



Letters to the Editor

Rheumatic Polymyalgia With Pleuropericardial Effusion: An Uncommon Association[☆]



Polimialgia reumática con derrame pleuropericárdico: una asociación infrecuente

Dear Editor,

We have carefully read the article by Sánchez Ruiz-Granados et al. Published in your journal and which touched upon a case of Polymyalgia Rheumatica (PMR) and a pleuropericardial effusion.¹ It described the case of an 80-year-old male with joint pain on the scapular and pelvic girdles, with constitutional signs, who presented a pleuropericardial effusion and an elevation of acute phase reactants. Among other diagnosis, elderly onset rheumatoid arthritis (EORA) was ruled out due to the affection of the scapular and pelvic girdles and rheumatoid factor negativity. This conclusion led to our unease and we would like to comment on a series of facts.

PMR is an inflammatory disease of elderly patients characterized by pain and morning stiffness in the cervical region as well as the scapular and pelvic girdles and elevated erythrocyte sedimentation rate. Immunological testing is typically negative and X-rays show no alterations. Due to the absence of specific tests, its excellent response to steroids is considered part of the diagnosis.² Recent echographic and magnetic resonance tests have shown the almost constant presence of extra-articular inflammation in the form of subacromial bursitis or bicipital tendinitis, leading to the inclusion of these alterations in the new diagnostic criteria for PMR.³ As may be understood, the diagnostic difficulty for PMR is inherent in its definition and the absence of specific testing. Therefore, in the differential diagnosis the clinician must take into account other rheumatic and non-rheumatic diseases that may simulate PMR.^{4–6} Among non-rheumatic causes one finds some infections and tumors that in the case at hand were reasonably ruled out. However, among the rheumatic causes, EORA stands out and we consider that it cannot be ruled out. EORA differs from rheumatoid arthritis of younger patients because it affects women with less frequency, has a more acute onset of disease, is more commonly accompanied by constitutional symptoms and, frequently, affects large joints, especially the shoulders, simulating PMR, with less affection of metacarpophalangeal joints. It presents with high erythrocyte sedimentation rate and the percentage of patients with positive rheumatoid factor is reduced in relation to younger patients. Due to these characteristics, its onset may be undistinguishable from PMR.^{7,8}

In the case at hand, although the patient has PMR criteria, the presence of a pleuropericardial effusion leads to doubts on the

diagnosis. The literature describes the association of PMR and pleuropericardial effusion rarely,^{9–13} as commented on by the authors, while the association of EORA –both seronegative and seropositive –with the presence of pleural or pericardial effusion, has been widely described.^{14,15}

On the other hand, as has been commented, a girdle syndrome similar to PMR is seen with relative frequency in EORA due to shoulder affection as a presenting manifestation, along with constitutional syndrome and elevation of acute phase reactants. The good response to steroids is also one of the characteristics shared with PMR.^{7,8} Therefore, contrary to what is commented by the case authors, the clinical presentation is perfectly compatible with an EORA diagnosis. Lastly, it is important to remember that negativity for anti-cyclic citrullinated peptide antibody negativity is also important (ACPA) is also useful to rule out EORA, something not reported by the authors. In any case, the absence of rheumatoid factor (RF) or ACPA would not rule out EORA either, because seronegative forms exist.¹⁶

Therefore, we believe that it is risky to diagnose PMR in the patient described due to its clinical similarity with EORA. It would be important to know if there was peripheral arthritis, ACPA titers and echographic or magnetic resonance evaluation of the patients' shoulders in order to rule out one entity or the other. Obviously, clinical follow up will also contribute fundamental data for a final diagnosis because, in many cases of elderly onset arthritis, disease evolution is the only key to reach a concrete diagnosis.

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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 (+) ($\pm 4.20\%$ 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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