

Original article

Calciophylaxis: Beyond CKD-MBD[☆]

María Fernández, Enrique Morales*, Eduardo Gutierrez, Natalia Polanco, Eduardo Hernández, Eva Mérida, Manuel Praga

Servicio de Nefrología, Hospital Universitario 12 de Octubre, Madrid, Spain

ARTICLE INFO

Article history:

Received 1 November 2016

Accepted 6 February 2017

Available online 28 September 2017

Keywords:

Calciophylaxis

Chronic renal failure

Mortality

Non-renal forms

Oral anticoagulants

ABSTRACT

Introduction: Calcific uraemic arteriopathy (CUA), also called calciophylaxis, is a rare but potentially fatal vascular disorder that almost exclusively affects patients with chronic renal failure. The objective of this study was to analyse various risk factors for developing CUA and its subsequent clinical course according to the treatment received.

Materials and methods: A retrospective study that included patients diagnosed with CUA from December 1999 to December 2015. Various risk factors, clinical course and treatment options were analysed.

Results: A total of 28 patients (53.6% females) with a mean age of 67.2 ± 11.8 (38–88) years were included. At the time of diagnosis, 53.6% were on haemodialysis, 25% were kidney transplant patients and 21.4% had normal renal function. The use of steroids (100%, $p = 0.001$) was the main risk factor in renal transplant patients. Skin lesions resolved in 60.7% (especially in those receiving multitargeted therapy). Patient survival at 12 months was 29% in transplant patients, 57% in haemodialysis patients and 100% in normal renal function patients (log-rank 6.88, $p = 0.032$). Chronic renal failure ($p = 0.03$) and hypoalbuminaemia ($p = 0.02$) were the main risk factor for CUA mortality.

Conclusions: Although the incidence of CUA remains low, CUA mortality is very high, special attention to its occurrence in kidney transplant patients and “non-renal” CUA forms is required. Oral anticoagulants and steroids appear to be the main risk factors, CUA is a challenge; a registry of patients and determining standard therapy are required.

© 2017 Sociedad Española de Nefrología. Published by Elsevier España, S.L.U. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

[☆] Please cite this article as: Fernández M, Morales E, Gutierrez E, Polanco N, Hernández E, Mérida E, et al. Calciophylaxis: más allá de CKD-MBD. Nefrología. 2017;37:501–507.

* Corresponding author.

E-mail address: emorales@senefro.org (E. Morales).

2013-2514/© 2017 Sociedad Española de Nefrología. Published by Elsevier España, S.L.U. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Calcifilaxis: más allá de CKD-MBD

R E S U M E N

Palabras clave:

Calcifilaxis
 Insuficiencia renal crónica
 Mortalidad
 Formas no renales
 Anticoagulantes orales

Introducción: La arteriopatía urémica calcificante (CUA), también llamada calcifilaxis, es un trastorno vascular poco frecuente pero potencialmente mortal que afecta casi exclusivamente a pacientes con insuficiencia renal crónica. El objetivo de este estudio fue analizar los diferentes factores de riesgo para el desarrollo de CUA y su posterior evolución según la terapia recibida.

Material y métodos: Estudio retrospectivo que recoge aquellos pacientes con diagnóstico de CUA desde diciembre de 1999 hasta diciembre de 2015. Se analizaron diferentes factores de riesgo, evolución y diferentes opciones terapéuticas.

Resultados: Se incluyeron 28 pacientes (53,6% mujeres) con una edad media de $67,2 \pm 11,8$ años (38-88). En el momento del diagnóstico, el 53,6% estaba en hemodiálisis, un 25% eran pacientes con un trasplante renal y el 21,4% presentaba función renal normal. En los pacientes trasplantados, el consumo de esteroides (100%; $p=0,001$) fue el principal factor de riesgo. La resolución de lesiones cutáneas se produjo en el 60,7% (especialmente en los que recibieron tratamiento *multitarget*). La supervivencia de los pacientes a los 12 meses fue de 29, 57 y 100% en los pacientes trasplantados, hemodiálisis y con función renal normal respectivamente (log-rank 6,88; $p=0,032$). La presencia de insuficiencia renal crónica ($p=0,03$) e hipoalbuminemia ($p=0,02$) fueron los principales factores de riesgo de mortalidad CUA.

Conclusión: Aunque la incidencia de la CUA sigue siendo baja, su mortalidad es muy elevada, por lo que debe prestarse especial atención a la presentación de la CUA en los trasplantados renales y en las formas «no renales». Los anticoagulantes orales y los esteroides aparecen como principales factores de riesgo. La CUA es un reto: necesitamos un registro de nuestros pacientes y establecer una terapia estándar.

© 2017 Sociedad Española de Nefrología. Publicado por Elsevier España, S.L.U. Este es un artículo Open Access bajo la licencia CC BY-NC-ND (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Introduction

Calciphylaxis, also called calcific uraemic arteriopathy (CUA), is an uncommon but serious condition with a high rate of mortality.^{1,2}

CUA develops in the dermis and subcutaneous tissue, generally in areas of adiposity. It is characterised by pruritic or painful skin lesions, purplish nodules, ulcers and sores that develop into ischaemic necrosis of the skin.³ A histological examination may reveal medial calcification and intimal proliferation in the small arteries, with or without endovascular fibrosis, extravascular calcification and vascular thrombosis.⁴

It occurs more frequently in patients with advanced chronic kidney disease on dialysis or in renal transplant patients; however, cases of calciphylaxis have been reported in subjects with normal renal function, classified as “non-renal” CUA.⁵

The pathogenesis of this vascular disease is complex and not well understood, but there are several mechanisms thought to be involved in its development. Over the years, other risk factors have been added to the traditional factors of bone and mineral metabolism (hyperparathyroidism, hypercalcaemia and hyperphosphataemia) and its high prevalence in patients with low bone metabolism.^{6,7} These include: female gender, diabetes, obesity, local trauma, hypoalbuminaemia, hypercoagulable states and exposure to active vitamin D, calcium chelators, corticosteroids and vitamin K

antagonists.⁸⁻¹¹ This amalgam of factors has generated a new patient phenotype that shows lesions from calciphylaxis that are unlike the classic image of this condition.

Despite the poor prognosis of this condition, the use of multidisciplinary treatment in recent years (treatment of pain and of superinfections, intensification of haemodialysis, bisphosphonates, sodium thiosulfate, non-calcium chelators, cinacalcet, vitamin K and pentoxifylline) has seen survival increase in these patients.¹²⁻¹⁴

The objective of this study was to analyse the current epidemiology of calciphylaxis in the region of Hospital 12 de Octubre, the risk factors for its development and the patients' clinical courses depending on the treatment received.

Material and methods

An observational and retrospective study was performed on a series of patients diagnosed with calciphylaxis between December 1999 and December 2015 at Hospital Universitario 12 de Octubre in Madrid.

The following data were recorded: gender, age, Charlson comorbidity index, status at diagnosis (patients on haemodialysis, with a normally functioning renal transplant or without kidney disease), aetiology of chronic kidney disease (CKD), time to renal replacement therapy (haemodialysis and renal transplantation), cardiovascular risk factors

Download English Version:

<https://daneshyari.com/en/article/8774191>

Download Persian Version:

<https://daneshyari.com/article/8774191>

[Daneshyari.com](https://daneshyari.com)