PS35

Uspješno liječenje bolesnice s kardiogenim šokom uzrokovanim razvojem plućne arterijske hipertenzije u sklopu sistemskog eritematoznog lupusa

Toni Kelava[1]; Zrinka Madunić[1]; Marija Vučić[2]; Mateja Janković Makek[1,3]

1 Medicinski fakultet Sveučilišta u Zagrebu
2 Dom zdravlja Zagreb Zapad
3 Klinika za plućne bolesti Jordanovac, Klinički bolnički centar Zagreb

ID Toni Kelava 0009-0005-8862-0355; Zrinka Madunić 0009-0004-5318-9115; Marija Vučić 0009-0000-0685-5606; Mateja Janković Makek 0000-0002-0171-764X https://doi.org/10.26800/LV-145-supl7-PS35

KLJUČNE RIJEČI: sistemski eritematozni lupus; plućna arterijska hipertenzija; kardiogeni šok

UVOD: Sistemski eritematozni lupus (SLE) je kronična multiorganska autoimunosna bolest. Plućna arterijska hipertenzija (PAH) je rijetka komplikacija SLE i dovodi do opterećenja desne klijetke i popuštanja desnog srca.
PRIKAZ SLUČAJA: Bolesnici je SLE dijagnosticiran 2014., u dobi od 27 godina. Narednih godina je nekoliko puta hospitalizirana zbog plućnog izljeva (u sklopu SLE) zbog čega je u 10/2020. pojačana imunosupresivna terapija. Početkom 2022. javlja zaduhu u naporu te je u 4/2022 u regionalnoj bolnici postavljena sumnja na PAH. Nakon kraće odgode zbog SARS-CoV-2 pozitiviteta, u 5/2022. je hitno hospitalizirana u slici kardiogenog šoka. Učinjenom kateterizacijom desnog srca potvrđena je teška PAH s visokom plućnom vaskularnom rezistencijom (PVR 15WU) i niskim srčanim indeksom (1.28 L/m2). S obzirom na težinu kliničke slike, odlučili smo se za brzo sekvencijsko uvođenje trojne PAH terapije. U intenzivnoj jedinici liječena je s dobutaminom i treprostinilom, a potom je uveden i sildenafil. Uz terapiju je došlo do oporavka srčane i bubrežne funkcije te pada vrijednosti NT-proBNP sa 16193 na 925. U 7/2022. uveden je i treći lijek – bosentan. U daljnjim kontrolama bilježimo poboljšanje funkcijskog statusa i nalaza 6-minutnog testa hoda (6MWT) uz normalizaciju vrijednosti NT-proBNP. Zbog opstruktivnih smetnji u spirometriji i sumnje na udruženu astmu, u terapiju je uvedena i dvojna bronhodilatatorna terapija (LABA + ICS). U daljnjim se kontrolama prati stabilno stanje uz odličan funkcijski kapacitet (NYHA I).
ZAKLJUČAK: Unatoč teškom kliničkom stanju bolesnice, postigli smo odličan učinak uz udarnu trojnu terapiju PAH i nastavak imunosupresivne terapije SLE.

Successful treatment of cardiogenic shock caused by pulmonary arterial hypertension in a patient with systemic lupus erythematosus

INTRODUCTION: Systemic lupus erythematosus (SLE) is a chronic multiorgan autoimmune disease. Pulmonary arterial hypertension (PAH) is a rare complication of SLE and leads to right heart failure.
REPORT: The patient was diagnosed with SLE in 2014, at the age of 27. In the following years, she was treated for pulmonary effusion (as part of SLE). Beginning in 2022, she developed exertional dyspnea and PAH was suspected in the regional hospital. Further work-up in our clinic was delayed due to her SARS-CoV-2 positivity, and she was then urgently hospitalized in cardiogenic shock in May 2022. Right heart catheterization confirmed severe PAH with high pulmonary vascular resistance (PVR 15 WU) and low cardiac index (1.28 L/m2). Considering the seriousness of the clinical picture, we opted for upfront triple PAH therapy. In the intensive care unit, she was treated with dobutamine and treprostinil. Soon after sildenafil was introduced, her heart and kidney function recovered and her NT-proBNP values dropped from 16193 to 925. In 7/2022, bosentan was added to therapy. Her functional status and 6-minute walk test (6MWT) improved on checkups, and NT-proBNP value normalized. Due to an obstructive pattern in spirometry and suspicion of associated asthma, dual bronchodilator therapy (LABA + ICS) was introduced to the therapy. She is now in a stable condition with excellent functional capacity (NYHA I).
CONCLUSION: Despite the severe clinical condition of the patient, we have achieved an excellent effect with upfront triple therapy of PAH and the continuation of immunosuppressive therapy for SLE.

KEYWORDS: pulmonary arterial hypertension; lupus erythematosus, systemic; shock, cardiogenic