PS34

Unilateralno odbacivanje transplantata pluća posredovano protutijelima kod bolesnice s cističnom fibrozom: prikaz slučaja

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KLJUČNE RIJEČI: cistična fibroza; transplantacija pluća; odbacivanje presatka

UVOD: Cistična fibroza je rijetka, nasljedna, multisistemska bolest egzokrinih žlijezda. Posljedica je mutacije CFTR gena (transmembranski regulator provodljivosti za cističnu fibrozu) koja dovodi do stvaranja viskozne i guste sluzi u žlijezdama. Bolest je progresivnog tijeka, a glavni simptomi su kombinacija kronične plućne bolesti i disfunkcije gastrointestinalnog sustava.
PRIKAZ SLUČAJA: 31-godišnjoj bolesnici 2019. godine učinjena je bilateralna transplantacija pluća zbog terminalne faze cistične fibroze. 36 sati nakon transplantacije nastaje reperfuzijski edem desnog plućnog krila, zbog stenoze/presavijanja vaskularne anastomoze, te je učinjena retransplantacija desnog plućnog krila. Daljnji klinički tijek kompliciran se akutnom mitralnom insuficijencijom zbog volumnog opterećenja uslijed resuscitacije plućnog krvotoka. Nakon oporavka liječena je po standardnom imunosupresivnom protokolu, a dvanaest mjeseci nakon transplantacije uveden je mikofenolat mofetil. Krajem 2020. godine dolazi do unilateralnog akutnog odbacivanja desnog plućnog krila posredovanog protutijelima. Nakon reindukcije alemtuzumabom uz podizanje doze imunosupresiva, u ožujku 2021. godine učinjena je kontrolna višeslojna kompjutorizirana tomografija (MSCT) toraksa. Time je potvrđeno značajno pogoršanje statusa desnog plućnog krila uz opsežna područja s infiltratima tipa zrnatog stakla. Nakon trinaest ciklusa imunoadsorpcije uz supstituciju imunoglobulina i pulsne doze kortikosteroida ne dolazi do poboljšanja stanja. Nakon prikaza na konziliju odlučeno je da se napravi desnostrana pulmektomija bez retransplantacije. Nažalost, tijekom operacije dolazi do perforacije plućnih vena i lijevog atrija uz opsežno krvarenje te naposlijetku smrtnog ishoda.
ZAKLJUČAK: Kod dijela bolesnika s cističnom fibrozom nastaju ireverzibilna oštećenja pluća te je transplantacija pluća jedina opcija za produljenje života. Iako ona može spasiti život, kod većine bolesnika dolazi do odbacivanja presatka unatoč imunosupresivnoj terapiji.

Antibody-Mediated Unilateral Lung Transplant Rejection in a Patient with Cystic Fibrosis: Case Report

INTRODUCTION: Cystic fibrosis is a rare, genetic, multisystemic disease that affects exocrine glands. It is caused by a mutation in the CFTR gene (Cystic fibrosis transmembrane conductance regulator), which causes thick, sticky mucus to build up in the glands. The disease is progressive and typically presents with chronic lung disease and gastrointestinal dysfunction.
CASE REPORT: A 31-year-old patient with end-stage cystic fibrosis underwent bilateral lung transplantation in 2019. However, 36 hours post-transplantation, the patient experienced reperfusion edema in the right lung caused by stenosis/folding of the vascular anastomosis. Consequently, retransplantation of the right lung was performed. During the patient's recovery, acute mitral insufficiency occurred due to volume overload during pulmonary circulation resuscitation. After recovery, the patient was treated with standard immunosuppressive therapy, and twelve months post-transplantation, mycophenolate mofetil was introduced. By the end of 2020, the patient experienced unilateral acute antibody-mediated rejection of the right lung. Reinduction with alemtuzumab, along with increased immunosuppressive doses, was performed. In March 2021, a multislice computed tomography (MSCT) revealed significant deterioration of the right lung, characterized by extensive ground-glass infiltrates. Despite thirteen cycles of immunoadsorption with immunoglobulin substitution and pulse doses of corticosteroids, there was no improvement. Following a multidisciplinary consultation, it was decided that the right pneumonectomy without retransplantation should be performed. Unfortunately, during

the surgery, there was a perforation of the pulmonary veins and the left atrium, resulting in extensive bleeding and ultimately, a fatal outcome.
CONCLUSION: For individuals with end-stage lung disease due to cystic fibrosis, lung transplantation may be the only life-saving option. However, even with immunosuppressive therapy, many patients experience graft rejection.

KEYWORDS: cystic fibrosis; lung transplantation; graft rejection