


Background: Behçet’s disease (BD) is a variable vessel vasculitis with a wide and heterogeneous set of signs and symptoms. The inhibitor of phosphodiesterase-4 Apremilast (APR) has demonstrated efficacy in the treatment of oral and/or genital ulcers.

Objectives: To assess the efficacy and safety of APR in BD patients with manifestations different from mucocutaneous ulcers.

Methods: National multicenter retrospective study on 32 BD patients treated with APR at maintained standard dose of 30 mg twice daily.

Results: From a cohort of 49 patients with oral and/or genital ulcers related to BD and refractory to conventional and/or biological treatment, we selected the cases with another clinical manifestation(s) (n=32, 23 women/9 men), mean age of 46.3+5.05 years. None-aphusic manifestations present at aprilestal onset were: arthralgia/arthritis (15), folliculitis/pseudofolliculitis (12), asthma (7), erythema nodosum (3), furunculosis (2), paradoxical psoriasis by TNFi (2), ileitis (2), deep venous thrombosis (2), erythematous and scaly skin lesions (1), fever (1), eating disorder (1), fibromyalgia (1), unilateral anterior uveitis (1) and neurobehçet (1). APR was used in monotherapy (n=3) or combined (n=29) with oral corticosteroids (20), colchicine (17), methotrexate (3), azathiaprine (3), dapsone (1), tocilizumab (1), hydroxychloroquine (1) and/or mesalazine (1).

Table: The outcome of the different clinical symptoms is shown in TABLE. The patient with neurobehçet kept stable (paresthesias) during the 6 months of follow-up. The 2 cases of deep venous thrombosis and the case of anterior uveitis resolved with anticoagulants and adjunct topical treatment, respectively. Furunculosis, folliculitis/pseudofolliculitis and ileitis were the manifestations that improved completely and rapidly. The cases of arthritis experienced improvement, while those with arthromyalgias presented a torpid evolution.

Conclusion: Our data show an improvement of the cutaneous follicular and intestinal clinic with APR and a stability of the neurological clinic, while the musculoskeletal manifestations were mostly refractory.

REFERENCES:
USE OF CONTRAST ENHANCED ULTRASOUND SONOGRAPHY (CEUS) IN LARGE VESSEL VASCULITIS (LVV)

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Background: C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) are important parameters in the monitoring of LVV. Since Tocilizumab is approved for treatment of LVV these cheap and easy repeatable parameters are worthless because of their normalisation by Tocilizumab. MRI and PET-CT as an alternative are not only much more expensive, they are also not arbitrarily repeatable and available. Thus, monitoring of LVV-Patients undergoing a Tocilizumab therapy remains unclear – especially upon showing a persisting thickened vessel wall.

Objectives: CEUS can improve the visibility of tissue perfusion, particularly if there is a very slow bloodflow, which cannot be detected by (power)-doppler sonography.

Methods: In this proof of concept study we investigated patients with active and inactive LVV (aLVV/iLVV) with CEUS. After injection of ultrasound contrast agent we measured the contrasted area of large vessels in a transverse section first if the lumen was completely contrasted and once again 4-8 seconds later. If the vessel wall incorporated the contrast agent the contrasted area increased (Fig 1). The increase of the contrasted area (CA) was correlated with CRP and ESR. Patients were only included if they were not treated with Tocilizumab and therefore ESR and CRP were usable to evaluate the disease activity.

Results: aLVV of 16 patients (11 male; 5 female; 3 patients with LP-7 days apart, 1 OGC prescription within 6 months of the first GCA diagnosis (index date – date of first GCA prescription) followed by a second OGC prescription, no other autoimmune disease requiring leukin-6 therapies, ≥ 1 C-reactive protein (CRP)/erythrocyte sedimentation rate (ESR) lab test and 12 months of data available pre- and post-index.

Potential AEs assessed during the 12 months post-index were descriptively summarized across cohorts of patients based on quartiles (Q) of mean daily dose of OGCs measured over 6 months post-index among this patient sample (Q1: 1.00 to £ 13.75 mg; Q2: > 13.75 to £ 25.00 mg; Q3: > 25.00 to £ 40.00 mg; Q4: > 40.00 mg). Potential AEs included type 2 diabetes (T2D) diagnosis, hemoglobin A1c (HbA1c), blood glucose level, serious infections, cataracts, gastrointestinal bleeding or ulcer, and increases in body mass index (BMI). Actual OGC use by patient could not be confirmed and is a limitation of this study.

Results: Mean age of the 785 eligible patients was 76 years (SD 9); 70% were female. Mean Deyo Charlson Comorbidity Index score at baseline was 1.57 (SD 2.01). The most common baseline comorbid conditions were cerebrovascular disease, diabetes, chronic pulmonary disease, and renal disease. Mean daily OGC dose was 28.9 mg during the first 6 months post-index. Mean (SD) CRP and ESR during the 12-month follow-up was 5.1 (13.6) and 26.5 (20.7), respectively. The proportion of patients with newly diagnosed T2D or with HbA1c ≥ 7.5 during the 12-month follow-up ranged from 7.5% to 24.5% from OGC daily dose Q1 to Q4 cohorts. The proportion of patients with glucose ≥ 200 mg/dL ranged from 7.5% to 15.0% from Q1 to Q4. Serious infections ranged from 16.8% to 24.8% from Q1 to Q4 and cataract ranged from 12.0% to 21.7% from Q1 to Q4. The proportions of patients with gastrointestinal bleed/ulcer ranged from 6.0% in Q1 to 11.8% in Q4. An increase in BMI of 5 ranged from 4.1% to 6.4% from Q1 to Q4.

Conclusion: In patients with GCA, potential OGC-related AEs increased with increased daily OGC dose. This highlights the need for effective therapies that reduce the exposure and potential risk of OGCs.

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