that they were polymedicated patients taking 6.4 drugs (excluding dicoumarinic anticoagulants). The fact that there was a control group and that the subjects from this group had an average QOL measured according to the EQ-5D test or the Barthel index, that was similar in both groups, rules out the idea that our study sample had a worse QOL. One of the limitations of the study is its small sample size (n = 22), but it represents 100% of the population treated with oral anticoagulants, representing 4.3% of the population, or that 36% of patients are actively working (n = 8).

The patients who participated in this study benefited from the agility and accessibility to the health system of their townships, with a good problem-solving rate, reflected by the patients’ good INR control, without waiting lists and with closer contact. In some cases, a change of treatment could be suggested, but in view of the results, it does not seem to be necessary due to the closely followed attention that patients receive and the awareness of therapeutic compliance generated by monitoring INR figures. Therefore, new oral anticoagulants may coexist with dicoumarinic anticoagulants depending on the clinical and social situation of patients.

References


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Scleromyxedema with neurological symptoms: Successful treatment with immunoglobulins

crossmark

Scleromyxedema asociado a síntomas neurológicos: tratamiento satisfactorio con inmunoglobulinas

de editor

Dear Editor:

Scleromyxedema is a variant of the lichen myxedematous, quite infrequent, which appears as whitish papules or a widespread skin-coloured rash, with 1–3 mm diameter, related to skin thickening. It is frequently associated with extra cutaneous manifestations, out of which paraproteinemia type IgG λ is the most frequent. ¹

There is neurological deterioration in 10–15% of cases, without consensus on the most effective type of intervention for patients who present it. ²

We present the case of a 59-year-old female patient, with a 5-month history of developing erythematous-plaque-type injuries infiltrated in the face, upper third of the torso and upper extremities, with bilateral oedema of the hands (Fig. 1), significant consciousness deterioration, and aphasia and dysarthria 2 days before admission.

During the neurological assessment, the patient was awake, fully space-oriented but partially time-oriented, with bradypsychia and inattention. She followed simple commands, with preserved nomination and repetition, but significant ideomotor apraxia. The cranial pairs assessment did not show any significant alteration. The motor assessment showed symmetric bilateral postural fine tremor, without any additional alterations. Sensitivity was preserved, with no signs of brain deterioration or meningeal signs.

Her admission tests showed thyrotropin 1.24 UI/l (normal range 0.3–4.2), vitamin B levels ⊳ 280 pg/ml (normal range 200–900), normal cerebrospinal fluid (glucose 60 mg/dl, protein 42 mg/dl, leukocytes 3 mm−3 negative for oligoclonal bands, adenosine deaminase>3 U/l, negative cultures, antistreptolysin O 79.9 UI/ml (normal <200), creatinine 0.53 mg/dl (normal 0.5–0.9), C-reactive protein 0.22 mg/dl (normal <0.5), haemogram and plasma electrolytes within normal ranges, negative rheumatoid factor, C3 and C4 complement components within normal ranges, negative antiphospholipid antibodies and anti-B2-glycoprotein 1. The brain MRI, electroencephalogram, and lumbar puncture did not show significant findings.

Scleromyxedema was suggested as a diagnostic hypothesis. A skin biopsy was performed, showing skin with normal orthokeratosis and epidermis. Dermis with fibrosis, mucin reserves, and band superficial dermis proliferation of fibroblasts. Superficial and deep perivascular mild lymphocytic infiltrate. Preserved annex structures. The immunohistochemical test with monoclonal antibodies showed a positive reaction for CD34 and a negative reaction for actin and CD10, confirming the suspected diagnosis of scleromyxedema. An intravenous methylprednisolone pulse treatment was started (500 mg for 2 days). The patient showed progress, with a decrease in skin injuries, but suffered an episode of convulsion, and qualitative consciousness deterioration. The test was completed with electrophoresis and protein immunofixation in blood and urine, showing the presence of the IgG λ mono-
clonal component, with a myelogram showing bone marrow global hyperplasia. In the face of these clinical findings, and given the progressive deterioration of the patient's consciousness, an intravenous immunoglobulin treatment was initiated at 0.5 g/kg/day dose for 4 days, in addition to the oral prednisone at a 1 mg/kg/day dose. The patient showed progress, with a decrease in skin injuries and consciousness deterioration. No side effects associated with the indicated treatment were detected.

Neurological deterioration related to scleromyxedema includes confusion, dizziness, and dysarthria, ascending paralysis, memory loss, acute psychosis, convulsions and coma.3 The triad of high fever, convulsions and coma, with a prodrome of flu-like symptoms, is known as dermato-neuro syndrome. Most cases of scleromyxedema associated with neurological symptoms, such as the case we present here, do not meet the criteria to be considered dermato-neuro syndrome.

There are many treatments for scleromyxedema, including glucocorticoids, retinoid, thalidomide, and extracorporeal photopheresis, phototherapy, cyclosporine, cyclospamide, melfalan, bortezomid, and stem cell autologous transplantation. The treatments that have shown better results in terms of neurological recovery for patients are plasmapheresis, systemic glucocorticoids and immunoglobulins.1–5

In conclusion, according to evidence in the medical literature and to the positive results obtained by means of the treatment administered to our patient, we recommend, as a first-line treatment, the use of intravenous immunoglobulins at a dose of 0.5 g/kg/day, in addition to the use of systemic glucocorticoids, in patients with neurological deterioration associated with scleromyxedema.

References

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** Ictus pontocerebeloso como primera manifestación de un linfoma B difuso de células grandes intravascular de localización renal

Dear Editor:

Intravascular diffuse large B-cell lymphoma (IVDLBCL) is a rare variant of the extranodal diffuse large B-cell lymphoma characterised by a selective growth of lymphomatous cells at the blood vessels lumen area, especially in capillaries, avoiding large vessels.1 This type of lymphoma is mainly diagnosed in adults of approximately 67 years, on average, and is not predominant between genders.1 There are 2 main clinical cases: Western cases, where damages are predominantly neurological and cutaneous, and Asian cases, which present multiple organ failure, pancytopenia, and hemophagocytic syndrome.2 This type of lymphoma has a limited response to chemotherapy, and its prognosis is poor.3,4

We present the case of a woman, 66 years, with no allergy to medications and no toxic habits, who was admitted to the Neurology Department of the Hospital Sant Joan Despí Moisés Broggi hospital (HSJDMB) in October, 2012, with an acute cerebellepontine ischemic infarction, diagnosed by MRI, and who recovered from it ad integrum. During her hospitalisation, she had episodes of fever,