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Schnitzler Syndrome☆

Síndrome de Schnitzler

To the Editor:

Schnitzler syndrome, described in 1974, is an autoimmune chronic urticaria syndrome associated with a characteristic monoclonal IgM component, in addition to fever, joint pain and lymphadenopathy.[1]

Several authors have reported patients with urticaria, fever, joint pain and increased erythrocyte sedimentation rate (ESR) and an IgG monoclonal component, suggesting that this could be a variant of Schnitzler syndrome. Clinical manifestations do not appear to differ between the typical disease and its variants. The diagnostic criteria can therefore be extended to include the variant IgG.[2–4] We report the case of a 38-year-old woman first seen in 2001, who presented a persistent IgG monoclonal component and maculo-papular erythematous lesions, non-painful or itchy, at trunk level (Fig. 1) and upper limbs, as well as perforation of the nasal septum. During her progression, she developed diarrhea accompanied by abdominal pain, paresthesias, hepatosplenomegaly, livedo reticularis, bone pain and rapidly progressive sensorineural hearing loss. Laboratory tests showed a non-regenerative anemia, ESR 100 mm/h and a gamma monoclonal IgG band 3580 mg/dl (vn: 600–1650), Bence-Jones proteinuria (−), ANA-HEp2 (−), normal complement and normal anti neutrophil cytoplasmic antibodies (ANCA) cytoplasmic (cANCA) and perinuclear pattern (pANCA), TSH, T3, T4 and thyroid antibodies, with a negative Hansen test. Abdominal subcutaneous fat biopsy: congo red negative. X-rays of the skull, pelvis, dorso-lumbar spine, chest, mento-naso and fronto-naso were normal, a normal tomography of the abdomen with contrast, and an electromyography of the 4 extremities showing an axonal, asymmetric and distal neuropathy. Skin biopsy observed necrotizing leukocyte vasculitis. Given the different hematological findings, a new bone marrow aspirate was performed with a negative cytogenetic study for lymphoproliferative diseases. All other causes of monoclonal gammopathy (collagen disease, amyloidosis, Hansen, POEMS and neoplasms) were ruled out leading to the diagnosis of Schnitzler's syndrome (Lipsker criteria), diagnosed in 2009 (Table 1).[5] This syndrome can be mimicked by other diseases such as cryoglobulinemia, urticarial hypocomplementemic vasculitis, acquired C1 inhibitor deficiency, hyper-IgD syndrome and adult Still's disease. In 2010, the patient presented anemia, increased plasma cells in the bone marrow (25%), increased IgG and positive Bence-Jones proteinuria again, with flow cytometry showing a heterogeneous group of plasma cells which expressed an intense CD 138 CD 38+ (+) CD19 (−), CD 56 (+) (±4.20% of total cells) immunophenotype, which corresponded to atypical plasma cells, leading to the diagnosis of multiple myeloma. We must consider, in the differential diagnosis, other entities characterized by monoclonal gammopathy and chronic urticaria, namely amyloidosis, chronic auto-inflammatory syndromes: Muckle–Wells or Sweet syndrome and neoplasms. Other symptoms that may be present include hearing loss, chronic inflammatory demyelinating polyneuropathy, headache, depression, dizziness, peripheral neuropathy associated with anti-AMG (anti-myelin associated glycoprotein), thrombophilia, antiphospholipid syndrome and hyperhomocysteinemia. The overall prognosis of Schnitzler syndrome depends on the possible progression to a lymphoproliferative disorder (15%–20%), either lymphomas, including lymphoplasmacytic lymphoma, Richter type lymphoma, marginal zone lymphoma, myeloma or Waldenstrom's disease. The latter may occur 10–20 years after the onset of symptoms. The patient underwent chemotherapy without an adequate response. She is currently awaiting a bone marrow transplant.

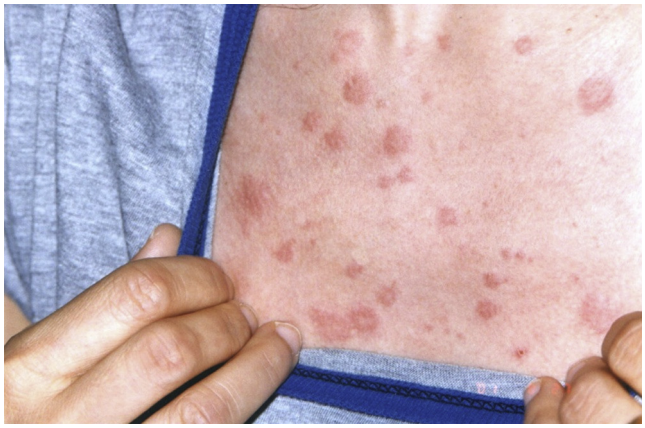

Fig. 1. Erythematous maculopapular lesions, non-painful or itchy, at trunk level.

Table 1
Criteria for the Diagnosis Schnitzler Syndrome.

Major criteria (both are required): chronic urticarial dermal rash and monoclonal gammopathy (IgM or IgG)	Minor criteria (at least 2): intermittent fever, joint pain or arthritis, bone pain, palpable lymphadenopathy, splenomegaly or hepatomegaly, elevated ESR, leukocytosis and bone abnormalities (X-ray or histological)

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The treatment of this disease is difficult and disappointing. The remarkable effect of inhibition of IL-1 has led to new expectations for these patients.[5–7] There is currently no Anakinra available in Argentina.

The interest of this article lies in the presence of a monoclonal IgG variant, together with the clinical and histological lesions and the subsequent progression to multiple myeloma.

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Listeria monocytogenes Meningoencephalitis During the Treatment With Rituximab and Mycophenolate Mofetil in a Patient With Pediatric-onset Systemic Lupus Erythematosus☆

Meningoencefalitis por Listeria monocytogenes *durante el tratamiento con rituximab y micofenolato de mofetilo en una paciente con lupus eritematoso sistémico de inicio pediátrico*

Mr. Editor:

We have pointedly read the excellent review of cases of central nervous system (CNS) due to *Listeria monocytogenes* (*L. monocytogenes*) infection in patients with systemic lupus erythematosu (SLE published by Horta-Baas et al.[1]) and would like to describe an additional case of meningoencephalitis in pediatric onset SLE (pSLE) during treatment with rituximab and mycophenolate mofetil (MMF).

A 12 year old girl from Ecuador, residing in Spain since the age of 4, with a history of uncomplicated hydrocephalus was diagnosed with pSLE in August 2008 at the age of 10, based on fever, anemia, oral ulcers, finger vasculitis, joint pain, pericarditis, pneumonitis, positive ANA and anti-DNA antibodies. She was initially treated with prednisone 2 mg/kg/day in a descending dose and hydroxicloroquine 200 mg/day with a good response but later developed type IV nephritis, pericarditis, constitutional symptoms, neurologic involvement (headache and psychiatric alterations) and type B insulin resistance syndrome (hyperinsulinism, hypoglycemia, acantosis nigricans and polycystic ovaries). She received 3 monthly boluses of cyclophosphamide (500 mg/m^2 iv) between February and April 2009 without improvement and therefore was treated with rituximab (375 mg/m^2/week iv 4 doses) starting in May and, since July, with MMF (1.5 g/day) plus prednisone 35 mg/day, with renal function and disease activity improvement (ΔSLEDAI 18→6), administering a new cycle in October. She underwent an oophorectomy in November because the increase in size of annexal masses carried a high risk of complications or malignity, the latter excluded by histopathological analysis. During this internment the patient presented bacteremia associated to a urinary catheter caused by *Candida parasilopsis* and a relapse in lupus activity, being treated with 3 boluses of methylprednisolone (30 mg/kg iv) and increasing oral prednisone to 45 mg/day (1 mg/kg/day). A month after this episode she came to the emergency room due to fever, forceful vomiting which had lasted for 12 h, with no prior symptoms. At the time she was using the same dose of MMF and prednisone 35 mg/day, but maintained important lupus activity (SLEDAI 10). Upon examination she was hypotensive, with pus coming out of the subclavian catheter insertion site, a reduced consciousness level (Glasgow 13), meningeal irritation signs and asymmetric bilateral paresia of the VI pair. Blood tests showed hemoglobin 10.8 g/dl, leukocytes 20.100/mm^3 (neutrophils 94.5%), C reactive protein 103 mg/dl and procalcitonin 12 ng/dl. Cerebrospinal fluid (CSF) analysis showed pleocytosis (200/mm,^{3} 80% mononuclear and 20% polymorphonuclear), hypoglucorrachia (2 mg/dl), hyperproteinorrachia (1.07 g/l) and a Gram stain that was negative for microorganisms. She was hospitalized, receiving empirical treatment with cephotaxim, vancomycin and iv steroids (methylprednisolone 50 mg/day). *Staphylococcus epidermidis* was isolated in the subclavian catheter and *Staphylococcus aureus* in a blood culture, while the CSF culture was positive for *L. monocytogenes*. The diagnosis of rhomb encephalitis was proposed, due to the neurologic focalization, but the cranial magnetic resonance image (MRI) did not demonstrate compatible alterations on the brain stem or cranial nerves. Antibiotics were changed to gentamycin for 7 days plus ampicillin for 4 weeks, afterward receiving amoxicyllin for two weeks after discharge. Vancomycin was maintained for coverage of catheter associated infection. The patient's consciousness level improved 24 h after hospitalization, but the oculomotor affection took longer, until the fourth week. MMF was suspended definitely, but the patient required 3 additional cycles of rituximab in May 2010, January and June 2011 for control of nephritis. She later presented other infectious complications, including herpes zoster and subdural empiema due to *Streptococcus pneumoniae*, dying as a consequence of the latter in January 2012.

The mean age of the 26 patients in the series of CNS infection due to *L. monocytogenes* in SLE by Horta-Baas et al. was 33.5 ± 11.8 years.[1] The only pediatric case corresponded to a patient of 16 years of age previously treated with steroids, azathioprine and methotrexate, who presented meningitis with a confusional state, bilateral diplopia, nistagmus, VII cranial nerve paralysis and weakness of the upper right extremity.[2] Alterations in consciousness appear in 75% of cases of meningitis de to *L. monocytogenes* and the focal neurologic signs appear in 35%–40%, with the term "meningoencephalitis" being more adequate.[3] Cranial neuropathies most commonly seen affect cranial nerves III, VI, VII, X and XI,[2] but

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