AB0676

EFFICACY OF RITUXIMAB THERAPY AGAINST ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY-RELATED HYPERTROPHIC PACHYMEINGITIS: A CASE SERIES

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Background: Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis presents with various symptoms. ANCA-associated hypertrophic pachymeningitis (HP) is a very rare pathology.

Objectives: This study aimed to investigate the efficacy of rituximab (RTX) against patients with ANCA-related HP.

Methods: Seven patients were identified by retrospective chart review from local registries at four Hospitals in Japan. All patients met Chapel Hill 2012 Consensus Conference definitions of ANCA-associated vasculitis and were complicated with HP. We assessed the dose of prednisolone, CRP, and MRI findings of HP before and after RTX administration.

Results: Three female and 4 male were evaluated. Median age was 66 years-old. Four cases had HP at the onset of vasculitis. Relapse of HP before RTX administration was found in 2 cases. RTX was used as an initial treatment in one patient. Daily dose of prednisolone and CRP were significantly decreased from baseline levels 24 weeks after RTX treatment. Evaluation of HP by contrast MRI showed improvement in six of seven cases. No relapse after RTX treatment was observed during the follow-up period of 24 weeks. Severe adverse effects were not found in any patients.

Conclusions: Our case series highlight the efficacy of RTX against patients with difficult-to-treat ANCA-related HP. Future studies in this context in a prospective manner are definitely required to establish the B-cell depletion therapy by RTX as a treatment option for ANCA-related HP.

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AB0677

BONE MINERAL DENSITY AND GLUCOCORTICOID TREATMENT IN PATIENTS WITH GIANT CELL ARTERITIS AND/OR POLYMYALGIA RHEUMATICA

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Background: Glucocorticoids (GC) are widely used in treating giant cell arteritis (GCA) and polymyalgia rheumatica (PMR) because of their strong anti-inflammatory and immunomodulatory effects. However, considerable adverse effects like osteoporosis can occur, especially when GC are used as a long-term treatment and disease relapses are frequent. While additional treatment options with conventional immunosuppressive drugs such MTX showed modest effects, the most recent and promising evidence points towards use of biologic agents like the IL-6-receptor antagonist Tocilizumab (TCZ) as an efficient novel treatment option in GCA.

Objectives: Rh-GIOP is an ongoing prospective study monitoring GC induced osteoporosis in patients with inflammatory rheumatic diseases, established in 2015 at the Charité University Hospital (ClinicalTrials.gov Identifier NCT02719314). To date, our database comprises clinical data and bone mineral density (BMD) data measured by dual x-ray absorptiometry (DXA) collected from 592 patients with inflammatory rheumatic diseases. The objective of this cross-sectional analysis was to describe GCA and PMR patients in terms of their clinical characteristics, immunosuppressive therapies and BMD.

Methods: We evaluated data from the initial visit of 61 patients (37 female, 24 male), 43 (70.5%) with PMR, and 18 (29.5%) with GCA (with or without PMR symptoms). Statistical analyses were performed for the overall cohort (GCA +PMR) and separately for the subgroups GCA and PMR. For subgroup analyses non-parametric tests were used.

Results: The whole group showed a mean age of 70.4 years (± 9.0), BMI of 27.7 kg/m² (± 4.8), disease duration of 2.9 years (± 4.0), daily GC dose of 12.5 mg (± 19.6) and cumulative GC dose (CGCD) of 6.1 g (± 6.5). While 40 (93%) PMR patients and all 18 GCA patients received GC on a daily base, GCA patients took significantly higher median doses (8.75 mg, interquartile range (IR) 5/27.5 mg) than PMR patients did (5 mg, IR 3.5/10 mg, $p=0.03$). However, no significant difference was seen in the CGCD. Overall, 34 (55.7%) patients demonstrated osteopenia and 5 (8.2%) patients osteoporosis. 27 (44.3%) patients suffered from at least one fracture, while 10 (16.4%) of these cases were confirmed fragility fractures. Methotrexate (MTX) was the most common conventional immunosuppressive drug being used in 12 (27.9%) PMR and in 6 (33.3%) GCA patients. 2 (11.2%) GCA patients and 1 (1.6%) PMR patient were treated with TCZ.

Conclusions: No increased prevalence of osteoporosis or fractures was found in our patient cohort compared to a normal German population over 50 years.¹ This might be due to therapeutic approaches, which include optimal disease control, the use of lowest possible GC doses and GC-sparing co-medications, and a rigorous osteoporosis prevention and treatment strategy. Alternatively, the number of patients may be still too small to identify significant differences when comparing with a non-diseased population.

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PERIPHERAL ULCERATIVE KERATITIS ASSOCIATED TO AUTOIMMUNE SYSTEMIC DISEASES: VISUAL PROGNOSIS AND OCCURRENCE WHILE SYSTEMIC DISEASE IN REMISSION

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Background: Background: Peripheral ulcerative keratitis (PUK) is a corneal crescent-shaped inflammatory damage with/without concomitant scleritis. PUK can occur isolated or as part of an underlying systemic disease (SD).

Objectives: Objective: To characterise a cohort of PUK patients with a long-term follow-up and to evaluate clinical (ocular and systemic) parameters as predictors of ocular prognosis, in terms of relapses and visual outcomes.

Methods: Methods: Retrospective review (1996–2017) of patients with PUK from a multidisciplinary Uveitis Unit (Ophthalmology/Autoimmune Diseases). Data recorded included ophthalmological features, clinical assessment and laboratory markers at disease onset, and therapeutic interventions, ocular relapses and visual outcomes during follow-up.

Results: Results: Among 18 PUK patients evaluated, 3 were idiopathic, 3 infectious and 12 (67%) associated with systemic diseases (SD-PUK): 8 rheumatoid arthritis (RA), 2 ANCA-vasculitis, 1 SLE and 1 Takayasu's arteritis. Among SD-PUK patients, sex ratio favoured women (rate 9:3) with a median age of 72 (range 33–85) years. Unilateral/bilateral involvement occurred in 7/5 patients and associated scleritis in 50% (11 eyes). All patients presented with eye pain/redness and visual impairment. Four (33%) patients (5 eyes) suffered ocular perforation and required surgery. All patients received topical glucocorticoids (GC), 75% systemic GC, 33% additional immunosuppressant, and 42% biologic therapy. Mean follow-up were 7.3 (range 0.5–12) years. The annual relapse rate was 0.3. Final visual acuity worsened in 42% (8 eyes). In 10 (83%) patients PUK onset occurred previous to SD diagnosis or with SD in remission, and only 3 (25%) were receiving