posteriormente leveriracetam y gabapentina en el manejo del status epiléptico en pacientes con PAI[9]. Sin embargo, ningún fármaco debiera ser descartado por completo si su uso resultase esencial. Con frecuencia tras el diagnóstico nos encontramos con fármacos considerados no seguros sin efectos adversos. No obstante, es importante recordar su posible interacción futura con otros fármacos que pudieran precipitar un ataque agudo.

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Successful treatment of systemic capillary leak syndrome with intravenous immunoglobulins

Tratamiento eficaz del síndrome de extravasación capilar sistémica con inmunoglobulinas intravenosas

Sr. Director

Systemic capillary leak syndrome (SCLCS) or Clarkson's Syndrome is a rare and life-threatening disorder characterized by repeated episodes of severe hypotension, hypoalbuminemia and hemoconcentration. Attacks are characterized by marked shifts of plasma and proteins to the extravascular space. Mortality during episodes is high (by persistence of shock or pulmonary oedema to reverse the syndrome, in the recruitment phase). In almost every case reported, a monoclonal immunoglobulin has been present in the serum, but its role in the disease has not been clarified.[1]

We present a patient affected by this rare entity, with a prolonged survival in which the crises have disappeared temporarily after treatment with melphalan–prednisone on two occasions, and lately with intravenous immunoglobulins (IVIG).

A 48-year-old woman was first admitted to hospital in 1997 with severe hypotension, hemoconcentration (hemoglobin 21 g/l) and hypoalbuminemia (23 g/l). The patient was managed in the Intensive Care Unit (ICU) with volume and dopamine; she remained hypotensive for 48 h, oligoanuric and with progressive oedema. She finally recovered her blood pressure with progressive polyuria, disappearance of oedema and normalization of hemoglobin and albumin. A serum monoclonal band of Ig Kappa of 3.5 g/L was documented, with a normal bone marrow examination. She was found to be hypothyroid, with a thyrotropin of 69 ng/dL (normal values: 0.8–1.8), thyroxine of 5.3 pmol/L (normal values: 10.29–23.16) and positive antimicrosomal, antiperoxidase and antithyreoglobulin antibodies.

The patient was diagnosed with SCLCS and treated with therbutaline and aminophylline. During follow-up to year 2000 she presented 20 similar episodes. In 2001, given the lack of therapeutic response, and after a normal new bone marrow biopsy, she was treated with melphalan/prednisone monthly for a year. With this treatment, the frequency and intensity of the episodes diminished until their total disappearance. In 2005 she relapsed with 6 new episodes. Serum levels of chromogranin A were found to be high (188 ng/mL), and did not change during the crises, with a NTF-α that remained normal. An abdominal computerized tomography, nuclear magnetic resonance of the adrenal glands and scintigraphy with octeotride In111 and with Iodine123 metaiodobenzylguanidine were all within normal limits. After a repeat bone marrow biopsy that was unrevealing, she restarted treatment with melphalan/prednisone monthly for a year. The patient remained newly asymptomatic for the next 24 months. The monoclonal band was greatly diminished (0.19 G/) and levels of chromogranin A remained normal (60 ng/mL). Finally, in 2008 the patient presented 2 new episodes of severe shock which reversed promptly with IVIG. Because the patient had a refractory SCLS we finally decided to administer intravenous immunoglobulin (2 g/kg) every 6 weeks. Since the beginning of this treatment, the

patient has had complete resolution of recurrent episodes and has been asymptomatic up to the present.

SCLS are characterized by recurrent crises of massive extravasation of plasma and its constituents to extravascular space as a result of a sudden reversible increase in capillary permeability. About 120 cases have been described, but recurrent episodes of shock were found in only 40 patients.[1] Capillary permeability is normal in quiescent periods. Usually no myeloma has been documented, although in four cases myeloma appeared during follow-up.[2] No curative treatment is available for SCLS, although different regimens have been tried with variable success. The most frequently used is the combination of terbutaline and aminophylline,[1,3] which was unsuccessful in our patient. Interestingly, in patients who developed myeloma during their evolution, treatment with chemotherapy is accompanied by a disappearance or decrease of the attacks.[3] Based on these results some authors have proposed the use of chemotherapy in patients with repeated attacks.[3] However to the best of our knowledge, the patient reported here is the first in whom this regimen has proved effective, although only temporarily. More recently, Lambert et al. described a patient with scleroderma and chronic refractory SCLS in whom the monthly administration of IGIV was followed by disappearance of symptoms, and two other patients with SLCS were also controlled with the same regimen during the acute phase of the disease.[4] At least one other patient with repeated episodes of SCLCS has remained stable for three years with chronic administration of IGIV.[5] More recently Gousseff et al.[6] had reported a similar data in other 18 patients recruited in a European Register. In conclusion, our observation suggests for the first time that there is a beneficial effect from therapy with melphalan–prednisone, which is reversible with the finalization of treatment, and strengthens the hypothesis of the action of intravenous immunoglobulins.

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Síndrome de reconstitución inmune en un caso de tuberculosis diseminada, tras retirada del tratamiento con adalimumab

Immune reconstitution syndrome in a case of disseminated tuberculosis after withdrawal of treatment with adalimumab

Sr. Director:

Una de las complicaciones más graves de los tratamientos biológicos con agentes antifactor de necrosis tumoral-alfa (anti-TNFα) es el desarrollo de una tuberculosis (TBC)[1]. Por otra parte, en relación con el inicio del tratamiento antirretroviral de la infección por el virus de la inmunodeficiencia humana (VIH) se ha descrito un síndrome de reconstitución inmune (SRI), especialmente frecuente en la infección tuberculosa, que se caracteriza por un empeoramiento paradójico del estado general, con aparición de fiebre, anorexia, adenomegalias y aumento de los reactantes de fase aguda. Este SRI se ha descrito en otras alteraciones del sistema inmune como el tratamiento con interferón de la esclerosis múltiple o tras la retirada de un tratamiento anti-TNF en la artritis reumatoide[2–4]. Presentamos un caso de tuberculosis miliar secundario al empleo de adalimumab, un anti-TNFα, utilizado en el tratamiento de la enfermedad de Crohn y, tras la retirada del adalimumab, de un SRI, consecuencia poco conocida derivada de su supresión.

Varón de 38 años afecto de enfermedad de Crohn diagnosticada a los 18 años de edad. Desarrolló estenosis intestinales a pesar del tratamiento con corticoides y azatioprina, motivo por el que se inició tratamiento con adalimumab. El Mantoux era negativo y la radiografía de tórax normal. Tras 10 meses de tratamiento, el paciente comenzó con astenia, anorexia, fiebre de hasta 39 °C, sudoración profusa nocturna y pérdida de peso. Por dicho motivo se suspendió el adalimumab y se administraron antibióticos de amplio espectro sin mejoría. Dos meses después se añadió disnea progresiva y fue hospitalizado. A la exploración destacaba la postración del enfermo, la intensa desnutrición, fiebre de 38,5 °C, y a la auscultación crepitantes inspira-