patient's low serum tacrolimus levels despite high doses, a pharmacogenetic test was undertaken, which revealed that the patient is a quick metabolizer of CYP3A4 and CYP3A5 substrate medications, explaining the necessity for larger tacrolimus doses than usual. On the 31st postoperative day, or the 89th day of hospitalization, the patient was discharged home in good general condition, hemodynamically and rhythmologically stable.

Conclusion: Despite the fact that combined heart and liver transplantation is one of the most demanding surgical procedures, the morbidity and mortality rates are low, and the survival rates are encouraging.[3] Good outcomes are the effect of the appropriate indication, prompt diagnosis and treatment initiation, as well as a multidisciplinary strategy that provides optimal patient care.

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LITERATURE

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