THE MANY FACES OF ATYPICAL HAEMOLYTIC-UREMIC SYNDROME

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Introduction

Haemolytic uremic syndrome (HUS) is caractherized by haemolytic microangiopathic anaemia and uremia with signs or symptoms in other organs. But the presentation is not always so clear and is important have in mind this diagnosis in order to make a good differential diagnosis and start prompt therapy.
We describe 4 patients with aHUS with disctint presentation ways and different evolution, all with good clinical response to eculizumab

Case 1

Family history

Sister diagnosed of membranoproliferative glomerulonephritis at 14 years old. She developed progressive renal insufficiency and started peritoneal dialysis on 2010. Then she presented neurological impairment and died on august, 2012 due to respiratory infection. Not definitive diagnosis about underliying disease was reached despite necropsic study

Patient history

At 12 months age she presented anaemia and kidney failure with spontaneous remission without secuelae

15 years old: non nephrotic proteinuria

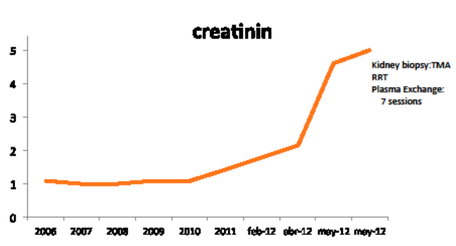
Arterial hypertension, anaemia without haemolytical changes. ESA therapy and antiproteinuric agents

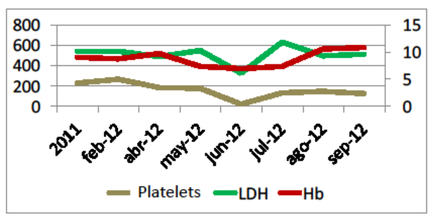
KIDNEY BIOPSY

Mesangial IgM Glomerulonephritis

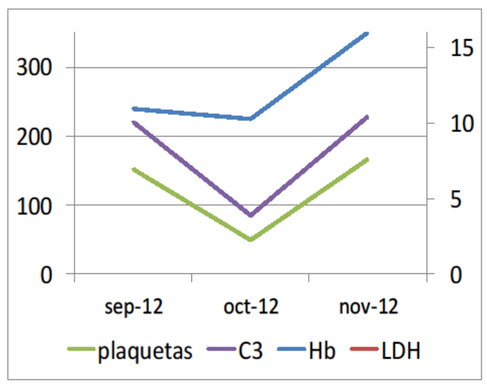
26 years old

Proteinuria and anaemia impairment, worst arterial hipertensión control, increase in serum creatinine (from 0.7 to 2 mg/dl)





On dialysis…



Loss of memory
Car crash due to absence crisis

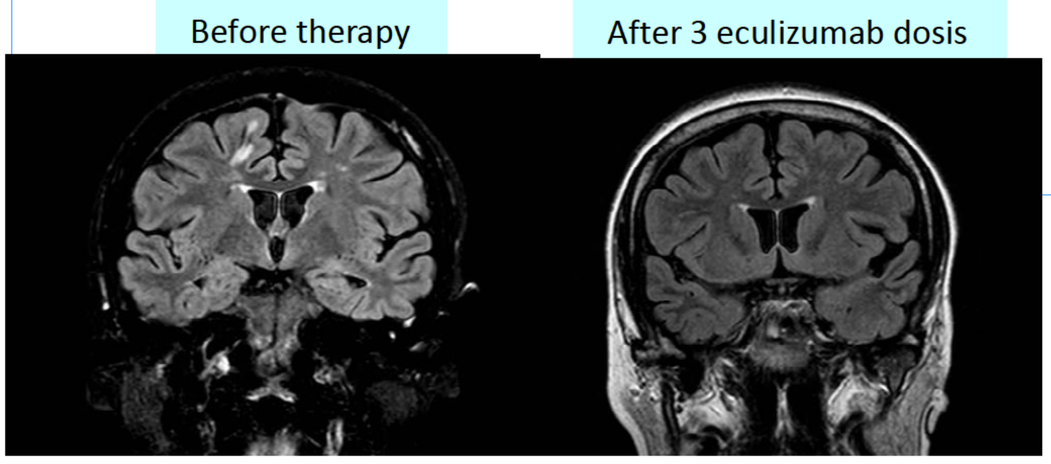
Haptoglobin <5
Schistocytes 1-2/hpf
ADAMST 13 normal

Plasma Exchange. 3 sessions

Eculizumab

Before therapy

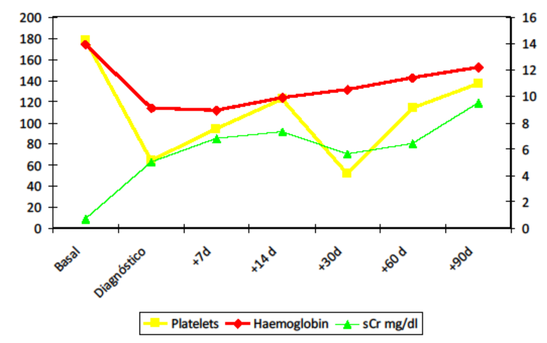
After 3 eculizumab dosis

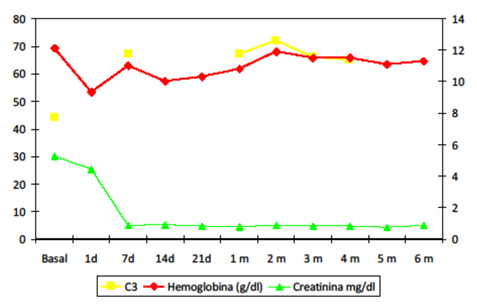


She received a donor kidney transplant, without aHUS recurrence, maintaining eculizumab therapy

Case 2

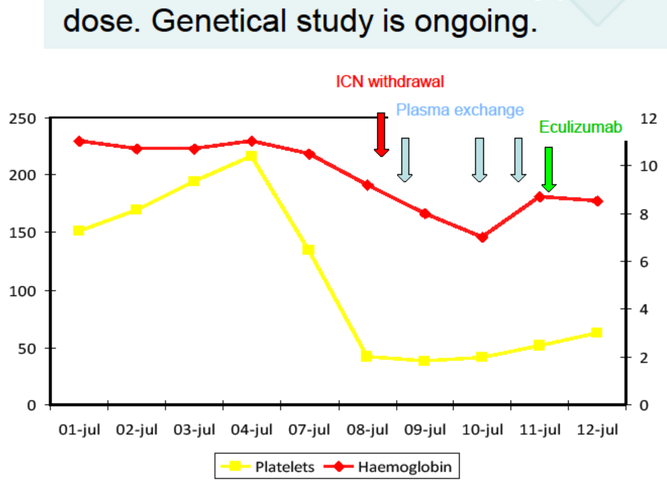
Woman 32 years old. Acute haemolytic microangyopathic anaemia and renal failure that evolved to end stage renal disease in one week. She started haemodialysis, with clinic stability during five years. She presented complement Factor I mutation (reccurrence moderate risk after transplant). When eculizumab was disposable, she was transplanted from a cadaveric kidney donor, receiving profilactic eculizumab. After 2 years of transplant, the patient remains stable

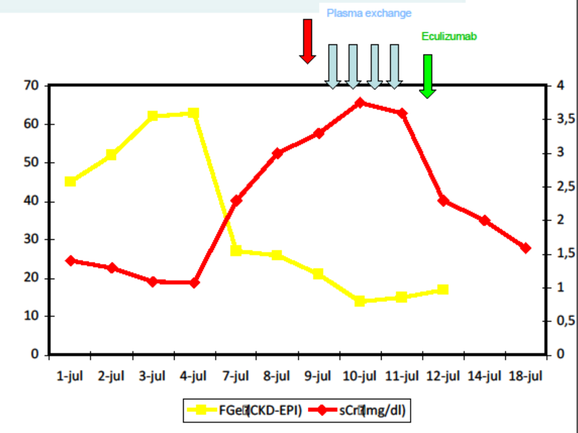




Case 3

Woman 43 years old. Kidney transplant due to ESRD of unknown origin. Excelent evolution the first week after transplant, reaching sCr 1 mg/dl. On the 2nd week she developed thrombotic microangiopathy without response to calcineurin inhibitor withdrawal and plasma exchange (7 doses). Then eculizumab was started, resolving postransplant TMA after one single dose. Genetical study is ongoing.





Case 4

Man, 34 years old. Admitted to ICU for septic shock due to E. Coli infection. After antibiotic and supportive therapy (including hdvvc), the patient presented stablished fidney failure requiring dialysis, coma grade III of unknown origin and anaemia without haemolytic signs. 6 months after initial episode eculizumab therapy was started. 3 weeks after, progressive neurological improvement was observed, but he remains on dialysis. The patient is now on peritoneal dialysis, independent for daily life activities. Genetic study is ongoing

CONCLUSIONS

- aHUS can be presented in different ways (abrupt onset TMA, “smoldering” TMA, making diagnosis difficult
- The kidney is one of the main organs affected, but, as in two of the cases presented, extrarrenal, mainly neurological involvement can be present
- Eculizumab is an effective treatment for aHUS, classic or smoldering
- Sometimes is difficult to distinguish between aHUS and seccondary TMA (as in kidney trasnplant), but eculizumab can be effective in the management of both cases
- The duration of eculizumab therapy is yet a question to resolve

REFERENCES:

1. Loirat C, Frémeaux-Bacchi V. Atypical haemolytic uremic syndrome. Orphanet J Rare Dis 2011; 6: 60
2. Avila et al. Clin Kid J. Clin Kidney J. 2015 Apr;8(2):232-6

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