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EFFICACY OF RITUXIMAB THERAPY AGAINST ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY-RELATED HYPERTROPHIC PACHYMENINGITIS: 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) as a treatment option for 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.

REFERENCES:

Disclosure of Interest: None declared


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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 as 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 2016 at the Charité University Hospital (ClinicalTrials.gov Identifier NCT02719314). To date, our patient cohort compared to a normal German population over 50 years. This study 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.

RESULTS:

Conclusions: No increased prevalence of osteoporosis or fractures was found in our patient cohort compared to a normal German population over 50 years. This study 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: 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 glaucocorticoids (GC), 75% systemic 3C, 33% additional immunosuppressant, and 42% biologic therapy. Median 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