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Belimumab in systemic lupus erythematosus: Experience in clinical practice settings in a regional hospital☆

Belimumab en lupus eritematoso sistémico: experiencia en práctica clínica en un hospital comarcal

Dear Editor,

Belimumab (BLM), a soluble human monoclonal antibody which inhibits the stimulator factor of lymphocyte B (BLyS), is the only biologic drug approved for the treatment of system lupus erythematosus (SLE). It is recommended in patients with active SLE (excluding patients with severe renal compromise or compromise of the central nervous system), with positive antibodies and high grade disease activity despite standard treatment.

We now present the clinical experience of BLM use in a regional hospital servicing a population of 165,000 inhabitants.

Eleven patients with SLE who had received BLM at some time were included. One hundred per cent were female, with a mean age at lupus diagnosis of 31.6 ± 9.7 years. Regarding clinical manifestations that presented during the course of the disease, joints were most commonly affected (100%), followed by cutaneous (81%), haematological (64%), renal (27%), pulmonary (9%) and cardiac (9%) manifestations. One hundred per cent of patients presented positivity for antinuclear antibodies, with 27% being positive for anti-DNA native antibodies, 45% for anti-SSA antibodies and 36% for anti-SSB antibodies. 45% of patients presented with positivity for antiphospholipid antibodies and over one third presented with hypocomplementemia.

Regarding treatments prior to the initiation of BLM, 100% of patients had received antimalarial drugs, over 80% methotrexate and 27% azathioprine. Twenty seven per cent had received anti-TNF drugs, 18% cyclophosphamide and 18% leflunomide. One of the patients had received treatment with tacrolimus and rituximab. The mean age of the patients at treatment initiation with BLM was 38.9 ± 9.6 years. The main manifestations for which treatment was prescribed were joint symptoms followed by cutaneous symptoms. Over 60% of patients underwent an improvement of cutaneous and joint symptoms, with no resolution of lymphopenia being observed in our patients. In 4 of them (37%), treatment was suspended due to ineffectiveness after a median duration of 12.2 ± 7.3 months. Particular mention is of one patient who developed a type IV lupus nephritis during treatment. Treatment was not definitively suspended due to side effects in any cases but was temporarily suspended in one patient (9%) due to a respiratory infection. With regard to concomitant treatments, in 3 of them (27%) treatment with BLM led to reduced doses of concomitant treatment (methotrexate, mycophenolate) and it was not possible to assess the possible corticoid sparing effect given the retrospective nature of the study.

There have been several reports of patient cohorts in U.S.A., Canada and Germany[1–3] treated with BLM with favourable results on the reduction of activity, improvements in lab tests and steroid sparing. However, few data on clinical practice is available in Mexico. The OBSERVE[4] study which included 64 patients with SLE, showed an improvement of ≥20%, ≥50%, ≥80% in 72%, 52% and 27% of cases, respectively. The BIOGEAS[5] study which included 10 patients with SLE refractory to antimalarial drugs and at least one other immunosuppressant, where manifestations of BLM were mucocutaneous, reported a response rate to the drug of 80%, higher than that reported by our study and by the OBSERVE study.

To conclude, in clinical practice BLM has proven to be an alternative therapy to consider in patients with LES with cutaneous manifestations or joints refractory to standard immunosuppressants.

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Livedoid vasculopathy in a patient with bullous pemphigoid and primary Sjögren's syndrome☆

Vasculopatía livedoide en una paciente con penfigoide ampolloso y síndrome de Sjögren primario

Dear Editor,

Livedoid vasculopathy is a chronic, recurrent and painful skin disease that usually affects the lower limbs. It was first described by Bard and Winkelmann in 1967,[1] as a vaso-occlusive disorder affecting the small vessels of the dermis.

We present the case of an 82-year-old woman under dermatological follow-up for bullous pemphigoid, which was stable on low doses of prednisone. Of interest in her personal history was a prior diagnosis of primary Sjögren's syndrome with positive anti-Ro antibodies, meeting the diagnostic criteria set established by Vitali et al.[2]

During a check-up visit, coinciding with more xerophthalmia and xerostomia than usual, reticulated erythematous macules of livedoid appearance together with painful ulcerated nodules and other areas of atrophie blanche were observed on both lower limbs bilaterally and symmetrically (Fig. 1).

Due to a clinical suspicion of livedoid vasculopathy, a diagnostic biopsy was undertaken showing thickening and hyalinisation of the vessel walls, with no inflammatory component.

A complete blood test highlighted: anaemia with a haemoglobin of 10.4 g/dl, positive ANA 1/1280 with speckled cytoplasmic patterns, positive anti-SSA/Ro antibodies (>240.0 U/ml), reduced C3 and C4 (43 and 3 mg/dl, respectively). The remaining parameters showed no anomalies.

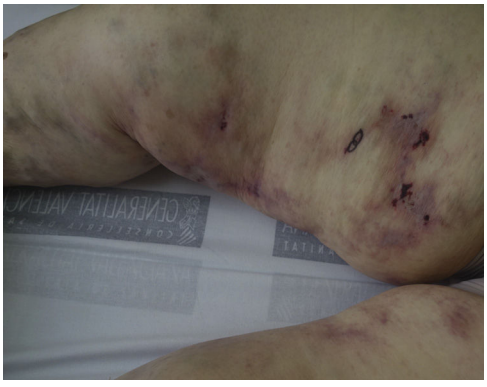

Fig. 1. Clinical image of the skin lesions.

Therefore, given the characteristic clinical skin symptoms and compatible histological findings, the diagnosis of livedoid vasculopathy was confirmed, and possible haematological alterations that would explain a prothrombotic condition were ruled out.

Treatment was started with pentoxifylline 400 mg every 8 h, with 100 mg of aspirin daily, achieving a good response and gradual resolution of the lesions.

Livedoid vasculopathy is a rare, chronic and painful disease, characterised by the presence of macules or purpuric papules and plaques with a tendency to form irregular ulcers that develop into star-shaped atrophic scars and peripheral hyperpigmentation, described as atrophie blanche.[3] It usually affects the lower limbs, with a bilateral, symmetrical distribution. It is characteristically, although not always, associated with livedo reticularis.

It can manifest at any time of life, and is more frequent in young women, with a 3:1 ratio over males.[3] However, our patient was older than the average.

The most frequent histopathological finding is hyalinising vascular changes of the inner layers of the dermal vessels, generally with little inflammation, together with thrombosis inside the blood vessels.[4] These signs enable the diagnosis to be confirmed, and other processes that present with similar skin symptoms to be ruled out.[5]

The condition's aetiopathogenesis remains unknown. However, the presence of thrombophilic alterations is considered increasingly relevant, and complementary tests are needed to rule out prothrombotic conditions.

In turn, it has been related to systemic diseases, such as scleroderma, systemic lupus erythematosus, rheumatoid arthritis, cryoglobulinaemia, and mixed connective tissue disease.[6,7] However, only one case relating to Sjögren's syndrome[8] has been published, and its simultaneous coexistence with two autoimmune conditions has not been recorded to date.[9]

To conclude, we present a case of livedoid vasculopathy in a patient with bullous pemphigoid and primary Sjögren's syndrome with positive anti-Ro antibodies and complement consumption. The co-existence of this disease with both autoimmune conditions reinforces the hypothesis that there is an association with systemic diseases that can be aggressive to the endothelium, triggering the onset of this vaso-occlusive disease. However, studies with a larger number of patients are necessary to obtain definitive results.

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