CRITICAL MANAGEMENT OF THE PREGNANT PATIENT WITH AN ATYPIC HEMOLYTIC UREMIC SYNDROME

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Background:

Up to 86% of patients with hemolytic uremic syndrome (HUS) secondary to pregnancy have **mutations in the alternative pathway of complement**, the underlying disease being an atypical HUS (aHUS)². It is an **ultra-rare** disease that usually evolves to a terminal chronic renal failure, which makes it an easily underdiagnosed and **potentially lethal pathology**¹.

Case Report:

Pregnant woman of 19+4 weeks that consults in the Emergency Room for right lumbar pain, micturition syndrome and fever. The sediment is compatible with urinary tract infection, which is corroborated with abdominal ultrasounds. *E. coli* is isolated, both in blood and urine cultures, so it is admitted under suspicion of acute obstructive pyelonephritis. The physical state and the analytical tests corroborate severe sepsis. The patient is transferred to the intensive unit care with perfusion of Noradrenaline at 0.8mg/h.

It appears hemolytic intravascular anemia with negative Coombs, worsening thrombocytopenia and renal function. The schistocyte visualization, with negative Adamts13, confirms **aHUS**. Immunoglobulin 1g/kg is administered, achieving normalization in 14 days. In week 38+6, cesarean section is scheduled, which takes place without incidents, under intradural anesthesia.

Discussion:

aHUS is an entity that is defined by the clinical triad of non-autoimmune hemolytic anemia, thrombocytopenia and acute renal failure. The choice of the anesthetic technique in **pregnant women with thrombocytopenia** is multifactorial. The **number of circulating platelets and their function** determines the **safety of regional anesthesia**³. Subarachnoid blockade with small caliber needles makes it a much less traumatic technique and a good choice for patients with aHUS. General anesthesia will be a good alternative in caesarean sections if there is evidence of bleeding or coagulation alteration.

Key Point:

In every pregnant woman affected by HUS, a **thorough preanesthetic evaluation** must be carried out, weighing the advantages and disadvantages of the different anesthetic techniques. The safest anesthetic management depends on a deep knowledge of the disease and its implications.

Bibliografía:

¹ Josep M. Campistol et al. Actualización en síndrome hemolítico urémico atípico: Diagnóstico y tratamiento. Revista Nefrología 2013;33(1):27-45

² Saad AF et al. Pregnancy-Associated Atypical Hemolytic-Uremic Syndrome. AJP Rep. 2016 Mar 6(1):e125-8

³ Kam PC. Thrombocytopenia in the parturient. Anaesthesia. 2004 Mar;59(3):255-64

Tabla 1. Clasificación etiológica de las microangiopatías trombóticas

PTT asociada a alteraciones genéticas o inmunes del ADAMTS13 (actividad $\leq 5\%$)

- Genéticas
- Anticuerpos (asociado al tratamiento con ticlopidina y clopidogrel)
- SHU asociado a infecciones (STEC y STEC-like)
- SHU por infección de STEC (VTEC) cepa O157:H7 y otras cepas no O157:H7, *Shigella dysenteriae* type I
- SHU asociado a infección por *Streptococcus pneumoniae* (neuraminidasa)

SHU atípico asociado con alteraciones genéticas o inmunes del sistema del complemento

- Mutaciones en CFH, MCP, CFI, THBD, CFB y C3
- Anticuerpos anti-CFH

MAT secundarias

- A embarazo
- Síndrome HELLP
- Posparto
- A enfermedades:
- Sistémicas
- LES

Síndrome antifosfolípido

Esclerodermia

- Otras

Infección por VIH

Glomerulopatías

Hipertensión arterial maligna

Infección por H1N1 (influenza A)

Neoplasias

Aciduria metilmalónica con homocistinuria

- A tratamientos

- Quinina

- Mitomicina C

- Gemcitabina

- Cisplatino

- Radiación ionizante

- Interferón

- Inhibidores de VEGF y tirosina quinasa (sunitinib, imatinib y dasatinib)

- Ticlopidina y clopidogrel

- Inhibidores de la calcineurina (ciclosporina, tacrolimus)

- Sirolimus

- Valaciclovir

- Anticonceptivos orales

- Al trasplante de órgano sólido o de médula ósea

VAGINAL BIRTH:

- If platelet count > 70.000 and there is no other contraindication: Locoregional anesthesia
- If platelet count < 70.000 and / or contraindication of neuraxial block: Opioids systemically

CAESAREAN SECTION:

- If platelet count > 70.000 and there is no other contraindication: Locoregional anesthesia
- If platelet count < 70.000 and / or contraindication for neuraxial block: General anesthesia