Background: Giant Cell Arteritis (GCA) is the most common form of primary systemic vasculitis, mainly affecting adults over 50 years old. Permanent visual loss (PVL) is one of the most feared complications, occurring in about 20% of cases, typically prior to initiation of high-dose glucocorticoid (GC) therapy. Color-duplex sonography (CDS) of temporal arteries (TAs) and large vessels (LVs) is recognized as a first-line diagnostic tool for patients with suspected GCA. A fast-track approach (FTA), incorporating CDS has been associated with a significant reduction of PVL in two retrospective studies.

Objectives: To assess the impact of FTA on PVL and risk of relapses during follow-up compared to conventional care prior to the introduction of the FTA in our rheumatology clinic.

Methods: Patients with new-onset GCA evaluated in our department from January 1998 to September 2019 were included in the study. The FTA approach for GCA was implemented since October 2016. The diagnosis of GCA was based on positive TAs and/or LVs CDS and/or a positive TA biopsy and clinical signs and symptoms of GCA. All patients were clinically examined by the same rheumatologist who performed the CDS. PVL was defined as total visual impairment in one or both eyes. Data on baseline clinical features and later outcomes were collected.

Results: 153 patients were included: 115 females (75.2%), mean age at diagnosis 71.6±8.2 years. Of these, 112 patients (73%) were evaluated conventionally and 41 (27%) with FTA. Patients in the FTA group were older (P=0.0002), presented more frequently with polymyalgia rheumatica symptoms, weight loss, jaw or tongue claudication and scalp tenderness (P<0.05 for all comparisons). The median duration of follow-up in the FTA group was shorter compared with the conventional group (1.5 vs 5.8 years). PVL occurred in 22 (19.6%) patients in the conventional group compared to 5 (12.2%) in the FTA, leading to a reduction of 37.9% in the relative risk of PVL with the FTA approach. Cumulative incidence of relapses and time to first relapse did not change after FTA introduction (P>0.05) (Fig. 1).

Conclusion: The application of a FTA in GCA resulted in a significant reduction of PVL. However, the relapse rate did not seem to be influenced by the FTA, highlighting the need to implement further management strategies, besides earlier diagnosis and prompt initiation of GC, that would impact the course of the disease during long-term follow-up.

Disclosure of Interests: None declared

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ANCA-ASSOCIATED VASCULITIS WITH RENAL INVOLVEMENT: THE ROLE OF A COMBINED HISTOPATHOLOGICAL ASSESSMENT AS PREDICTOR OF PATIENTS' PROGNOSIS

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Background: Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis often affect the kidney and renal involvement has a considerable clinical impact on patient's prognosis. Currently used histopathological classifications are basically focused on the glomerular damage and assessing chronic damage progression, but their prognostic role presents some limitations.

Objectives: To combine the Berden Classification, the ANCA Renal Risk Score (ARRS) and the Mayo Clinic-Renal Chronicity Score (RCS) with the inflammatory interstitial infiltrate and to evaluate the prognostic value of the combined assessment in patients with AAV.

Methods: We included 19 AAV patients with renal involvement (mean age 63±13.2 years; disease duration 4.9±5.2 months) that underwent renal biopsy. Patients were classified according to age, sex, disease duration, ANCA positivity. The histopathological evaluation was performed assessing the Berden category, Risk group (low, medium, high) according to the ARRS and Chronicity class according to the RCS, we also assessed the % of inflammatory interstitial infiltrate. Each patient was follow-up for 12 months; we considered the stage IV (eGFR < 30 ml/min/m²) of the KDIGO CKD Classification as renal outcome.

Results: 8 (42.1%) AAV patients were p-ANCA and 11 (57.9%) c-ANCA. 12 months after renal biopsy, 8 patients (42.1%) had a GFR <30 ml/min. According to the ARRS, 10 (52.6%) patients were in low, 7 (38.6%) in medium and 2 (10.5%) in high risk group. According to the RCS, 2 (10.5%) biopsies had minimal, 10 (52.6%) mild and 7 (38.6%) moderate chronic changes, no one presented severe chronic changes. According to the Berden classification, 6 (31.6%) samples represented the focal, 2 (10.5%) the crescentic and 11 (57.9%) the mixed category, no one represented the sclerotic class. The % of inflammatory infiltrate was 37.4±25.2. The interstitial inflammatory infiltrate showed a direct correlation with the severity of the Berden category (R=0.51, p=0.025), the % of sclerotic glomeruli (R=0.6; p=0.007) and the number of fibrocellular crescents (0.46; p=0.05) and an inverse correlation with the GFR at 12 months (R=-0.48; p=0.045). A ROC curve study identified a 22.5% cut-off of inflammatory infiltrate to predict the outcome of GFR at 12 months <30 ml/min (sensitivity 88%, specificity 97.5%). Patients in focal class developed less frequently a GFR<30 (x2=9.1; p=0.003), but there were no differences in the outcomes between the crescentic and mixed class. ARRS could differentiate risk group with regard to the renal outcome stage IV (β=9.0 p=0.01) as well as the chronicity Score (β=8.1; p=0.017). Finally, we built a matrix combining the different histopathological scores and the % of inflammatory infiltrate to predict the outcome; we found that an inflammatory infiltrate wider than 22.5% characterizes most of patients developing stage IV chronic renal failure at the 12th month. In fact, more than 75% of patients with eGFR < 30 ml/min had inflammatory infiltrate wider than 22.5% despite they were in the low risk class (ARRS) and in minimal changes class (RCS).

Conclusion: Our results underline the importance of the inflammatory infiltrate in renal outcome and histology. Despite the limited number of patients, our data suggest that a combined histological score assessing the chronicity and activity of renal disease from both glomerular and interstitial perspective could better predict patients’ global and renal prognosis.

References:

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PENTOXIFYLLINE GEL FOR ORAL ULCERS IN PATIENTS WITH BEHÇET’S SYNDROME

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Background: Oral ulcers, the hallmark lesion of Behçet’s syndrome (BS) can be disabling and impair eating, drinking and speaking. Despite recent advances in systemic medications for the treatment of oral ulcers, some patients do not achieve complete remission. Topical agents may help such patients by decreasing the frequency and duration of oral ulcers. Pentoxifylline (PTX) is a methyloxanthine derivative that inhibits phosphodiesterase and is thought to have immunomodulatory effects in addition to improving blood flow which is its main reason for use in peripheral vascular disorders.

Objectives: The aim of this study is to assess the efficacy and safety of PTX gel for oral ulcers in patients with BS. We also aimed to explore the best tools for the assessment of treatment response to topical agents in randomized controlled trials (Clinicaltral.gov ID: NCT 03888846).