Conclusion: The most common ocular manifestation in patients with SNV was episcleritis, which occurred in almost one third of patients. The serum levels of ET-1 were decreased in patients with SNV with eye involvement compared with patients without eye involvement, but this cannot be used for diagnostic purposes.

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FRI0289 THE CYCLOPHOSPHAMIDE-SPARING ROLE OF AN INTENSIFIED B-CELLS DEPLETION PROTOCOL IN ANCA-ASSOCIATED VASCULITIS

Dario Roccatello^{1,2}, Savino Sciascia², Roberta Fenoglio¹, Daniela Rossi². ¹San Giovanni Bosco Hospital, Turin, Italy; ²University of Turin, Turin, Italy

Background: ANCA-associated vasculitis (AAV) are systemic diseases with relapsing chronic course. The management of AAV requires the use of immunosuppressive drugs whose use is associated with potential toxicity.

Objectives: This case-control study aims to evaluate the immunosuppressive-sparing effect of rituximab (RTX) used with cyclophosphamide (CYC), compared to a traditional regimen based on high-dose CYC.

Methods: 26 patients (pts) with AAV with a necrotising extracapillary glomerulonephritis were prospectively enrolled. 13 pts received the intensified protocol of B-lymphocyte depletion therapy (IBCDT) "4+2" with RTX and CYC (4 weekly infusions of RTX followed by 2 monthly followed by prednisone tapered to 5 mg/day in 3 months). 13 pts treated with the high-dose CYC treatment protocol followed by azathioprine as maintenance therapy were enrolled as controls.

Results: In the 13 cases treated with the IBCDT we observed a significant reduction in mean values of parameter of disease activity. After administration of RTX, a significant reduction of the mean s-creatinine values and BVAS was observed. All pts had full B-cell depletion on peripheral blood after the first IBCDT protocol after 1 year. No further maintenance therapy was given. In the cases, a response was observed in 8/13 cases. 4 pts did not respond and a death was observed for cardiovascular causes. No significant difference was observed in terms of response to therapy between the two groups. The IBCDT allows a significant reduction in the cumulative dose of CYC to which each patient was exposed during follow-up, reaching statistical significance levels ($p < 0.001$). Calculated on a monthly basis, the "4+2" protocol allowed an average reduction in the CYC cumulative dose equivalent to 827 mg/month.

Conclusion: In a selected sample of patients with AAV with renal involvement, the IBCDT regimen appeared to be non inferior in terms of the efficacy when compared to CYC-based standard regimens. Moreover, the IBCDT regimen allowed a net reduction in the cumulative average dose of CYC to which pts are exposed, quantifiable in approximately 1g/month.

Disclosure of Interests: None declared

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FRI0290 EFFICACY AND SAFETY OF TOCILIZUMAB IN PATIENTS WITH GIANT CELL ARTERITIS AND VISUAL DISTURBANCES

Andrea Rubbert-Roth¹, Scott Tschuppert², Thomas Neumann¹, Ulf Benecke³, Ian Pirker¹, Johannes von Kempis¹. ¹Rheumatology Clinic, Kantonsspital St Gallen, St Gallen, Switzerland; ²Ophthalmology Clinic, Kantonsspital St Gallen, St Gallen, Switzerland; ³Angiology Clinic, Kantonsspital St Gallen, St Gallen, Switzerland

Background: Giant cell arteritis (GCA) affects cranial arteries and, as a complication, may lead to visual disturbances and ultimately blindness. Tocilizumab (TCZ) has recently been approved for treatment of GCA. Data on its efficacy and safety in patients who present with visual affection are limited.

Objectives: To study the outcome of patients with GCA and visual affection treated with TCZ.

Methods: We performed this retrospective single center study of all patients with GCA and visual disturbances consecutively seen in our clinic between April 2013 and January 2019 who underwent treatment with tocilizumab in addition to glucocorticosteroids (GC).

Results: 16 GCA patients (13 women, 3 male) with a mean age of 76.3 ± 9.7 yrs at GCA diagnosis and 25 affected eyes were treated with tocilizumab in addition to GC.

2/16 patients presented with visual disturbances under treatment with under prednisone, and 1/16 patient under leflunomide for polymyalgia. 2 patients experienced unilateral blindness while receiving iv pulse GC. AAION was diagnosed in 20/25 eyes, PION in 1/25 eyes and occlusion of the central retinal artery in 4/25 eyes. Loss of vision below 10% occurred in 12/25 eyes. None of the patients had bilateral blindness at baseline. 5/25 eyes were affected by hemi- or sectorial anopsia, blurred vision was reported in 8/25 eyes. 14 patients were treated with TCZ iv 8mg/kg every 4 weeks, 2 patients received sc TCZ at 162mg every 2 weeks.

All patients with visual symptoms received intravenous GC boluses, followed by prednisone 1mg/kg/day with subsequent stepwise reduction. Concomitant treatment consisted of low dose ASS in 11/16 and anticoagulants/NOAKs in 5/16 patients. Statins were used in 9/16 patients. Mean disease duration before initiation of tocilizumab was 3.8 ± 5.7 months. 13/16 patients started with TCZ within 2 months after diagnosis of GCA, in 3 patients TCZ was started because of refractory and/or relapsed disease.

Mean duration of TCZ therapy was 14.8 ± 9.4 months. 9/16 patients were able to stop GC after a mean duration of 14.8 ± 9.5 months and have been steroid-free for an average time of 14.7 ± 10 months (as of January 2019). 4/7 patients with a disease duration of less than 6 months are still on a GC taper at present. In addition to cessation of GC, 4 patients have discontinued TCZ and are drug-free at present for 4, 9, 31 and 35 months, respectively. In 2 additional patients, TCZ dose was decreased and/or the dosing interval extended. None of the 12 eyes with vision < 10% have recovered, but no new vision disturbances occurred during TCZ or after cessation of either TCZ/or GC.

Overall, TCZ was well tolerated with no major side effects. 22 patients experienced vascular complications (stroke, development of an aortic aneurysm) during treatment with TCZ.

Conclusion: Although TCZ was unable to reverse unilateral blindness, no new visual symptoms occurred during or after TCZ treatment and the majority of patients were able to stop or reduce GC.

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