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Brzo progresivni tijek idiopatske plućne arterijske hipertenzije: prikaz slučaja i terapijske opcije

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KLJUČNE RIJEČI: idiopatska plućna arterijska hipertenzija; transplantacija pluća; vazodilatatori

UVOD: Idiopatska plućna arterijska hipertenzija (iPAH) rijetka je bolest koja označava povećanje tlaka u plućnoj cirkulaciji, posljedično srčano popuštanje i, nerijetko, smrtni ishod. Liječenje se temelji na primjeni vazodilatatora.
PRIKAZ SLUČAJA: 41-godišnja pacijentica hospitalizirana je krajem 2020. zbog znakova desnostrane kardijalne dekompenzacije uz prisutnost perikardijalnog izljeva. Postavljena je sumnja na PAH no liječena je amlodipinom i furosemidom. U svibnju 2021. dolazi do pogoršanja srčanog popuštanja i pojave značajne respiratorne insuficijencije (NYHA IV). Obradom u našoj Klinici, potvrđena je iPAH i započeta dvojna terapija (sildenafil i macitentan) uz povećanje diuretske terapije i oksigenoterapiju protokom O2 2L/min. U srpnju 2021. uveden je i treći lijek za iPAH – treprostinil. Uz terapiju se prati poboljšanje funkcijskog statusa (NYHA II/III) uz poboljšanje nalaza 6-minutnog testa hoda (6MWT), NT-proBNP-a i ehokardiografskog nalaza te poboljšanje respiratorne insuficijencije. Nadalje, uz dijetnu prehranu i pojačanu diuretsku terapiju, smršavila je 30 kg (trenutna TT 85 kg). Stabilno je bila do prosinca 2022. kad je evidentirana značajna progresija dilatacije desnog ventrikula i porast NT-proBNP-a. Tada je povišena doza treprostinila, a sildenafil zamijenjen riociguatom. Zbog perzistencije simptoma, u lipnju 2023. započeta je i obrada za transplantaciju pluća. Od dodatnih medikamentoznih opcija, u očekivanju je registracija lijeka (sotatercept) koji predstavlja četvrti, različiti put djelovanja i nova je nada u terapiji bolesnika s iPAH.
ZAKLJUČAK: iPAH je bolest koja često ima progresivan tijek unatoč terapiji i transplantacija pluća tada je jedina preostala opcija. No, s obzirom na relativno kratki medijan preživljenja nakon transplantacije pluća, istu se pokušava odgoditi maksimizacijom medikamentozne terapije.

Rapidly Progressive Course of Idiopathic Pulmonary Arterial Hypertension: A Case Report and Therapeutic Options

INTRODUCTION: Idiopathic pulmonary arterial hypertension (iPAH) is a rare disease characterized by increased pressure in the pulmonary circulation, resulting in heart failure and, often, death. It is treatable with vasodilators.
CASE REPORT: A 41-year-old female patient was hospitalized in late 2020 due to signs of right-sided cardiac decompensation with the presence of pericardial effusion. PAH was suspected, but she was treated with amlodipine and furosemide. In May 2021, the patient's condition deteriorated (NYHA IV). Evaluation at our clinic confirmed iPAH and dual therapy (sildenafil and macitentan) was initiated, along with intensified diuretic therapy and oxygen therapy with a flow rate of 2L/min. In July 2021, a third medication, treprostinil, was introduced. Triple combination therapy improved the patient's functional status (NYHA II/III), 6-minute walk test (6MWT) results, NT-proBNP levels, echocardiography findings, and respiratory insufficiency. In addition, with dietary changes and intensified diuretic therapy, she lost 30kg (current body weight 85kg). The patient remained stable until December 2022, when clinical check-up revealed significant progression of right ventricular dilation and an increase in NT-proBNP levels. At that point, the dose of treprostinil was increased, and sildenafil was substituted with riociguat. In June 2023, the clinic initiated a lung transplant evaluation. As for additional treatment options, the fourth medication (sotatercept) is on the verge of approval and offers new hope for treating iPAH.

CONCLUSION: iPAH often has a progressive course despite therapy, with lung transplantation becoming the only remaining option. However, considering the relatively short post-transplant median survival, efforts are being made to delay it by maximizing medical treatment.
KEYWORDS: Lung Transplantation; Pulmonary Arterial Hypertension; Vasodilator Agents