Mrežnica, Friday, May 16 2025, 8:00-9:00
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Chairs: Polona Jaki Mekjavić and Ivana Gardašević Topčić

OR-028
Susac syndrome - a case report
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47-years old patient with known billateral sensorineural hearing loss and central nervous system dysfunction (cognitive impairment and emotional disturbance), was seen by an ophthalmologist for the first time due to visual disturbances in the right eye.
The examination showed inferior visual field defect and in the right eye and occlusion of the right superior branch of the central retinal artery with relatively good visual acuity and normal colour vision.
Fluorescein angiography showed large acapillary area in the right eye corresponding to the superior branch retinal artery occlusion and signs of bilateral numerous segmentally occluded arterioles and areas of contrast leakage.
Based on fluorescein angiography and known accompanying systemic signs – encephalopathy and hearing loss, the diagnosis of Susac syndrome was confirmed, along with MRI lesions of the brain.
Neurologists started with aggressive immunosuppressive and antiplatelet therapy.
The patient had numerous fluorescein angiographies performed in the following years with phases of improvement and deterioration of avascular areas and areas of contrast leakage. As a consequence of systemic corticosteroid therapy, cataract and secondary glaucoma developed. She had cataract surgery in both eyes and has intraocular pressure lowering medications, after which the pressure normalized.
After 4 years of follow up, the patient condition is stabile. She is receiving intravenous immunoglobulins every 4 weeks and is regularly followed with MRI of the brain, audiometry and fluorescein angiography, which still shows small acapillary areas and contrast leakage.
The prognosis of the Susac syndrome depends on early and aggressive immunosuppressive treatment. In rare cases, the condition can cause permanent complications like vision and hearing loss.

Susac syndrome is autoimmune microangiopathy affecting the brain, retina and inner ear.
Diagnosis is based on the presence of the clinical triad of central nervous system dysfunction, branch retinal artery occlusions and sensorineural hearing loss. Patients are treated with aggressive immunosuppressive agents and multidisciplinary approach is necessary - a good cooperation between ophthalmology, neurology and ear nose throat specialty.

Susacov sindrom - prikaz kliničnega primera
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47-letna pacientka z znano obojestransko senzorinevralno naglušnostjo in encefalopatijo (težave s kognicijo in motnjami razpoloženja), je bila prvič pregledana pri oftalmologu zaradi poslabšanja vida na desno oko.
Ugotovljen je bil izpad v spodnji polovici desnega vidnega polja in zaporo zgornje veje mrežnične arterije desno, ob relativno dobri vidni ostrini desno in normalnem barvnem vidu.
Fluoresceinska angiografija je pokazala večje akapilarno področje desno po zapori zgornje veje mrežnične arterije in obojestransko številne segmentno okludirane arteriole z barvanjem žilne stene oziroma puščanjem kontrasta.
Glede na fluoresceinsko angiografijo in pridruženimi sistemskimi znaki – encefalopatijo in izgubo sluha, pa je bila potrjena diagnoza Susac sindroma, ki je bila skladna tudi z MRI lezijami v možganovini.
Nevrologi so začeli z intenzivno imunosupresivno in antiagregacijsko terapijo. Pacientka je opravila več kontrolnih fluoresceinskih angiografij s fazami izboljšanja in poslabšanja avaskularnih področij in področij puščanja kontrasta. Ob sistemski kortikosteroidni terapiji je prišlo do katarakte in sekundarnega glavkoma. Potrebna je bila operacija sive mrene obojestransko. Ob lokalni antiglavkomski terapiji, pa je očesni pritisk urejen.
Po 4 letih je pacientkino stanje stabilno, trenutno je zdravljena z vzdrževalnimi odmerki intravenskih imunoglobulinov na 4 tedne, redno se spremlja z MRI glave, audiometrijo in flouresceinskimi angiografijami, ki pa še vedno pokažejo posamezna akapilarna področja in področja puščanja kontrasta.
Za prognozo Susak sindroma je ključno zgodnje in agresivno imunosupresivno zdravljenje, bolezen pa lahko, kljub terapiji, povroči trajne posledice – izguba sluha ali vida.

Susak sindrom je avtoimunska mikroangiopatija, ki se kaže s prizadetostjo možganov, mrežnice in notranjega ušesa. Gre za disfunkcijo centralnega živčnega sistema, zaporo vej mrežničnih arterij in senzorinevralno izgubo sluha. Potrebno je agresivno imunosupresivno zdravljenje, hkrati pa multidisciplinarni pristop več vrst specialnosti - oftalmologija, nevrologija, otorinolaringologija.