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Antisynthetase syndrome without myositis secondary to AA amyloidosis: a non-described association

Nefrologia 2011;31(1):117-9
doi:10.3265/Nefrologia.pre2010.Sep.10469

To the Editor,

Antisynthetase syndrome (AS) is a rare disease in the idiopathic inflammatory myopathy group and is characterised by the presence of antisynthetase antibodies. The clinical presentation of antisynthetase syndrome is varied and includes polymyositis or dermatomyositis, polyarthritis, diffuse interstitial lung disease, Raynaud's phenomenon and erythematous-violaceous hyperkeratotic skin lesions on metacarpophalangeal and interphalangeal joint areas.[1,2] AS is due to IgG antibodies directed against the enzyme synthase. Seven autoantibodies have been identified: anti-Jo-1, anti-PL7, anti-PL12, anti-OJ, anti-EJ, anti-KS and anti-Wa, with anti-Jo-1 being the best known.

Amyloidosis is a protein metabolism disease characterised by extracellular deposition of fibrillar protein set in beta fold arrangement. The most important are primary amyloidosis (AL), consisting mainly of fragments of light chain of immunoglobulins, and secondary amyloidosis (AA), consisting of protein A1-3 fibrils. Renal involvement is common in secondary amyloidosis, with a wide variety of signs and symptoms: isolated proteinuria, nephrotic syndrome, hypertension, hypotension, renal failure, etc. Amyloidosis secondary to chronic rheumatic diseases are the most common type of secondary amyloidosis.

Only one case of AS and secondary AA amyloidosis has been reported in the literature, but this patient had a lymphoma.[3-5]

Case report

We report the case of a 72-year-old man diagnosed by the rheumatology department with AS antiJo-1 positive without myositic damage and with impaired renal function. This patient had a history of hypertension, interstitial neuropathy, moderate mitral regurgitation and left ventricular hypertrophy with septal birefringence. In a previous hospital stay, the cardiology department performed a subcutaneous fat biopsy after suspicion of amyloidosis, which was negative. In this current hospitalisation, he has been referred for the study of renal failure, with creatinine 2.9mg/dl, proteinuria 1.25mg/24h, and no other biochemical changes. A physical examination revealed the telangiectasias of the eyelids and erythematous-violaceous hyperkeratotic skin lesions (Gottron sign) on the metacarpophalangeal and interphalangeal joints ("mechanic's hands", Figure 1). The remaining physical examination was normal. The autoimmunity study was completed (ANA, negative ANCA, normal C3 and C4), blood and urine immunofixation and serum protein studies were also performed and there were no apparent abnormalities.

A chest x-ray was performed, which showed cardiomegaly at the expense of atria and fissural thickening with right

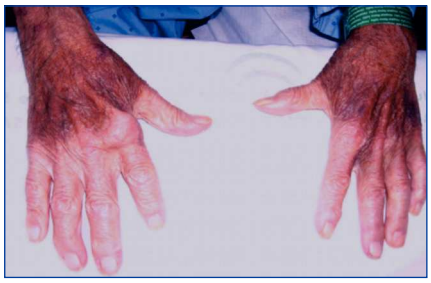

Figure 1. Mechanic's hands, characteristic of antisynthetase syndrome.

costophrenic impingement. An abdominal ultrasound showed the right kidney to be 9.2cm with an echogenic cortical area and a small simple cyst of 1cm in the lower pole. The left kidney was 9.3cm, with similar characteristics. In the upper pole, there was a simple cyst of about 3cm, and one of 1.6cm in the lower pole, with calcified internal septation. It was decided to perform a renal biopsy for diagnosis, and 9 sclerotic glomeruli were seen. Also detected were amorphous deposits in the glomeruli, vessel and interstitial, Congo red-positive and AA amyloid + (IHC) in glomerular-interstitial, nodular and predominantly perivascular areas. The pathologic diagnosis was renal AA amyloidosis with glomerular, interstitial and mainly vascular affectation (Figures 2, 3, 4 and 5).

Discussion

This was an AS without clinical or analytical data of muscle disease or myositic affectation, which is associated with deterioration of renal function. Amyloidosis secondary to chronic rheumatic diseases are now the most common type of secondary amyloidosis caused by deposition of amyloid beta protein. However, there is only one case associated with AS, and that patient also had a lymphoma.

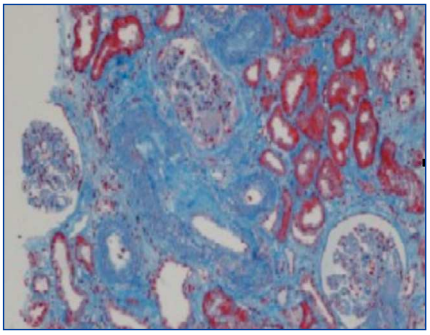

Figure 3. Masson's trichrome showing intense fibrosis.

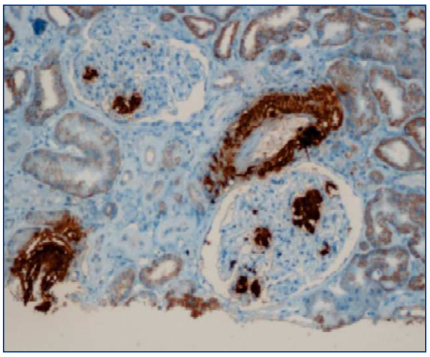

Figure 4. Immunohistochemical study with AA protein deposits in vessels and glomeruli.

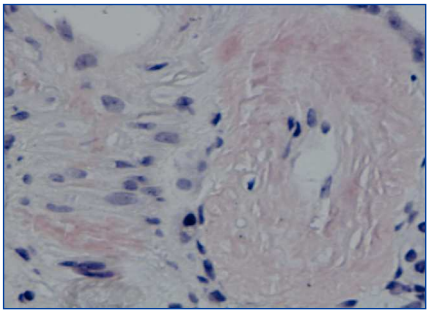

Figure 5. Congo red stain showing the characteristic amyloid deposits in glomeruli and vessels.

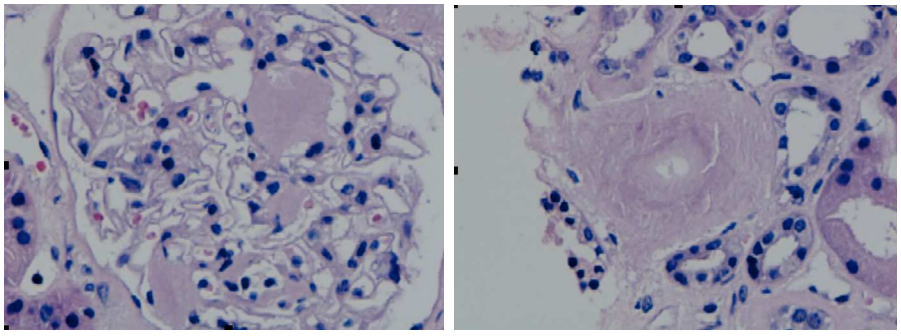

Figure 2. Haematoxylin-eosin test showing amorphous nodules in glomeruli and vessels.

In the literature, we found inflammatory myopathies related with amyloidosis, such as amyloidosis due to inclusion bodies, however, this association is not described in the case of AS.

The treatment of renal amyloidosis was symptomatic. The patient was monitored by the rheumatology department, and treated with methotrexate, risedronate, and calcium and vitamin D supplements. He had a good response and did not need immunosuppressive therapy.[5-7]

As recommended for other long-duration inflammatory diseases, we believe it necessary to perform a renal biopsy in patients with anti-Jo-1 positive antisynthetase syndrome and an unexplained deterioration of renal function.

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Cytomegalovirus colitis

Nefrologia 2011;31(1):119-20
doi:10.3265/Nefrologia.pre2010.Oct.10678

To the Editor,

Our patient was a Bolivian woman diagnosed with systemic lupus erythematosus (SLE) in 2003. In July 2008, she developed type IV lupus glomerulonephritis and treatment was started with i.v. methylprednisolone followed by oral prednisone at doses of 1mg/kg/day and mycophenolate mofetil (MMF). She was hospitalised for a month due to respiratory symptoms. *Nocardia spp* were isolated from the sputum culture, antibiotic treatment begun and immunosuppressive medication was reduced.

During hospitalisation, the patient developed fever, abdominal pain, vomiting and diarrhoea. Culture studies, parasite study, identification of *Clostridium difficile* toxin and antigenaemia for cytomegalovirus (CMV) were performed on two occasions, and the results were negative. An abdominal CT (Figure 1) showed a thickening of the wall of the colon with occlusion of the lumen from the cecum to the rectum-sigmoid junction, which was compatible with diffuse pancolitis. A fibrocolonoscopy was requested (Figure 2), which revealed oedematous mucosa with multiple soft nodular lesions compatible with pneumatosis *coli*. The colon biopsy revealed viral inclusions which were confirmed for CMV by immunohistochemistry (Figure 3). The third antigenaemia determination for CMV was positive and the definitive diagnosis was CMV colitis. Treatment with i.v. ganciclovir was begun and the MMF treatment withdrawn, with clinical improvement and a negative antigenaemia for CMV.

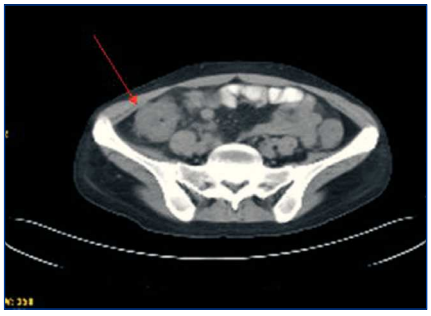

Figure 1. Abdominal TC.

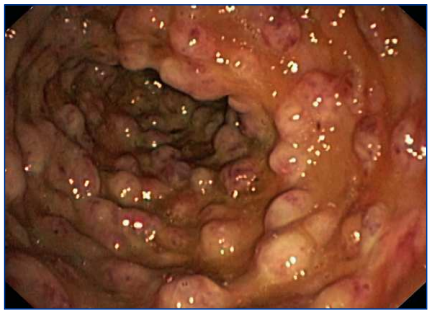

Figure 2. Fibrocolonoscopy.

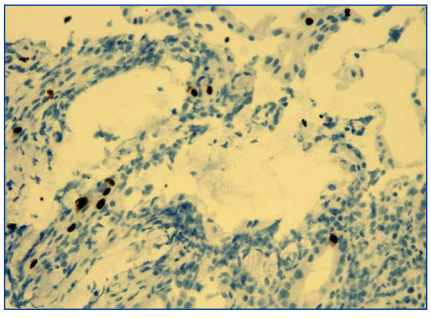

Figure 3. Colon biopsy.

SLE is a chronic autoimmune inflammatory disease of unknown cause with a wide variety of clinical presentations.

It is characterised by an alteration in the immune system which involves the synthesis of autoantibodies and the formation of immune complexes which cause tissue damage, along with the action of inflammatory mediators.

Lupus disease itself and the use of immunosuppressive agents increase the risk of opportunistic infections causing increased morbidity and mortality. In general, patients have bacterial infections, however, there is an increase in viral infections, due especially to CMV, but also to human parvovirus B_{19}, simplex herpes, varicella zoster and hepatitis A.[1]

Viral infections may have symptoms identical to those of SLE, such as malaise, fever, arthralgia, a rash, lymphadenopathy and cytopaenias, so it may be confused with an outbreak of SLE.[2,3] This may lead to increasing immunosuppressive therapy, thereby worsening the latent infection.

In our case, the patient developed an invasive CMV disease despite reducing the immunosuppressive treatment. The persistence of abdominal symptoms without encountering an infectious cause, the finding of non-specific lesions in the colonoscopy and reduced immunosuppression, made us suspect that it was an outbreak of SLE with abdominal involvement. The anatomopathological study allowed us to make the diagnosis of CMV colitis, and administer appropriate treatment to prevent further immunosuppression.

We believe it important to maintain a high degree of suspicion for both bacterial and viral opportunistic infections that mimic outbreaks of lupus disease in immunosuppressed patients. They will benefit from prompt diagnosis and appropriate treatment.