

patients' preferences and the right to receive all information required.

Even though most people over 80 may not benefit from starting dialysis treatment, the decision to start or not start dialysis treatment should be personalised, as it was the case of our patient. The right of each and every individual, regardless of age, to choose their own present and their own future should be respected and valued.

## REFERENCES

1. Swidler MA. Geriatric renal palliative care. *J Gerontol A Biol Sci Med Sci.* 2012;67:1400-9.
2. Murtagh F, Marsh J, Donohoe P. Dialysis or not? A comparative survival study of patients over 75 years with chronic kidney disease stage 5. *Nephrol Dial Transplant.* 2007;22:1955-62.
3. Murtagh F, Burns A, Moranne O, Morton R, Naicker S. Supportive care: comprehensive conservative care in end-stage kidney disease. *Clin J Am Soc Nephrol.* 2016;11:1909-14.
4. Murtagh F, Addington-Hall JM, Edmonds PM, Donohoe P, Carey I, Jenkins K, et al. Symptoms in advanced renal disease: a cross-sectional survey of symptom prevalence in stage 5 chronic kidney disease managed without dialysis. *J Palliat Med.* 2007;10:1266-76.
5. Rebollo-Rubio A, Morales-Asensio JM, Pons-Raventos ME, Mansilla-Francisco JJ. Revisión de estudios sobre calidad de vida relacionada con la salud en la enfermedad renal crónica avanzada en España. *Nefrología.* 2015;35:92-109.

6. Abdel-Kader K, Unruh M, Weisbord SD. Symptom burden, depression, and quality of life in chronic and end-stage kidney disease. *Clin J Am Soc Nephrol.* 2009;4:1057-64.
7. Ley 41/2002, de 14 de noviembre, básica reguladora de la autonomía del paciente y de derechos y obligaciones en materia de información y documentación clínica («BOE» núm. 274, de 15 de noviembre de 2002. Referencia: BOE-A-2002-22188).

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# Development of C3 glomerulopathy in a patient with acquired partial lipodystrophy<sup>☆</sup>

## Desarrollo de glomerulopatía C3 en paciente con lipodistrofia parcial adquirida

Dear Editor,

Dysregulation of the activation of the alternative pathway of the complement is involved in the development of C3 glomerulopathy (C3G), a high percentage of cases are positive for the C3 nephritic factor autoantibody (anti-C3Nef),<sup>1</sup> the cause of the complement abnormality. This antibody may be associated with adipose tissue abnormalities, causing acquired partial lipodystrophy (APL),<sup>2</sup> which may appear before or after the onset of C3G.

We present the case of a 52-year-old man with a medical history of hypertension and APL of recent onset, being treated with enalapril 5 mg/24 h. He was referred to a nephrol-

ogy clinic due to detection of albuminuria with levels that had been rising for years. The patient was asymptomatic. Physical examination revealed a lack of adipose tissue on the cheekbones, neck, upper limbs and trunk; normal blood pressure; and no oedema in the lower limbs. Given the differential diagnosis of APL, we ruled out causes associated with panniculitis, autoimmune diseases such as Barraquer-Simons syndrome, acquired generalised lipodystrophy (Lawrence syndrome), membranous and membranoproliferative glomerulonephritis (GN) (following kidney biopsy), drugs (cortisol, insulin), and viral infections such as HIV infection. Complementary tests showed that the patient had normal kidney function with creatinine 0.89 mg/dl, urea 38 mg/dl, sodium 139 mEq/l,

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potassium 4.2 mEq/l, total protein 6.7 mg/dl, total cholesterol 240 mg/dl, decreased C3 < 15 mg/dl and C4 28.1 mg/dl, and negative immunoglobulins, serum protein electrophoresis, serology (HIV, HCV, HBV) and autoimmunity (ANA, ANCA, APO, anti-PLA2R). Urine testing revealed microhaematuria, albuminuria 3.7 g/24 h and proteinuria up to 3.8 g/24 h. A kidney ultrasound showed kidneys of a normal shape and size with normal differentiation between the cortex and the medulla and no other findings. Antiproteinuric therapy was intensified, but the patient's proteinuria remained in the nephrotic range during the follow-up. A kidney biopsy revealed pathology findings of granular deposits in the membrane with spread to the mesangium, with a widespread and diffuse pattern and positivity for C3 (3+), C4d (2+) and fibrinogen (1+), with a more segmental pattern and intensity  $\pm 1$  for IgM and lambda light chains. IgG, IgA, C4, C1q and kappa light chains were negative. The histology diagnosis was C3G with a lesion pattern more similar to membranous GN than to mesangiocapillary GN.

Anti-C3Nef autoantibodies were detected in serum (IdiPAZ).

Treatment was started with corticosteroids on a down-titration regimen and mycophenolate mofetil 1 g/12 h. At present, the patient has normal kidney function with slight improvement in his albuminuria and proteinuria, 2.5 g/24 h and 2.8 g/24 h, respectively.

APL is a very uncommon disease characterised by a gradual loss of adipose tissue of the head, neck, trunk and upper limbs.<sup>2</sup> Up to 75-90% of patients have decreased levels of the component C3 of the alternative complement pathway linked to the presence of anti-C3Nef, for which more than 80% of patients test positive.<sup>1,3</sup>

The most common form of presentation of APL associated with C3G is dense deposit disease,<sup>4</sup> although in some cases the histology study shows a histology pattern similar to IgA GN.<sup>5</sup> We believe that our case is interesting first due to the diagnosis of C3G with a histology pattern similar to membranous GN, this type of presentation has not been previously reported, and second due to the development of APL, probably subsequent to kidney involvement.

In C3G case cohorts, only a limited percentage of patients have APL.<sup>6</sup> However, among patients with lipodystrophy, up to 25% may develop C3G in the medium to long term.<sup>2,4,7</sup> In these cases, the common mechanism is dysregulation of complement system, although the possibility that other associated factors are involved have yet to be determined and cannot be ruled out.<sup>8</sup> In conclusion, C3G is a rare disease, and in the presence of signs such as lipodystrophy, a proper differential

diagnosis and follow up of these patients is essential to prevent or promptly intervene should nephropathy develop, if not present already.

## REFERENCES

1. Levy Erez D, Meyers KE, Sullivan KE. C3 nephritic factors: a changing landscape. *J Allergy Clin Immunol.* 2017;140:57-9.
2. Garg A. Acquired and inherited lipodystrophies. *N. Engl J Med.* 2004;350:1220-34.
3. Hussain I, Garg A. Lipodystrophy syndromes. *Endocrinol Metab Clin North Am.* 2016;45:783-97.
4. Misra A, Peethambaram A, Garg A. Clinical features and metabolic and autoimmune derangements in acquired partial lipodystrophy: report of 35 cases and review of the literature. *Medicine (Baltimore).* 2004;83:18-34.
5. de Lucas-Collantes C, Pozo-Román J, Aparicio-López C, de Prada-Vicente I, Argente J. Acquired partial lipodystrophy (Barraquer-Simon syndrome) and IgA nephropathy. *Nefrología.* 2016;36:556-8.
6. Vargas R, Thomson KJ, Wilson D, Cameron JS, Turner DR, Gill D, et al. Mesangiocapillary glomerulonephritis with dense «deposits» in the basement membranes of the kidney. *Clin Nephrol.* 1976;5:73-82.
7. Sissons JG, West RJ, Fallows J, Williams DG, Boucher BJ, Amos N, et al. The complement abnormalities of lipodystrophy. *N Engl J Med.* 1976;294:461-5.
8. Corvillo F, López-Trascasa M. Acquired partial lipodystrophy and C3 glomerulopathy: dysregulation of the complement system as a common pathogenic mechanism. *Nefrología.* 2018;38:258-66.

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