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A Chain Journey: From Kidney to Eye

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A case report of a 57-year-old Cuban woman with a longstanding history of uncontrolled arterial hypertension, hyperlipidemia, mild left ventricular diastolic dysfunction and advanced chronic renal failure who presented with mildly decreased visual acuity on both eyes with correction 0,7 (Snellen chart). Slit-lamp biomicroscopy showed quiet anterior segments. Dilated fundus examination revealed bilateral macular serous detachment with generalized yellow gray subretinal deposits with hypopigmented areals, visible on fundus autofluorescence as a "leopard spot-like" pattern. Spectral domain OCT showed striking globular subretinal deposits along the RPE Bruch's membrane complex and subfoveal subretinal fluid. No vascular abnormalities were observed on FA, ICG, and OCT angiography. Electrophysiological testing was within normal limits. Visual field examination showed a central scotoma bilaterally . Eye and orbit ultrasound was within normal limits. Serum laboratory tests showed poor renal function, mild anaemia and thrombocytopenia. Proteinuria was present in the urinary sediment. Immunological tests (ANA, anti-ENA, ANCA) were performed and were negative. IgG antibodies to Toxoplasma gondii, Toxocara sp., HVS1 and HSV2 were detected. Quantiferon test was negative. Serum and urine protein electrophoresis showed significant free monoclonal immunoglobulin κ light chains (LCs). Bone marrow biopsy showed 10% plasma cell infiltration with restriction to κ chains. The kidney biopsy was negative for amyloid, but showed LC deposition disease (LCDD), with advanced chronic lesions already present. The haemato-nephrological specialists suggested an initial progression to disseminated plasmacytoma with advanced renal impairment. Hematological treatment was indicated, treatment was introduced with DARA-VCD scheme, including Daratumumab, Bortezomib, Lenalidomide and Dexamethasone. After 3 weekly applications, we observed a morphological ocular improvement with initial regression of serous macular detachment with stable visual acuity on both eyes with correction 0,7 (Snellen chart) and stable renal function. Continuation of treatment is planned. LCDD is a rare form of monoclonal gammopathy. The patient's ocular course mirrored the severity of her renal dysfunction. Based on the available literature, this is one of few cases, first one in Slovenia, described before of histopathological confirmed deposition of kappa LCs around the glomeruli, associated with bilateral pigment epithelial immunoglobulin LC deposition.

Verižna pot: od ledvic do oči

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Prikaz kliničnega primera 57-letne Kubanke z dolgoletno zgodovino neurejene arterijske hipertenzije, hiperlipidemije, blage diastolične disfunkcije levega prekata in napredovale kronične bolezni ledvic, z zmanjšano vidno ostrino obojestransko s korekcijo 0,7 (po Snellenu). Sprednja biomikroskopija ni prikazala sprememb sprednjega segmenta. Zadnja biomikroskopija je pokazala obojestranski serozni odstop makul z rumeno-sivimi subretinalnimi depoziti obdanimi s hipopigmentiranimi areali, na avtofluorescenci vidnimi kot vzorec geparda.
OCT makul je pokazal hiperreflektivne grčaste depozite vzdolž kompleksa RPE-Bruchove membrane in subfovealni serozni odstop. Na FA, ICG in OCTA ni bilo vidnih žilnih nepravilnosti. Elektrofiziološko testiranje je bilo v mejah normale. Pregled vidnega polja je pokazal centralni skotom obojestransko. Ultrazvok zrkel in orbit je bil v mejah normale. Laboratorijski serumske preiskave so pokazale slabo delovanje ledvic, blago anemijo in trombocitopenijo. V urinskem sedimentu je bila prisotna proteinurija. Opravljeni so bili imunološki testi (ANA, anti-ENA, ANCA), ki so bili negativni. Prisotna so bila IgG protitelesa za Toxoplasma gondii, Toxocara sp., HVS1 in HSV2. Kvantiferonski test je bil negativen.
Elektroforeza beljakovin v serumu in urinu je pokazala povišane proste lahke verige imunoglobulinov κ. Biopsija kostnega mozga je pokazala 10% infiltracijo s plazmatkami z restrikcijo na κ verige. Biopsija ledvic je bila negativna na amiloid, vendar je pokazala bolezen odlaganje lahkih verig (LCDD) z napredovalo ledvično okvaro.
Hematološko-nefrološki konziliji je podal mnenje, da je prišlo do napredovanja v diseminirani plazmocitom z napredovalo ledvično okvaro. Indicirano je bilo hematološko zdravljenje, uvedeno je bilo zdravljenje po shemi DARA-VCD, ki vključuje daratumumab, bortezomib, lenalidomid in deksametazon. Po treh tedenskih aplikacijah smo opazili morfološko očesno

izboljšanje z začetno regresijo seroznega makularnega odstopa, s stabilno ostrino vida obojestransko s korekcijo 0,7 (po Snellenu) in stabilno ledvično funkcijo. Predvideno je nadaljevanje zdravljenja. LCDD je redka oblika monoklonske gamopatije. Očesna simptomatika bolnice je sovpadala z ledvično prizadetostjo. Glede na razpoložljivo literaturo je to eden izmed redkih, prvi v Sloveniji, doslej opisanih primerov histopatološko potrjene bolezni odlaganja lahkih verig κ okoli glomerulov, povezanega z obojestranskim odlaganjem lahkih verig na pigmentni epitel.